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FOREWORD

It is with great pleasure that we are sharing our first edition of this medical bulletin - Zulekha Healthcare Medical Journal. This is an endeavour from the medical fraternity at Zulekha Healthcare Group including contributions from clinical experts at Zulekha Hospital, UAE and Alexis Multispeciality Hospital, India. With our facilities in UAE and India equipped to be super-hubs for critical healthcare, the journal includes a collection of case studies and experiences of our expert clinicians that will help global healthcare professionals and patients.

The Zulekha Healthcare Medical Journal aims to advocate the highest quality of medical care, enable efficient decision making in clinical practices, educate clinical and non-clinical audience and showcase our high-end capabilities. The journal is a platform to present key subjects and innovative advancements in healthcare.

Amidst the global pandemic qualms, it is a privilege and responsibility of healthcare providers to emerge stronger with significant discoveries. We believe the rough terrains always make us stronger and create opportunities to excel and better manage complex diseases. Clinicians have certainly excelled in their quality of work and endorse their abilities to manage high-risk procedures as a team. While we hope this educational resource helps all of you reading it, we invite you to contribute towards its development and in turn ensure continued improvement of the global healthcare system.

Wishing you well,

Taher Shams
Managing Director
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COLONIC GANGRENE FOLLOWING BEVACIZUMAB – A RARE COMPLICATION

A case report by **Zulekha Hospital,** Dubai

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ABSTRACT

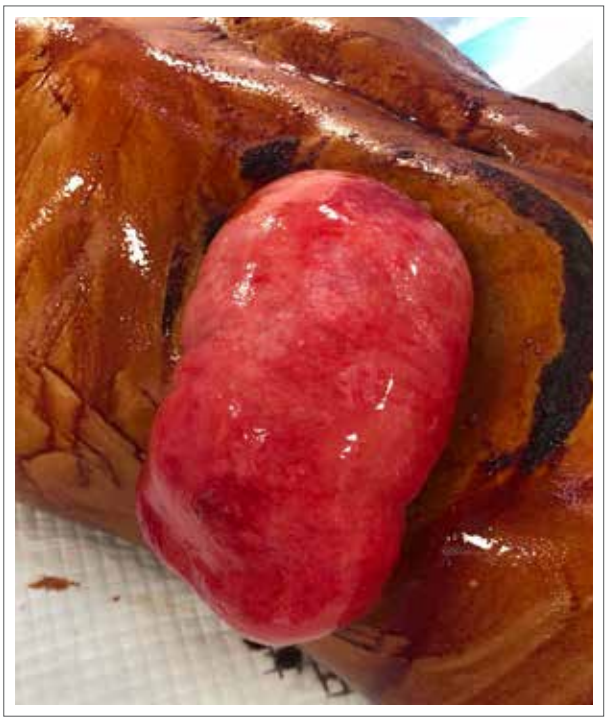
Recently, Bevacizumab has become an essential drug for the treatment of metastatic colorectal cancer. Molecularly targeted agents such as Bevacizumab can cause life-threatening adverse effects, though they are generally considered less toxic than cytotoxic drugs. Here we review a case of a 36-year-old male who experienced bowel necrosis after three days of administration of Bevacizumab and then was taken for emergency life-saving surgery.

Bevacizumab is a monoclonal antibody against vascular endothelial growth factor, which is a crucial element for tumour angiogenesis. Although an excellent safety profile against standard chemotherapy has been demonstrated for the drug, specific rare side effects such as arterial thromboembolism and gastrointestinal perforations have been reported.(1) We present a young male treated with Bevacizumab added to 5 fluorouracil and Irinotecan who developed bowel ischaemia after three days of treatment. Combination of cytotoxic agents and Bevacizumab is the most frequently used regime for the treatment of metastatic colorectal cancer.(2) Combination with Bevacizumab helps in improving response rates to chemotherapy, progression-free survival (PFS) and overall survival (OS) compared to treatment with cytotoxic agents alone.(3,4)

PATIENT DESCRIPTION

We present a 36-year young male who was diagnosed as a case of carcinoma colon in September 2018. He presented with intestinal obstruction for which emergency laparotomy with Hartman's procedure and end transverse colostomy was done. The mass was near splenic flexure. His postoperative histopathology confirmed the presence of poorly differentiated carcinoma, margins negative, pT4N2aMx disease with 4/14 lymph nodes positive for metastasis. During staging CT scan of chest and abdomen, he was found to have metastasis in multiple areas of the abdomen (liver and peritoneal cavity). He was given six cycles of CAPOX regime (oxaliplatin and capecitabine) with a plan for maintenance capecitabine later on. However, in March 2019, he developed the progression of the disease (adrenal metastasis with abdominal nodal metastasis) with rising CEA marker. Metastasis was evident on CT scan of the abdomen and pelvis. CT chest was unremarkable.

The patient was put on the second line FOLFIRI regime (5 fluorouracil with leucovorin with Irinotecan) along with Bevacizumab in the standard dose of 5 mg/kg body weight every 2 weeks. The patient received the fifth cycle of chemotherapy on 12th September 2019. However, three days later, late in the night the patient presented with severe abdominal pain, nausea and vomiting and was noted to have herniation of gangrenous bowel loop through the stoma. He was afebrile, pulse rate 96/min, respiratory rate 22/min and his blood pressure



143/67 mm of Hg. His pain score was 8. The patient had lower abdominal tenderness. The patient was immediately taken for surgery and underwent right-sided hemicolectomy, resection of gangrenous bowel loop, adhesiolysis and terminal end ileostomy. Intraoperative findings included gangrenous distended edematous colon with stomal herniation of around 15 cms and multiple inter bowel and mesenteric adhesions. No grossly visible peritoneal or nodal disease was noted. The cut end of the rectum was seen in the pelvis with no evidence of disease. There was no ascitic fluid. Small bowel loops were densely adherent with peritoneum and anterior abdominal wall. Entire small bowel loops were mobilised, and integrity was checked, and right hemicolectomy was done with end ileostomy. The patient was discharged on the fifth postoperative day. Post-operative histopathology showed patchy mucosal necrosis with haemorrhage, submucosal edema and vascular congestion with serosal edema. Margins were viable. Six reactive pericolic lymph nodes with sinus hyperplasia were seen, and no signs of malignancy were seen. Later on, the disease evaluation showed stable disease, and now patient is on follow-up with maintenance treatment.

DISCUSSION

In the case presented here, extensive bowel necrosis developed three days after the last treatment with Bevacizumab. Bevacizumab acts as an inhibitor of vascular endothelial growth factor which mediates angiogenesis. Thus by inhibiting angiogenesis, it promotes tumour regression and necrosis. This results in the more effective delivery of chemotherapy.(5) But given that Bevacizumab may impair wound healing, it may contribute to cytotoxic necrosis that may progress from mucosal layers through the full layers of the bowel wall. In our case the pathological report suggested patchy mucosal necrosis.

Life-threatening adverse effects by Bevacizumab are rare but well documented which include gastrointestinal perforation caused by trans mural necrosis, a serious condition induced by mucosal damage. In a meta-analysis of 17 randomised control trials, the incidence of bowel perforation was 0.9% (95% CI -0.7 to 1.2). Among patients receiving Bevacizumab, a twofold increased risk for perforation was noted with a mortality rate of 21.7% (95% CI 11.5-37).(6)

High-risk factors associated with bowel perforation with Bevacizumab are intact primary tumour, history of colonoscopy within one month of initiation of Bevacizumab therapy, pelvic radiotherapy, and those who receive higher doses of Bevacizumab 5 mg / kg /week.(6,7) From the report of 14 cases of Bevacizumab related gastrointestinal necrosis or perforation identified between 2003 to 2013 (8 to 16), surgical treatment was performed in ten cases, drainage in one case and conservative therapy in three cases.

The detailed mechanism of Bevacizumab related gastrointestinal perforation remains unclear. Likely aetiologies are tumour invasion(6) and damage of the structure and function of gastrointestinal vasculature resulting in ischaemic perforation.(6) Another report suggests inhibition of vascular endothelial growth factor mediated release of nitric oxide which might lead to vasoconstriction.(17) Thus, Bevacizumab has the potential to cause gastrointestinal ischaemia. Hence, we might hypothesise that insufficient blood supply caused by antiangiogenic treatment might have led to the bowel gangrene.

COMMENT

Based on our case and other reported cases of unexplained colonic necrosis post Bevacizumab, caution should be exercised in adding Bevacizumab to therapy. Colonoscopy should be deferred if possible, for one month after Bevacizumab. A high index of suspicion should be there when the patient presents with abdominal symptoms post Bevacizumab to rule out fatal complication like colonic necrosis or perforation. This case highlights the importance of teamwork and excellent communication between onco-physician and onco-surgeon.

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COVID-19 POSITIVE PATIENT WITH NODULAR SCLERITIS AND CONJUNCTIVITIS

A case report by **Zulekha Hospital,** Sharjah

Dr. Arif Adenwala
Specialist Ophthalmologist

SUMMARY

This case highlights a unique presentation of a COVID-19 positive patient with Nodular Scleritis with Conjunctivitis as a presenting feature. Patient initially had ocular symptoms and developed only mild systemic features subsequently which did not require hospitalisation. COVID testing done at different time points showed variable results which correlated with the ocular features. This patient was followed up during quarantine using tele-ophthalmology and hence highlights the importance of tele medicine during these unprecedented times. Several clinical studies have reported the presence of SARS-CoV-2 in tear specimens from individuals with COVID-19.(1) There has been discordance in reports regarding the proportion of COVID-19 patients with presence of virus in ocular specimens, possibly relating to factors including sensitivity of tests, type of ocular specimen and timing of specimen procurement in patients. In a large study including 1099 hospitalised patients with laboratory-confirmed COVID-19 from 30 hospitals in China, conjunctival congestion was documented in 9 patients (0.8%).(2) In another study of patients diagnosed clinically as COVID-19, one third had ocular symptoms and signs, including conjunctival hyperaemia, chemosis and epiphora.

Early recognition and detection of these cases can help in adequate protection and reduced transmission of the disease.(3,4) Although viral infection of ocular cells has not yet been reported in patients, a recent report found that SARS-CoV-2 can infect conjunctival epithelium in an ex-vivo culture system.(5)

This case report highlights an unusual presentation of a patient positive for COVID-19 and the need for treating every patient with red eye in this pandemic with adequate precautions to prevent inadvertent cross infection.

CASE REPORT

A 39-year-old male patient presented with redness and pain in the right eye for over 2 days. On examination, the patient had minimal chemosis with congestion in the right eye. He was diagnosed as conjunctivitis (Figure 1) and prescribed topical 0.5% moxifloxacin eye drops 4 times a day in the right eye. Since conjunctivitis has been described as a possible clinical feature in patients of COVID-19, the patient was tested for SARS-CoV2 with Real time-polymerase chain reaction (RT-PCR) on nasopharyngeal swab sample.

His symptoms increased and he had pain and redness in the eye. He was found to have localised congestion and swelling in the superomedial quadrant of the right eye and diagnosed to have possible nodular scleritis clinically. Nasopharyngeal swab sample was taken for SARS-Cov2 RT-PCR and was positive for COVID infection.

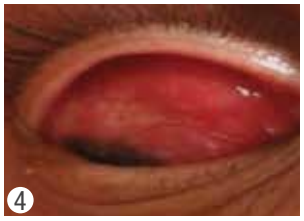
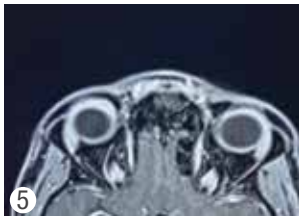
1: Initial presentation with conjunctival congestion
2: Conjunctival chemosis
3: Normal left eye

Patient was asymptomatic and had already taken oral antibiotics and so no oral treatment as given. Since patient had only mild symptoms, he did not require hospitalisation and was advised home quarantine.

Patient was started on Topical Betadine 0.25% drops made with tears eye drops every 8 hours. Systemic blood investigations to look for underlying autoimmune disorders which could cause a nodular scleritis

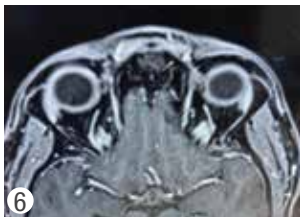
(Complete blood count, Random blood sugars, RA factor, ANA, c-ANCA, p-ANCA, ESR, CRP, urine microscopy) were advised and found to be within normal limits. Patient was followed up via telemedicine during the quarantine period.

Examination of the right eye via teleophthalmology showed increased conjunctival congestion with worsening of the nodular inflammation. Owing to non-resolving inflamed conjunctival/episcleral nodule an MRI orbit was advised to look for other possible causes of such a nodule like neoplasms and to rule out orbital extension. MRI contrast reported as a nodular lesion with scleral thickening and a possible inflammatory aetiology suggesting a diagnosis of Nodular Scleritis.

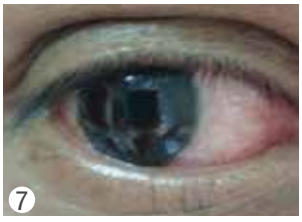
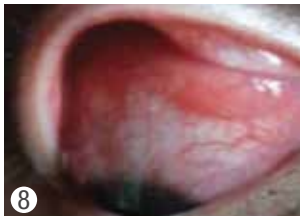
4: Large nodular scleritis
5: Initial MRI shows nodule at superior quadrant with thickening of sclera

Patient was started on topical 1% prednisolone eye drops 4 times a day along with 0.25% betadine drops and reported significant improvement in ocular symptoms and signs which was continued for a week followed by tapering of steroid eye drops. A repeat MRI (Figure 2) orbit done after 5 days showed resolution of inflammation and scleral thickening. The patient's systemic condition remained stable and resolved without worsening or requiring hospitalisation.



6: Post treatment MRI shows complete resolution of the nodule

Repeat COVID test done after the treatment were negative and on regular follow-up the signs and symptoms also reduced.

7: Follow-up reduced conjunctival congestion and chemosis
8: Reduced size of nodule

Patient was finally seen in the clinic after quarantine was over. His steroids were tapered and then stopped.


9: Final visit - No congestion is seen and eye is clear
10: Final visit - Complete resolution of nodule

DISCUSSION

This is the first reported case of nodular scleritis being the presenting feature in a patient with COVID-19 infection. Another unusual feature of the case is that the patient had very mild systemic features with only ocular features which resolved with supportive treatment and did not require hospitalisation.

There have been reports of patients with COVID-19 developing acute follicular conjunctivitis, conjunctival hyperaemia, chemosis, epiphora, and increased ocular secretions.(6/7) These manifestations however have been observed more frequently in patients with severe pneumonia and during the middle phase of illness.(4) Only 1 patient in a series of 38 cases was reported to have conjunctivitis as the initial manifestation of the disease.(4) There has also been a case report of a patient presenting with conjunctival congestion and then rapidly worsening to develop severe acute respiratory illness within few hours.(8) This the is first report of an associated Nodular Scleritis and Conjunctivitis with SARS-CoV-2. Though the patient did develop fever and cough, the patient was only mildly symptomatic requiring home quarantine and not hospitalisation in contrast to earlier reports of ocular manifestations presenting in patients with severe respiratory distress often requiring admission to the intensive care unit.(6)

Scleritis is an inflammatory process involving the outer coating of the globe which is characterised by focal or diffuse hyperaemia, moderate to severe pain, and possible impairment of vision. The autoimmune scleritis constitutes the majority of cases of scleritis. Topical and/or systemic corticosteroids are the management of choice in these cases. Seldom, scleritis may be caused by an infectious aetiology, seen in 5%-10% of cases. There has been an association of scleritis with herpes group of viruses, however there has been no evidence of association with SARS-CoV-2 yet.(9)

This case also highlights an unusual trajectory of the disease wherein the patient had a fluctuating COVID test positivity correlated with the fluctuating levels of

ocular manifestations as well. It is important to try and decipher the possible reasons for the varying COVID test results. Negative test result immediately followed by a positive test result within a short period of time, could be due to a false-negative test result. RT-PCR is commonly used to detect SARS-CoV2 in samples. There are a number of reasons for false negative results. It is well known that results from real-time RT-PCR can be affected by the variation of viral RNA sequences.(9,10) In addition, according to the natural history of the COVID-19 and viral load kinetics in different anatomic sites of the patients, sampling procedures can contribute to the false-negative results. A study has reported sputum as the most accurate sample for laboratory diagnosis of COVID-19, followed by nasal swabs, while throat swabs were not recommended for the diagnosis. The role viral load kinetics of SARS-CoV-2 was documented in two patients in Korea where they have shown a variation, suggesting a different viral load kinetics from that of previously reported other coronavirus infections.(11,12) The virus was detected from upper respiratory tract (URT) and lower respiratory tract (LRT) specimens 2 days after onset of symptoms, however an altered viral load led it to be negative after 5 days but on the 7th day due to a spike in the viral load the patient again turned out to be positive. These findings indicate the different viral load kinetics of SARS-coV-2, suggesting that sampling timing and period of the disease development play an important role in real-time RT-PCR results.

Conjunctival swabs from patients with ocular manifestations have proven positive for SARS-CoV-2 in only 5% patients.(13) This low detection rate could indicate a low prevalence of the virus in conjunctival secretions and tears or viral loads below the detection thresholds of existing PCR diagnosis techniques.(14) This could explain the initial negative result on RT-PCR. One limitation of this case report was that we did not demonstrate the virus from conjunctiva to conclusively link the nodular inflammation to SARS-CoV2.

This case also illustrates the usefulness and relevance of teleophthalmology procedures during the COVID-19 epidemics, which, in addition to preventing the transmission of SARS-CoV-2, could help detect potentially COVID-19 patients.

Ophthalmologists should be aware of these unusual ocular presentations of COVID-19 since they could precede the development of systemic manifestations and help early identification & treatment of these cases.

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ADOLESCENT ONSET HEPATOBLASTOMA: SUCCESSFUL TREATMENT WITH MULTIDISCIPLINARY APPROACH.

A rare case report by **Alexis Multispeciality Hospital,** Nagpur

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ABSTRACT

Hepatoblastoma, a primary neoplasm of liver, is commonly seen among the paediatric population. Adult onset of this disease poses a challenge in diagnosis and hence in timely treatment. Adult onset hepatoblastomas are rare and often shows a poor prognosis owing to delayed presentation, delay or misdiagnosis and delayed treatment. We present a case of adult onset hepatoblastoma which was diagnosed and treated with preoperative neoadjuvant chemotherapy followed by liver resection. High index of suspicion, timey biopsy and multidisciplinary team approach plays a vital role in successful outcomes.

CASE REPORT

A 22-year-old male patient was presented with recurrent abdominal pain and vomiting to the oncology clinic along with pain in upper abdomen.
On examination, he was stable vitally. There was palpable lump in epigastrium which was firm to hard in consistency and was moving with respiration. The lump was in continuity with palpable enlarged liver.
Sonographic evaluation revealed large mass arising from the left lobe of liver with areas of heterogenous echogenicity. Contrast enhanced CT was done which was suggestive of 14.2 cm x 8 cm x 9 cm heterogeneously enhancing lesion in segment III, IV and VIII with peripheral pooling of contrast on arterial phase, no evident portal phase washout with homogenous enhancement on delayed phase. There was another lesion in close proximity in segment II measuring 8.4cm x 6.4 cm x 8.3 cm with similar characteristics. Serum AFP levels were 1645 IU/ml.
In view of atypical findings and very high AFP levels, percutaneous USG guided biopsy was done from the left liver lesion. The biopsy was suggestive of nodular arrangements of nests, acini and trabeculae of hepatocytes with moderate nuclear pleomorphism which as suggestive of hepatocellular carcinoma. However, the immunohistochemistry marker study diagnosed it as hepatoblastoma and categorised it to be a fetal type HB.

On preoperative staging of the disease it was categorised into PRETEXT III since it was involving more than 3 sections and was closely related to right hepatic vein but not involving it. After thorough evaluation of routine blood investigations including complete blood count, liver function tests, renal function tests and coagulation profile, he was started on neoadjuvant

chemotherapy (NACT) with cisplatin and Adriamycin in consultation with medical oncologist.

He was reassessed with serum AFP and CECT abdomen after 3 cycles of NACT. AFP was reduced to 125 IU/ml and there was significant reduction in tumor size. He further received 2 more cycles of cisplatin and Adriamycin. Post NACT AFP reduced to 20 IU/ml while the follow up CT showed a significant reduction in size of lesion to 8.2cm x 5.2 cm x 5 cm in segment III, IV and VIII and 6.4 cm x 4.8 cm x 6 cm in Segment II. There was significant increase in the volume of right posterior segment which was approximately 50% of total liver volume and tumour had retraced significantly away from right hepatic vein. Preoperative liver function tests were normal.

He underwent extended left hepatectomy where in Segment II, III, IV A, IV B, V and segment VIII was removed. Tumour was found to be free from the right hepatic vein. The stumps of middle and left hepatic vein was adequate enough to ligate after taking a margin from the tumour. Postoperatively patient was monitored in ICU for one day. He improved gradually and his liver function test normalized on POD 4. Patient was discharged on POD 5. His final histopathology report suggested viable nests of foetal type of hepatoblastoma in the resected specimen with negative surgical margins. He also received 2 cycles of chemotherapy with same regime. On the 3rd month follow up, the AFP levels had reduced to 8 IU/ml.

DISCUSSION

Hepatoblastoma (HB) is a rare type of liver tumour, comprising of two third of all malignant neoplasm of liver in children less than 2 years of age. Although most of them are sporadic, some are associated with genetic syndromes like Beckwith-Wiedemann syndrome and familial adenomatous polyposis.(1)(2) The prevalence of this disease decreases after that age and is even rarer after age of 5 years.(3) This was a peculiar presentation of hepatoblastoma which presented in an adult patient. In literature about 40 cases have been described.(4) Although the AFP levels were very high, only IHC markers could help to differentiate it as foetal type of hepatoblastoma and differentiate it from hepatocellular carcinoma.

In as small review by Zheng et all, they found female preponderance in adult onset hepatoblastomas. They are commonly found as unifocal isolated lesion involving right or left lobe. However, as we noticed in our case multifocality of onset is known although rare. Multifocality was found in only one case in the review.(4)

The cross-sectional imaging forms a vital tool for diagnosis along with very high AFP levels. Any atypical presentation can be a ground for preoperative biopsy. In our patient the CT picture was atypical for the diagnosis of hepatocellular carcinoma as there was delayed enhancement on delayed phase and no portal phase washout which led us to investigate more to look for appropriate diagnosis. Hepatocellular carcinoma is one of the commonest differential for adult onset HB which was our differential diagnosis before complete histopathological evaluation.(5)

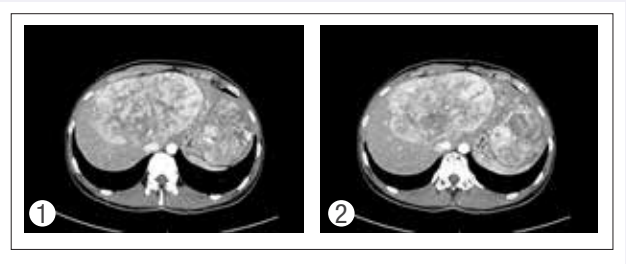
Surgery forms mainstay of curative therapy for hepatoblastoma. But NACT is essential to reduce the size of the tumour, prevent metastasis, prevent tumour related complications like bleed and gives time for the liver to have an adequate amount of remnant liver volume. In our patient the remnant liver volume of right posterior sector increased approximately from 20% to 50% in the span of 3 months during chemotherapy which definitely helps in post-operative recovery following major liver resection.

The prognosis of HB treated with NACT followed by surgery depends on preoperative staging, tumour biology, pre-operative AFP levels, R 0 resection of tumour and age of onset of disease. Our patient comes in moderately high-risk category as it was PRETEXT III stage preoperatively and adult age of onset. However, at the 3rd month follow-up, patient seemed to be doing well. His AFP levels had almost normalised (8IU/ml) from preoperative value of 1645 IU/ml.

Reports from literature where HB was diagnosed on post-operative histopathology, have shown poorer prognosis. Overall neoadjuvant chemotherapy improves operability and recurrence free survival.

CONCLUSION

High index of suspicion, a preoperative diagnosis by percutaneous biopsy of tumour, timely NACT and a favourable response to chemotherapy and a R0 resection of tumour can help improve the long-term outcomes.



1 & 2: Pre NACT-tumour involving segment II, III, IV and VIII with involved middle hepatic vein and closely abutting right hepatic vein.

3: Post NACT favourable response with right hepatic vein away from tumour.

4: Resected specimen

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BLASTOID VARIANT OF MANTLE CELL LYMPHOMA WITH LEUKEMIC PRESENTATION

A rare case report by **Alexis Multispeciality Hospital,** Nagpur

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Senior Consultant Medical Oncologist

Dr. Mahesh Deshmukh
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Dr. Shahab Sharif
Associate Consultant Radiologist

ABSTRACT

Mantle Cell Lymphoma (MCL) is a type of Non-Hodgkin's lymphoma and has a wide spectrum of histopathological subtypes of which the blastoid variant constitutes 10-15% of all cases. It is difficult to diagnose blastoid variant of MCL on the basis of morphology alone as it mimics lymphoblastic lymphoma and centroblastic large cell lymphoma, hence additional analysis like immunophenotyping and molecular studies aid in its diagnosis.

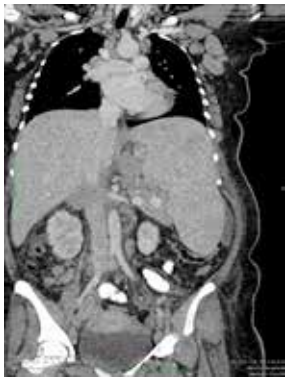
We present an extremely rare case of 45-year-old lady who presented to the Oncology clinic with chief complaints of fever, fatigability and neck swelling. Complete blood count showed pancytopenia with large cells showing high N:C ratio and dispersed chromatin. Bone marrow examination revealed 37% atypical lymphoid cells which on flow cytometric immunophenotyping expressed clonal B cell phenotype was diagnosed as blastoid variant of MCL. Cyclin D1 positivity was seen on immunohistochemistry, confirming the diagnosis. Patient was administered R-CHOP and underwent autologous bone marrow transplant. Patient is on a regular follow up for 18 months now post autologous transplant. This is the only case documented in the literature with a long survival.

Keywords: Flow cytometry, Hepatosplenomegaly, Blastoid, Non-Hodgkin's lymphoma

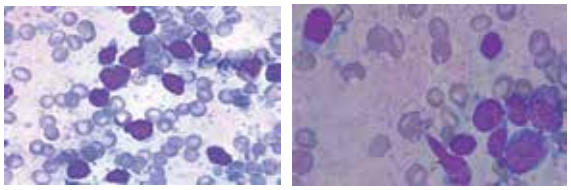
CASE REPORT

A 45-year-old female patient presented to oncology clinic with chief complaints of fever, easy fatigability and neck swelling of recent onset. Physical examination revealed enlarged bilateral cervical lymph nodes, splenomegaly (14 cm below the left costal margin) and hepatomegaly (8 cm below the right costal margin). Complete blood count showed pancytopenia and peripheral smear demonstrated 23% abnormal cells which resembled lymphoblasts. Computerized tomography (Figure 1) showed generalised lymphadenopathy. Bone marrow aspirate smears demonstrated 37% abnormal lymphoid cells which were medium to large sized with high N:C ratio with dispersed nuclear chromatin, scant cytoplasm (Figure 2) and few showing irregular nuclear margins and inconspicuous nucleoli. Trephine biopsy showed interstitial and focal nodular infiltration by abnormal lymphoid cells. Flow cytometric analysis was performed on bone marrow aspirate and the cells were bright CD45 positive and negative for CD34, which confirmed that the abnormal cells were mature lymphoid cells and not blasts. The abnormal lymphoid cells showed clonal B-cell phenotype (positive for CD5, CD20, CD19, CD22, CD10, and CD79a with surface kappa Figure 3 light chain restriction). The cells were negative for other T cell markers, myeloid markers, monocytic markers, and CD15. Immunohistochemistry was done on bone marrow biopsy which showed Cyclin D1 was positive in >10% abnormal cells. A diagnosis of Blastoid variant of Mantle Cell Lymphoma (MCL) was rendered. The patient completed six cycles of chemotherapy with Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP) regimen after which her disease was in complete metabolic and morphological remission. She underwent Autologous Hematopoietic Stem Cell Transplant (HSCT) in first complete remission

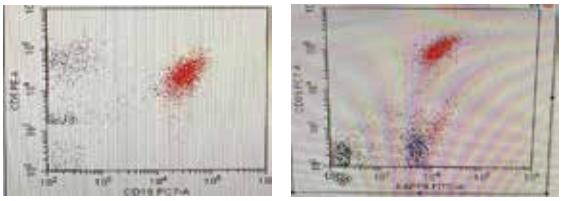
with BEAM as a conditioning regimen. She is doing well and is in regular follow up post 18 months transplant.



1: CT image shows hepato-splenomegaly with abdominal lymphadenopathy



2: Bone marrow aspirate shows lymphoid cells with dispersed chromatin and inconspicuous nucleoli. (Leishman's stain, 400x)



3: Flow cytometric analysis shows abnormal lymphoid population (in red) expressing CD5, CD19 and kappa light chain restriction.

DISCUSSION

MCL constitutes 3-10% of all Non-Hodgkins Lymphomas. It usually occurs in middle aged to older adults with a median age of 60 years and has a male predominance. MCL is a subtype of B-cell Non-Hodgkins lymphoma arising from naive pre-germinal center B cell within the mantle zone that surrounds normal germinal center. These cells show over expression of cyclin D1 due to the chromosomal translocation t (11;14) (q13;q32) (1). The presentation is usually at an advanced stage III or IV with lymphadenopathy, hepatosplenomegaly and bone marrow involvement. Our case was a young adult female with stage IV disease. A 30-50% of cases present with systemic symptoms like weight loss, fever and night sweat.(2) The primary site of involvement is lymph node which is often generalised. The other common sites involved are spleen and bone marrow with or without peripheral blood involvement. The frequently involved extra nodal site is gastrointestinal tract and Waldeyer ring.(3) The morphology of classical MCL is monomorphic proliferation of lymphoid cells with diffuse, vaguely nodular, mantle zone or in rare instances has a follicular growth pattern. MCL has a broad spectrum of several histopathological variants of which blastoid variant constitutes 10-15% of all cases.(4) The blastoid variant differs from the classical MCL as the cells resemble lymphoblast with more dispersed chromatin and have a very high mitotic activity (at least 20-30/10HPF). Our case was morphologically blastoid variant and showed positivity for CD5 and CD10. Another unique feature was kappa restriction. But Cyclin D1 positivity confirmed the diagnosis. The differential diagnosis of blastoid variant includes centroblastic large cell lymphoma and lymphoblastic lymphoma. But the characteristic immunophenotype in the blastoid variant reveals a mantle cell pattern.(5) The adverse prognostic factors include very high leukocyte count, high mitotic activity, increased ki 67 index, pleomorphic or blastoid morphology and overt peripheral blood involvement.(3) MCL has a poor prognosis with a median overall survival of 2 to 3 years with the administration of standard therapy. The blastoid variant is more aggressive and is refractory to chemotherapy. It has a very poor prognosis and poorer overall survival indicating that it is one of the worst forms of Non-Hodgkin's lymphoma.(2) In our case as the patient underwent autologous HSCT, we expect a good long-term overall survival. Our case emphasized the heterogeneity in blastoid MCL cases wherein in spite of being stage IV disease, she responded well.

CONCLUSION

The blastoid variant of MCL is a rare type of Non-Hodgkin lymphoma which carries an overall poor prognosis. Immuophenotyping and flow cytometry is a must for correct diagnosis because recognising blastoid variant carries prognostic significance.

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HARTNUP DISEASE – A RARE DERMOMETABOLIC DISORDER

A case report by **Alexis Multispeciality Hospital,** Nagpur

Dr. Bhaskar Gupta
Consultant Paediatrician and Neonatologist

ABSTRACT

Dermometabolic disorders due to inborn errors of metabolism in children can manifest in a variety of ways. Disorder due to Tryptophan metabolism being rare can cause symptoms which can be confused with a variety of skin diseases. One such disorder of Tryptophan metabolism which is autosomal recessively inherited known as Hartnup disease can be very distressing to the child and family and can cause severe morbidity and death due to complications. The characteristic findings are pellagra like dermatitis following exposure to sunlight, intermittent cerebellar ataxia, psychosis and constant aminoaciduria. A proper history with clinical findings and investigations can diagnose this disorder and with proper medications the flare ups and complications can be minimised. A case of Hartnup disease in which the child presented repeatedly with severe illness and his elder sibling had died due to similar presentation was thoroughly examined and investigated and thereby diagnosed and got relieved with due medications at Alexis Hospital, Nagpur; thus, highlighting the need to be alert with regards to various inborn errors of metabolism in children.

INTRODUCTION

Hartnup disease is an aminoaciduria that is usually clinically silent but can manifest with severe and intermittent episodes of pellagra like rash, cerebellar ataxia and psychosis. The reported incidence is 1 in 30,000 persons and is autosomal recessively inherited due to mutations in SLC6A19 gene coding an enzyme that transports neutral amino acids across the apical membrane of epithelial cells in the gut and kidneys. Due to this large amount of neutral amino acids including tryptophan are present in urine establishing the diagnosis. A 3-year-old child was presented with severe pellagra like skin lesions since the age of 9 months and was repeatedly hospitalised due to its complications and had failure to thrive. He was examined and investigated. With a simple medication of Nicotinamide tablets daily his symptoms were alleviated and he started with good growth and development. This case highlights the importance of always keeping in mind the rare inborn errors of metabolism in children which if diagnosed properly can reduce the mortality and morbidity.

CASE REPORT

A 3-year-old boy presented with multiple severe excoriating hyper-pigmented lesions on flexural aspect all over the body, perineum and neck, hands and feet with mild itching. The first episode was at 9 months of age with irritability, poor-feeding, fever and lesions and was admitted to the hospital in Chhattisgarh. He was investigated and treated with emollients, antibiotics and nutritional supplements and was provisionally diagnosed as staphylococcal scalded skin syndrome. The lesions gradually reduced but continued off and on till the time at age of 16 months he again had severe flare ups with poor feeding, stomatitis, cough and irritability and was taking oral antibiotics. He was admitted again at another hospital and treated with provisional diagnosis of Steven Johnson's syndrome due to the reaction of antibiotic that had been consumed. The skin lesions gradually reduced and his third admission at age of 2 years was with diarrhoea, fever and similar excoriating skin lesions. This time he was being treated with provisional diagnosis of Acrodermatitis enteropathica and was given steroids and zinc although the Serum and Urine zinc levels sent were normal.

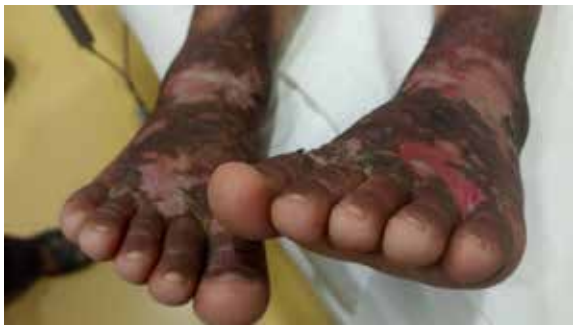
He was presented to Alexis Hospital in February 2018 with extensive excoriation, abdominal pain and anorexia. He was admitted and found to have weight of 11 kgs which is below 10th centile for expected age, erythematous plaques over thighs, hands and legs, perineum, ankles with bleeding at some spots and excoriation with hyperpigmentation. He was kept on IV fluids, emollients, zinc, and anti-allergics with antibiotics. His history revealed that he was born as full-term gestation with birth weight of 2.8 kgs with history of consanguinity and slightly delayed developmental milestones with emotional instability and irritability and mood swings. His examination revealed that he had sunlight exposure exacerbation and

hyperpigmentation with lichenification and stomatitis. Also, he was found to have mild gait instability and nystagmus. His investigations revealed high WBC count with neutrophils of 62% normal Liver function tests and ASO titre of 27, normal Calcium, phosphorous and alkaline phosphatase and low vitamin D levels. His Serum Zinc was 101 which were within normal limit and his urine for porphobilinogen was negative.

A detailed history revealed that his elder sibling presented with similar recurrent skin lesions, with poor nutrition, unstable gait, uncontrollable and irritable behaviour; died at age of 3 years.

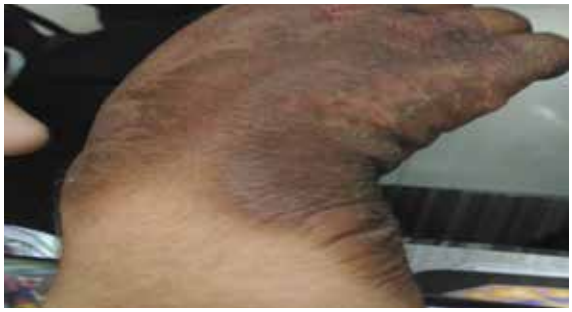
With due care his lesions improved and he was advised follow up after 4 weeks. At follow up again similar skin lesions were noted and his urine for Amino acid was sent which showed aminoaciduria with excess excretion of neutral amino acids alanine, serine threonine, tryptophan, valine leucine isoleucine and excretion of basic and acidic amino acids like proline, hydroxyprolene, and arginine were normal.

The clinical presentation of recurrent extensive photosensitive dermatitis with extensive lichenifation and emotional instability with nystagmus with aminoaciduria suggested a diagnosis of Hartnups disease. He was put on Tablet Nicotinic Acid 250 mg (Nicoglow) per day with high protein diet, Emollients and nutritional supplements with advice to avoid sun exposure and was followed up after 6 weeks. Subsequent follow ups at 6 weeks, 3 & 6 months and 1 year later revealed no skin lesions, good weight gain and appetite with reduced irritability.



Presentation with extensive lesions

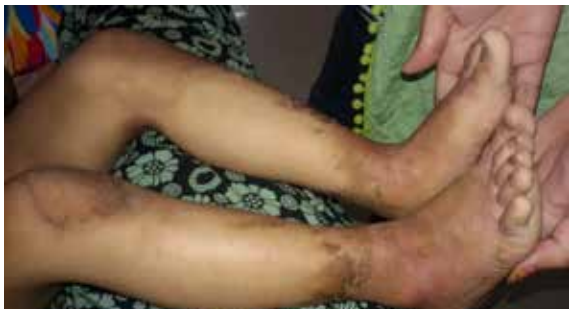
Healing lesions with only hyperpigmentation one month after starting treatment



Healing lesions with only hyperpigmentation one month after starting treatment

Healing lesions after treatment for 3 months

Presentation with extensive lesions



Healing lesions after treatment for 3 months

DISCUSSION

Acute dermatitis and blistering with secondary crusting and scarring with lichenification and hyperpigmentation following sun exposure are seen in pellagra which is due to Vitamin B3 deficiency and is uncommon now-a-days with triad of dermatitis, diarrhoea and dementia. This child presented with neurological features along with glossitis, stomatitis, nystagmus, emotional disturbance and aminoaciduria with familial occurrence thus suggestive of the diagnosis of Hartnup disease. The lesions are exacerbated during the period of malnutrition and intercurrent infections.

Hartnup disease is autosomal recessive in inheritance with the genetic defect in 5p15.33 locus encoding for SLC6A19 this gene was identified in the year 2004. The estimated incidence is 1 in 30,000 persons. The metabolic aberration in hartnups disease results from an error in the transport of monoamino monocarboxylic neutral amino acids that affects renal tubular reabsorption and intestinal absorption. There is deficient transport of neutral amino acids including tryptophan and thus large amounts of amino acids are excreted in urine. Large amounts of tryptophan remain in intestinal lumen where it is converted to Indolic compounds by bacteria and then absorbed. These compounds are toxic to Central nervous system thereby causing neurological manifestations. Large amounts of Indican excretion is noted which can be detected by urine chromatography. Lack of tryptophan absorption leads to niacin deficiency which results in pellagra like symptoms and photosensitivity.

Thus, the characteristic features of Pellagra like dermatitis following sunlight exposure with exacerbations during infections and poor nutrition along with intermittent cerebellar ataxia, neurological and emotional features with glossitis and stomatitis and constant aminoaciduria are characteristic features of Hartnup disease. Affected children are normal at birth but may have delayed milestones.

The differential diagnosis is acrodermatitis enteropathica which is an autosomal recessive nutritional dermatosis due to defective absorption of zinc from intestine presents with reddish hairs, alopecia blepharitis, corneal dystrophy poor wound health and zinc levels are very low.

Staphylococcal scalded skin syndrome known as Ritter's disease due to phage group 2 Staphyloccal strains 71 & 55 due to exfoliative toxins A & B presents in kids as bullous impetigo to generalised peeling and Nikolsky sing positive.

Biotindase deficiency is due to defective enzyme for degradation of valine isoleucine and lucine resulting in atopic dermatitis, alopecia, myoclonic seizures and hearing loss and diagnosed by enzyme estimation in serum.

CONCLUSION

Hartnup disease is an inborn error of tryptophan excretion which is named after the Hartnup family in whom it was first noted and is the second most inherited aminoaciduria after phenylketonuria. The intermittent nature of disease in the form of pellagra like dermatitis to face, neck, hands and legs with neurological manifestations and psychological disturbances are characteristic. Early suspicion of this disease is vital in reducing the mortality and morbidity and thereby nutritional and neurological outcome. A simple tablet of Nicotinamide daily with proper diet and avoiding exposure to sunlight is essential part of treatment with excellent results.

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KEYHOLE SURGICAL EVACUATION OF HEMATOMA IN A PATIENT WITH SPONTANEOUS INTRACEREBRAL HEMATOMA

A rare case report by **Alexis Multispeciality Hospital,** Nagpur

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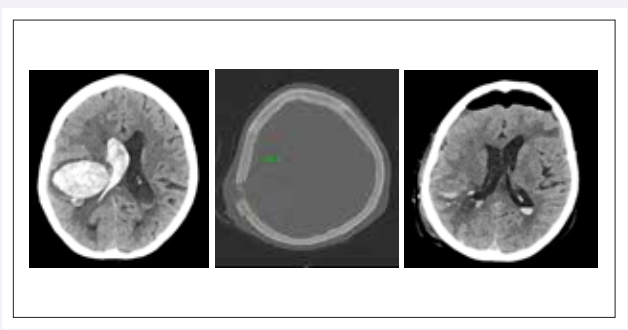
Dr. Abhishek Wankar
Consultant Neuro Physician

Dr. Manoj Pethe
Consultant Pulmonologist

ABSTRACT

Although the surgical management of spontaneous intracerebral hematoma (SICH) is a controversial issue, it can be lifesaving in a deteriorating patient. Surgical techniques have varied from the open large craniotomy, burr hole and aspiration to the minimally invasive techniques like stereotactic aspiration of the SICH, endoscopic evacuation and stereotactic catheter drainage. The authors report a case of SICH treated with keyhole craniectomy technique. A small craniectomy of 2.5 cm diameter was made using a vertical incision over a relatively silent area of the cortex closest to the clot. Using a small cortical incision, the hematoma was evacuated and decompression was achieved. Haemostasis was achieved using standard micro neurosurgical techniques. Good outcome was achieved with minimal blood loss during the procedure. The keyhole craniectomy technique is minimally invasive, safe and can achieve good clot evacuation with excellent haemostasis. It can be combined with microscope or endoscopic assistance to achieve the desired result.



INTRODUCTION

Spontaneous intracerebral hematoma (SICH) is one of the most devastating forms of cerebrovascular disease accounting for about 15% of all strokes. It is associated with high morbidity and mortality.(1) The role of surgery in the management of these cases is controversial. It is possible that some cases will benefit from surgical evacuation. Current practice favours surgical intervention in following situations: lobar haemorrhage, clot volume between 20 to 80 ml, worsening neurological status, relatively young patients, and haemorrhage causing midline shift or raised intracranial pressure (ICP).(2) Surgical indications in cerebellar hematomas however are more accepted. Hematomas above 3 cm diameter and those causing hydrocephalus, generally require surgical evacuation.(2,3,4)

Various surgical strategies have been adopted over the years for evacuation of the intracerebral hematomas ranging from the large open craniectomies and decompression to the more minimally invasive therapies like stereotactic evacuation of hematomas, endoscopic evacuation, stereotactic endoscopic evacuation, stereotactic fibrinolytic therapy etc.(5,9) Tsementzis has advocated a method of a small trephine craniotomy 3 cm in diameter and evacuation of hematoma through this craniotomy.(5) The authors present a case treated with a keyhole craniectomy in the evacuation of SICH.

CASE REPORT

A 78 years old woman was brought in an emergency with history of headache, vomiting, altered sensorium and left sided paralysis. Patient was a known case of hypertension, but on irregular treatment. CT scan brain was performed which showed massive right basal ganglia bleed with intraventricular extension with mass effect and midline shift. An emergency surgery was performed by doing a 2.5 cm keyhole craniectomy and hematoma evacuated under microscope. Post-operative CT brain showed minimal residual hematoma with decrease in intraventricular bleed without any mass effect or midline shift. Post-operatively, patient was on ventilatory support which was weaned off gradually. Patient improved gradually with improvement in paralysis and discharged in a stable condition.

DISCUSSION

SICH forms 15% of all strokes and carries a high morbidity and mortality.(4) Typically, the intracerebral hematomas secondary to hypertension are found in the basal ganglia, putamen and globus pallidus; thalamus; cerebral lobes; cerebellum and brain stem.(1,5) The role of surgical treatment in the management of these hematomas is controversial. Clear-cut indications and guidelines for surgical treatment are not available. However, it is considered by most, that surgery may be indicated in patients where the hematoma is large in the basal ganglia; lobar; where there is secondary neurological worsening; in young patients; in those with hydrocephalus and in those whom a secondary cause is suspected.(1,4,5) There is little to be gained by direct surgery in patients with thalamic and pontine haemorrhage.(1) However, the indications for surgery are more frequent for cerebellar hematomas as the risk of brainstem compression and hydrocephalus from ventricular obstruction are important.(4)

The authors advocate a keyhole craniectomy which is a minimally invasive technique by which significant evacuation of the hematoma (near total and subtotal evacuation) can be achieved with excellent haemostasis. The formal large craniotomy or decompressive large craniectomy and dural enlargement subsequent to hematoma evacuation, have proved to be very useful in a group of severely compromised patients with SICH. The evacuation of hematoma and haemostasis has been found to be excellent with this modality. However, associated morbidity of the craniotomy, prolonged operative time and blood loss have been noted as some of the disadvantages of this approach. The 'edge effect' resulting in compression of the brain and the cortical veins along the edges of the craniectomy has also been cited as one of the disadvantages. In the minimally invasive procedures, the morbidity of extensive craniotomy can be obviated, but the evacuation of hematoma and subsequent perfect haemostasis may be technically difficult. The keyhole craniectomy is a less invasive method requiring less operating time and blood loss. Perfect haemostasis can be achieved using the standard micro neurosurgical techniques. The 'edge effect' of a large craniectomy is obviated. Endoscopic usage is also possible through the same approach. The keyhole craniectomy can also be extended into a formal craniotomy if a secondary lesion such as an aneurysm or an arterio-venous malformation or a tumour is encountered. This surgical procedure can be performed at all neurosurgical centres with basic neurosurgical infrastructures, without any expensive instruments such as the stereotactic apparatus or the endoscope.

CONCLUSION

Keyhole craniectomy for the surgical evacuation of SICH is a less invasive and effective surgical modality. Good evacuation of the clot can be achieved with perfect haemostasis. Good results with this procedure can be obtained in young patients with lobar or large basal ganglionic hematomas with worsening neurological status. Surgery is not a preferred modality of treatment in patients with deep seated (thalamic and brainstem) hematomas. Poor results may be expected in patients with advanced age, hematoma volume of more than 60 ml, and GCS less than 6.

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AN UNUSUAL CASE OF A COMPLETE LEFT LUNG COLLAPSE TREATED SUCCESSFULLY AT ALEXIS MULTISPECIALITY HOSPITAL

A rare case report by **Alexis Multispeciality Hospital,** Nagpur

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INTRODUCTION

Foreign body aspiration is not an uncommon problem particularly in children, elderly and those with predisposing factors like neurological impairment. In adults the diagnosis is usually delayed as it is confused with other lung diseases.(1) Foreign body aspiration if undiagnosed can often lead to distal airway complications such as partial or whole lung collapse, post obstructive pneumonia etc. Once diagnosed, the management involves use of rigid or flexible bronchoscopy for prompt removal of foreign body. Here we present a case of a 75 years old male presented to us after almost one-month history of aspiration of betel nut pieces & complete left lung collapse. Removal of multiple pieces of betel nut with the use of rigid bronchoscopy & flexible bronchoscopy under general anaesthesia and thereafter successful management of this patient for post obstructive pneumonia & sepsis in our intensive care unit (ICU) are the highlights of this case.

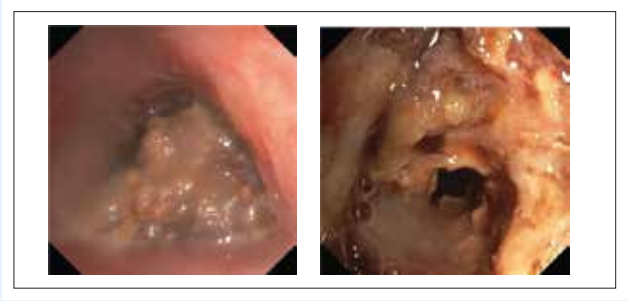
CASE REPORT

A 75-year-old male with no previous medical problems was admitted the in first week of June 2017 to Alexis Multispeciality Hospital with complaints of cough with expectoration and breathlessness on exertion of 1-month duration and history of intermittent fever of 1-week duration. On admission to Alexis Hospital he was afebrile, blood pressure (130/74mmHg), pulse rate (86/ min) and respiratory rate (20/min) were normal. Examination of respiratory system revealed absent breath sounds & dull note on percussion over left hemithorax. Other systemic examination was unremarkable. Posteroanterior radiograph of chest x-ray showed complete collapse of the left lung and there was cut off of left main bronchus.

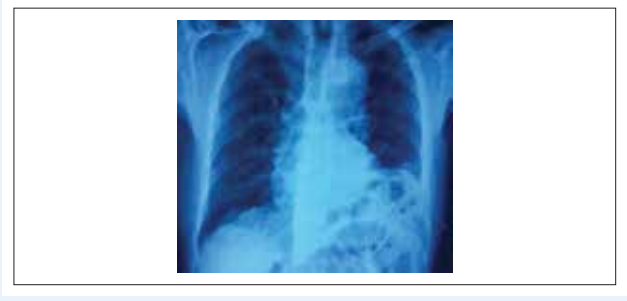
CT-scan examination of the thorax revealed complete collapse of left lung & endobronchial opacity in the left main bronchus suggestive of a foreign body which was almost completely occluding the lumen of the left main bronchus. Patient's history was reviewed again and he recollected history of foreign body aspiration (betel nut) one month back while chewing betel nuts after dinner. He had intractable cough & choking sensation after this episode of aspiration. Bronchoscopy was done, which showed aggregated beetle nut particles completely occluding left main bronchus. In view of this, rigid

bronchoscopy under general anaesthesia was performed next day and with rigid as well with a flexible bronchoscope multiple pieces of beetel nuts were removed from left main bronchus and segmental bronchi on left side.

This procedure lasted for 3 hours & during this procedure it was also noted that from 2 segments of left lower lobe bronchi, pus was oozing out. At the end of this procedure, it was ensured with flexible bronchoscopy that the all bronchi on in left lung were patent and there were no visible foreign bodies.



Following this procedure patient had developed hypotension secondary to sepsis due to post obstructive pneumonia in left lower lobe. He was treated successfully in our intensive care unit. Pus which was aspirated from left lower lobe bronchi during the procedure grew klebsiella Pneumoniae and patient was treated as per antibiotic sensitivity report. He recovered well with antibiotics & supportive care and was discharged after one week. His post procedure chest X-ray showed complete expansion of left lung.



DISCUSSION

Adults constitute approximately 20% cases of tracheobronchial aspiration. Among adults, the median age group reported is 60 years. This is attributed to higher incidence of cerebrovascular and neurodegenerative disorders in elderly that compromise swallowing and airway protective reflexes. Most commonly aspirated materials include vegetable matter, dental appliances/prosthesis, medical appliances, bone fragments etc.(2) A history suggestive of aspiration may not be forthcoming in most of the adults and some may recollect it retrospectively. Betel nut is a commonly incriminated agent in foreign body aspirations from the South East Asian region. Once diagnosed, prompt removal of foreign bodies from bronchial tree is essential to avoid irreversible complications.

This patient already had developed pneumonia due to obstruction of major bronchi in left lung and further delay in removing betel nut particles would have led to haemoptysis, lung abscess, scarring & destruction of lung and permanent collapse of left lung. Betel nut as it absorbs moisture while in bronchi, it swells up and causes more bronchial obstruction. It is known to cause excessive damage and scarring surrounding it and also can cause bronchial erosions.(3) Removal of multiple betel nut particles by the use of both rigid & flexible bronchoscope under GA & post procedure successful management sepsis due to post obstructive pneumonia in ICU make this case report unique.

A high index of suspicion is required in young adults and elderly for early diagnosis and management of endobronchial foreign bodies to avoid long term complications.

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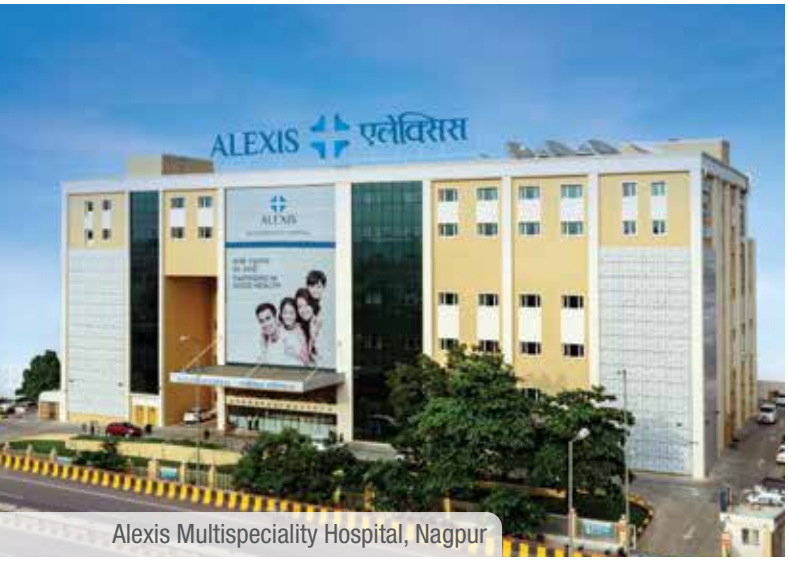

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