Case Report

UNCOMMON CAUSE OF ABDOMINAL LYMPHADENOPATHY – CASTLEMAN DISEASE

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ABSTRACT

No doubt it is rightly said that abdomen is a temple of surprises. The case presented here is one of the uncommon entity which a surgeon encounters . Abdominal mass always presents a diagnostic dilemma to the treating surgeon because of various structures from which it can arise. Mesentric mass in particular is one such aspect which always poses a difficullty to surgeon because of the extension of the mesentry right across the abdomen from right iliac fossa to left hypochondrium. The mass of the mesentry ranges from cystic mass to solid mass. The case that follows is a solid mass which is an uncommon variety of lymph node neoplasm- Castleman disease.

Keywords : castleman disease, lymph node neoplasm

CASE REPORT

A 45 year old female patient presented to the emergency department with lower abdominal pain for 20 days duration. She had no other symptoms corresponding to gastrointestinal system involvement. She had no comorbid illness and have undergone sterilization at 27years of age. She attained menopause 3 years ago and had no significant post menopausal symptoms. On examination she was moderatly built and had no pallor and jaundice. She was in no distress. On examination, abdomen was soft, no mass palpable, normal bowel sounds and no organomegaly. Per rectal examination was normal. Her cardiac examination and respiratory examination was normal and her vitals were within normal limits. Patient was admitted for observation.

On blood investigation, patient was found to be anemic Hb% - 8.6g%. Ultra sonogram of the abdomen revealed a well defined solid homogenous mass 5.9X4.8cm noted in right paraumbilical region moving with respiration. CECT (contrast enhanced computed tomography)of abdomen showed a 6.7 X 5 cm hypervascular lesion in the mesentry in the right lumbar region closely abutting the inferior wall of proximal transverse colon – with the possibilities of lymph node or carcinoid and multiple homogenously enhancing discrete lymph node in the mesentry in peripancreatic, perigastric and near transverse colon with the possibility of metastases [Fig. 1]. OGD (oesophago gastro duodeno) scopy was done which showed antral gastritis. CT (computed tomogram)guided FNAC (fine needle aspiration cytology) of the mass was inconclusive and only hemorrhagic fluid was present.

An exploratory laparotomy was planned. Under general anesthesia via midline incision, abdomen was explored and a firm mass of size 5X6 cm was present in the transverse mesocolon abutting the inferior surface of the proximal part of transverse

colon [Fig.2]. Rest of the abdomen was normal. Mass was excised and sent for HPE (histopathological examination). Abdomen was closed and post operative period was uneventful and patient was discharged on 10th post operative day. The biopsy report showed a solid mass with homogenous surface [Fig.3]and microscopic examination revealed a hyperplasia of lymphoid follicles with lollypop appearance and onion skin appearance [Fig. 4] increased hyaline collagen and a diagnosis of Castleman disease of hyaline variety made.

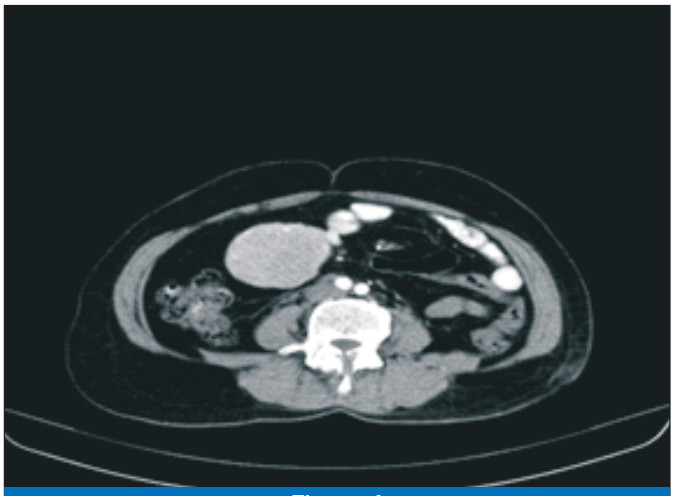

Figure 1

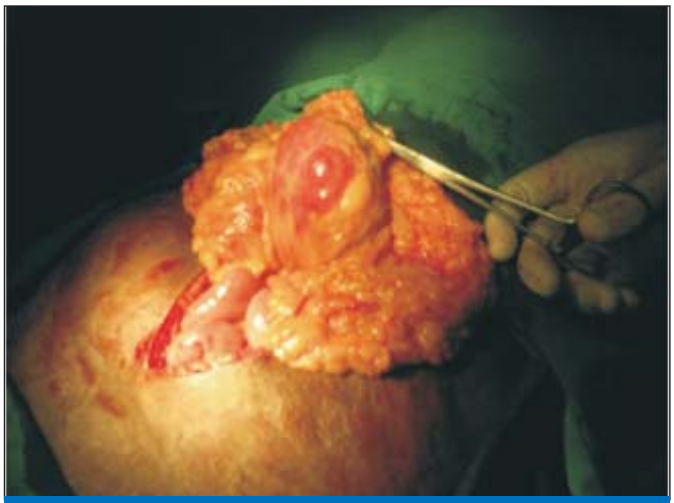

Figure 2

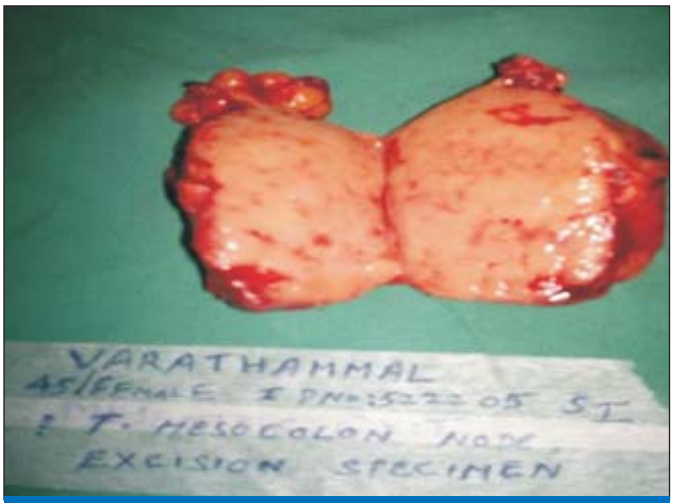

Figure 3

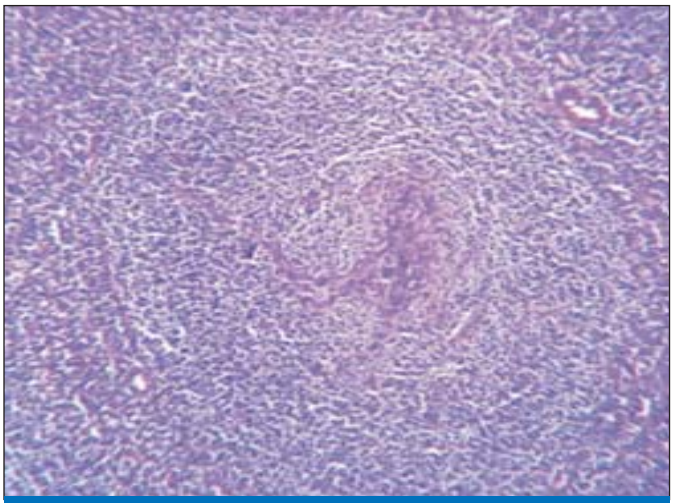

Figure 4

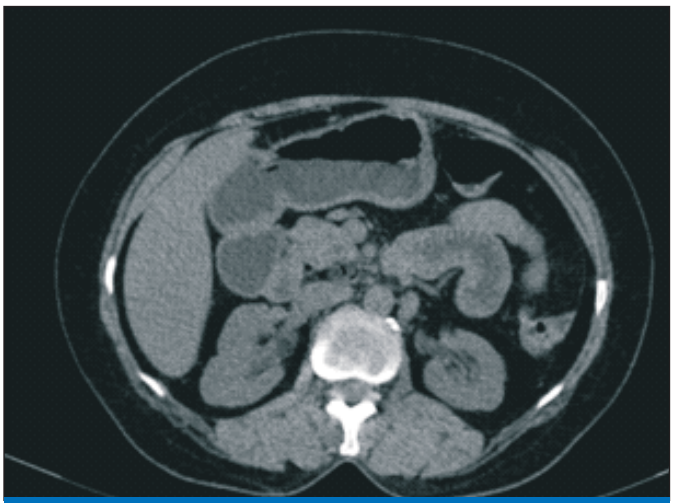

Figure 5

DISCUSSION

Castleman disease is a lymphoproliferative disorder first coined by Benjamin Castleman in 1956. It was formerly called as angiofollicular hyperplasia. It can be classified as unicentric vs mulitcentric, hyaline vs plasmacytic vs mixed cellularity and HIV negative vs positive[1]. The patient above discussed was unicentric, HIV negative and hyaline type. Unicentric tumors are usually located in mediastium or mesentry and symptoms are only due to pressure effects due to the bulkiness of the tumor. Multicentric variety on the other hand is widespread and presents with constitutional symptoms of fever , weight loss, fatigue and night sweats and peripheral edema. Untreated multicentric variety progress to non hodgkins lymphoma. Patients with multicentric disease usually have plasmacytic and mixed histology . If it is complicated by HIV the disease runs a very aggressive course. Human herpes virus 8(HHV 8) [2] is considered to be one of the causative agent of multicentric castleman disease because of its presence in lymph node and mononuclear cells in HIV positive patients. Detection of HHV 8 in HIV positive DNA antedates the development of multicentric disease by 5% to 35%. Cases of extrathoracic Castlemans disease with paraneoplastic phempigus and myasthenia gravis have been documented[3]. Curative therapy for unicentric disease is mainly surgical excision [4], [Fig.5- Post op status] and usually there is no progression to other tumors like lymphoma. If the lesion is too bulky pre operative steroids and rituxan can be tried. Radiotherapy also forms an important modiality of treatment in patients with unicnentric disease where surgery is not possible due to fixity to underlying structures and the response rate is near to 70% [5,6]. In case of multicentric disease it is mainly systemic therapy with intravenous immunoglobulins against Interlukin 6, Siltuximab [7,8]and other like rituximab for TAFRO (thrombocytopenia, anasarca, fever, reticulin fibrosis and organomegaly) [9]. In intractable cases stem cell transplantation can be tried.

CONCLUSION

The Patient is under regular followup for past 4yrs and is disease free. Latest CT abdomen showed normal study except few subcentimetric nodes in mesentry and in retroperitoneum. Though the presentation was clinically insignificant further investigations turned out to show one of the rare causes of lymphoproliferative disorder. Diagnosis of this disease is based on histopathological examination. The prognosis of unicentric castleman disease is very good, close to 100% disease free survival rate by 5years. On the other hand multicentric disease runs an aggressive course and at a risk of developing multiple autoimmune diseases like autoimmune hemolytic anemia, pemphigus ,amyloidosis and multiple myeloma. Thus this patient requires a life long follow up to find out possiblilty of unicentric transforming into a multicentric variety in a HIV negative patient.

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