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UNICENTRIC CASTLEMAN DISEASE PRESENTING AS A LARGE RETROPERITONEAL MASS-A CASE REPORT

KEYWORDS: Castleman disease, unicentric/multicentric, hyaline vascular variant, onion skin pattern, lollipop lesions, HHV-8, IL-6

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ABSTRACT *Castleman disease (CD) described by castleman as a large,benign,asymptomatic mass involving mediastinal lymph nodes.The causes of CD are mostly unknown.As patients with CD can have markedly different presentation and clinical courses,with some lesions requiring innovative approaches to therapy, the major unifying feature is the histologic appearance. It may be unicentric or multicentric, hyaline-vascular variant or plasma cell variant. Unicentric variant mostly present as a isolated lymph node mass ,often mediastinal or cervical. We present a case of unicentriccastleman disease (hyaline vascular variant) presenting as a large retroperitoneal mass.*

SUMMARY:
Castleman disease also known as Angiofollicular lymph node hyperplasia, giant lymph node hyperplasia,angiomatous lymphoid hamartoma, benign giant lymphoma presently defined as a heterogeneous disease with different etiologies that share common histologic patterns. Mostly it presents as lymph node mass often mediastinal or cervical. A patient presented to us with a large retroperitoneal mass without any systemic symptoms. After surgical excision of the mass, histopathology findings suggested it to be a case of hyaline vascular variant of castleman disease.

INTRODUCTION:
Castleman disease is a rare cause of lymphadenopathy. In original reports by Castleman et al,Castleman disease was described as a large ,benign,asymptomatic mass involving mediastinal lymph nodes.Subsequent to recognition of hyaline vascular variant,plasma cell variant of CD ,both localized and multicentric and association of human herpes virus 8(HHV-8) in a subset of multicentric cases ,CD is presently defined as heterogeneous and multiple distinct disease with different etiology that share common histological patterns. It must be differentiated from other causes of lymphadenopathy either clinically or pathologically like tuberculosis, lymphoma,HIV. We present a case of castleman disease presenting as large retroperitoneal mass which on excisional biopsy found to be a case of hyaline vascular variant of castleman disease.

CASE REPORT:
A 23 year old female presented to us with swelling in the lower abdomen for four years. The swelling was insidious in onset and gradually increasing in size. She complained of dull aching pain in the lower abdomen for 6month, not associated with vomiting,fever,weight loss,night sweats,bowel bladder irregularity or any other swelling in the body. She was having regular menstrual cycles with average regular flow.Her last menstrual period was 20 days back. P_2L_2.Last child birth 13month back. No history of tuberculosis,hypertension,and diabetes.

On examination BMI 19,pallor is present,vitals normal,per abdominal examination revealed a globular swelling of size 15×12×3 cm in hypogastric and right iliac fossa region which is non tender,having smooth surface,regular margin and firm in consistency,immobile. Swelling was found to be retroperitoneal.Investigations like complete blood counts,serum electrolytes,BUN, creatinine, fasting blood sugar, and liver enzymes normal. Serum LDH level was normal.Her serum for hepatitis markers and HIV was negative.Ultrasound of abdomen and pelvis suspected inflammatory lesion and advised CT-scan.. CT-scan of abdomen and pelvis showed well marginated soft tissue attenuating mass in the right iliac fossa, anterior to the right psoas muscle-? BENIGN NEOPLASTIC MASS

Patient underwent surgery under general anesthesia. Right paramedian incision given and retro peritoneum entered by incising along the White Line of Toldts. The mass was not fixed to any other structure. It was excised with proper hemostasis. Patient recovered uneventfully.

Histopathology report came out to be hyaline vascular variant of castleman disease.Lymphoid follicles are increased throughout the cortex and medulla, and often contain two or more germinal centers (so-called "twinning).The mantle zones are composed of concentric rings of small lymphocytes (onion skin pattern). Hyaline deposits prominent within germinal centers found. Sclerotic blood vessels radially penetrate the germinal centers,forming HV lesions also known as 'lollipop lesions' found.The interfollicular regions are composed of numerous high endothelial venules with plump endothelial cells and sclerotic walls.

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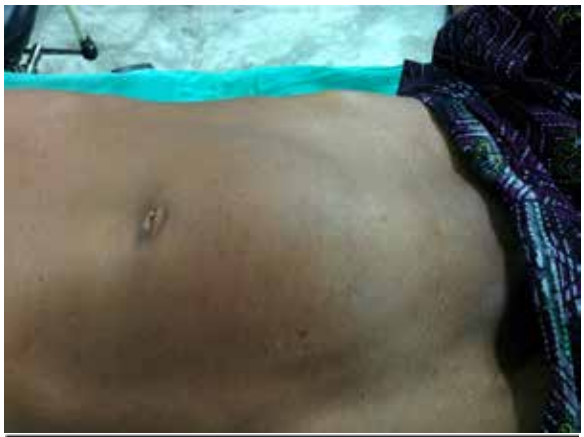
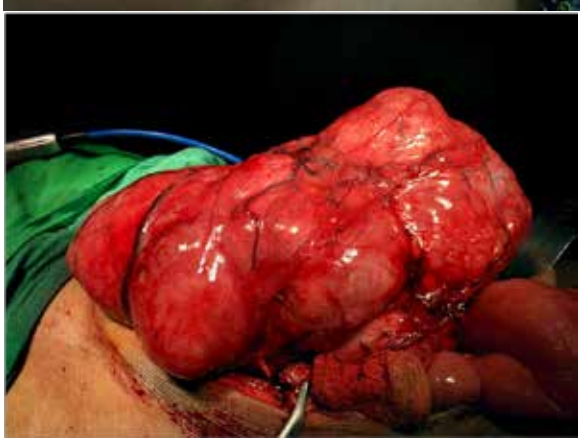
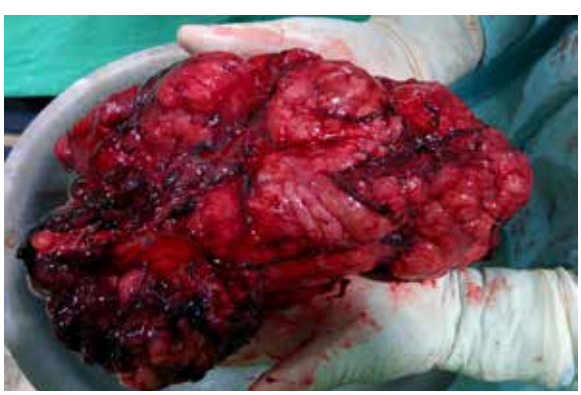
Patient is now on follow up.



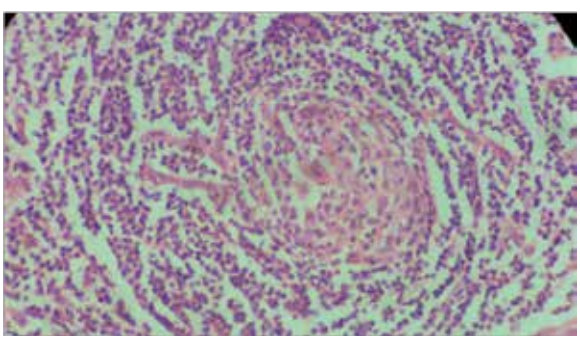




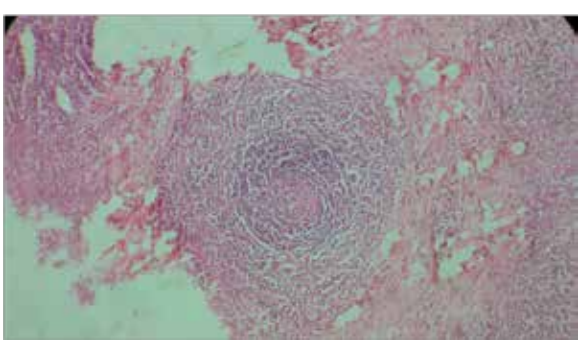
DISCUSSION:

Castleman disease is a rare disorder characterized by benign growth that involves lymph node tissue throughout the body.Most often,they occur in mediastinum,neck and abdomen but may also be found in peripheral sites containing lymph nodes. Clinically it can be unicentric affecting lymph nodes in only one part of body or multicentric affecting multiple parts of body. Histologically can be hyaline-vascular type (80-90%) and plasma cell type (10-20%). The causes of CD are mostly unknown. One known etiologic agent is HHV-8 (in 50% cases of plasma cell variant CD, including most multicentric cases, and in most cases with HIV infection). Abundant evidence indicates a role for IL-6 in the pathogenesis of PC variant of CD. Some evidence also suggests immune dysfunction in patients with PC-CD. Much less is known about pathogenesis of HV variant CD. Hyaline vascular variant usually presents as a large mass involving a lymph node (or a group of lymph nodes). Symptoms when present are related to size of the mass and compression of contiguous structures. Laboratory abnormalities other than elevated serum LDH levels in a subset of patients are rare. The plasma cell variant can be asymptomatic but systemic symptoms like fever, night sweats, weight loss, and malaise may be present. Laboratory studies most commonly show anemia or thrombocytopenia and increased serum IL-6 levels. Multicentric CD is mostly seen in patient with HIV infection and associated with HHV-8 infection. B-type symptoms (fever, night sweats, weight loss) occur in over 95% of patients. Splenomegaly (~ 75%) and hepatomegaly (~50%) are common. Edema, body cavity effusions, skinrash, and neurologic changes can occur in a small subset of patients.

The localised nature of HV-CD usually allows complete surgical excision, which is curative. Radiation therapy can also have been used in patients whom complete surgical excision could not be performed. For asymptomatic patients with localised PC-CD, surgical excision appears to be adequate. For symptomatic patients chemotherapy and steroid treatment along with surgical excision gives a good response. Patients with MCD without neuropathy, POEMS syndrome, or HIV infection can be treated with systemic chemotherapy and steroids with some success.For patients with these association requires innovative approach as prognosis is generally poor.



Hyaline vascular ("lollipop") lesion in which a follicle is radially penetrated by a sclerotic blood vessel.Hematoxylin and eosin stain.



The follicle is surrounded by a broad mantle zone composed of concentric rings of small lymphocytes (so called "onion skin").*Hematoxylin* and eosin stain

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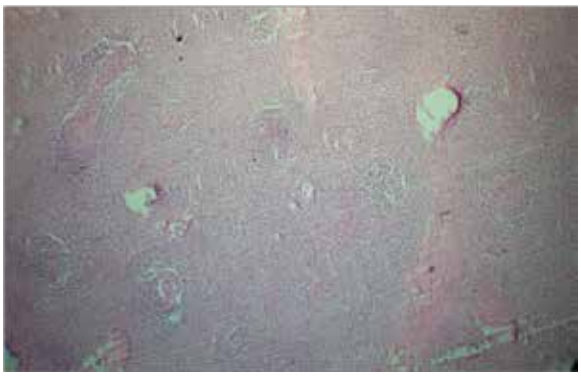

The follicle to the upper left of the field contains more than one germinal center ("twinning"). Hematoxylin and eosin stain

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