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Case Report - Retroperitoneal Castleman's Disease Masquerading as Tumour Mass

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Abstract

Castleman disease (CD) is a rare, localised or generalised lymphoproliferative disorder. More frequent in mediastinal location. In our case, the patient presented with retroperitoneal mass in the right lumbar region. After evaluating the patient (CECT KUB, MRI), surgical resection was performed. The diagnosis of Castleman's disease-hyaline vascular type was made after histopathological examination of the mass.

Keywords: Castleman's disease, retroperitoneal mass, multicentric disease

Introduction

Abdominal masses always pose a dilemma for the surgeon and due to varied presentation of the tumours pre-operative diagnosis is always difficult.[1]

Castleman's disease (CD) also known as an Angio follicular lymph node hyperplasia or giant lymph node hyperplasia is a rare benign tumour of lymphoid origin with unknown etiology.[2]

Case summary

An 18-year-old male presented to the medicine department with complaint of pain in the right leg for 3 months associated with tingling sensation which increased on walking. On examination no neurological deficit was noted.

He was evaluated for the same. CECT KUB [Fig.1] suggests 8.7 x 7.5 x 8cm lesion in the right paravertebral region extending from T1-L2 displacing portal vein, IVC, right crus of diaphragm, right kidney with fat planes maintained. Few sub centimeter lymph nodes in paracaval/ paraaortic/ aortocaval lymph nodes. MRI spine LS suggests 9.4x4x7.7 cm lesion, right paravertebral aspect of suprarenal region? Ganglio neuroblastoma? neurogenic tumour.

Patient was taken for surgery to remove the mass under general anaesthesia. Informed consent was taken before surgery. Intraoperative and postoperative period was uneventful. We have received specimen of right adrenal gland with attached mass in 10% buffered formalin. The

mass measures 8.5x7x5 cm, weighing 10.5 g. Externally mass was bosselated with intact capsule. Cut section was homogenous and grey white [Fig.2]. Tissue sections were processed and the slides were stained with haematoxylin and eosin(H&E). Histopathological examination of the mass showed follicles with atretic germinal centres [Fig.3] traversed by penetrating vessels (lollipop follicles) [Fig.4]. Mantle zone was thickened with lymphocytes arranged in layers giving onion – skin appearance. Interfollicular area showed extensive vascular proliferation with perivascular hyalinization along with infiltration of lymphocytes, eosinophils and plasma cells.

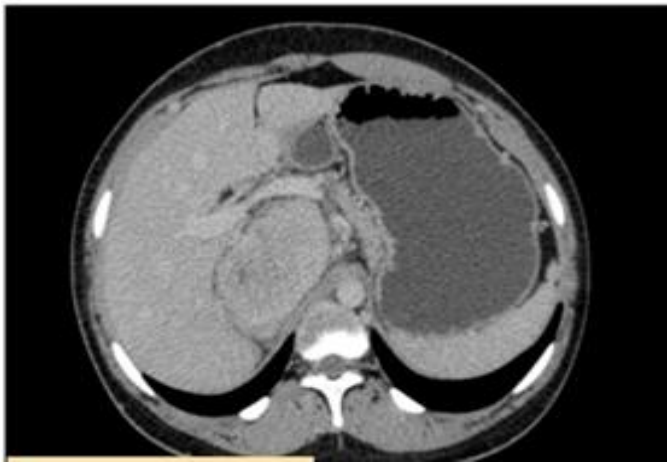

Fig 1: CECT KUB showing lesion in right paravertebral region.

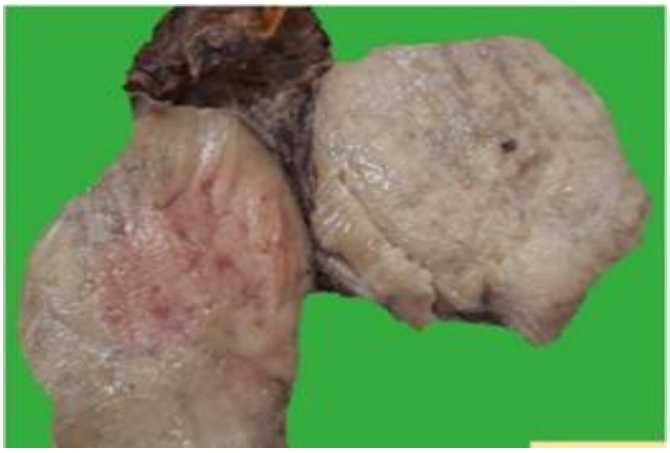

Fig 2: Gross specimen of adrenal gland with attached mass

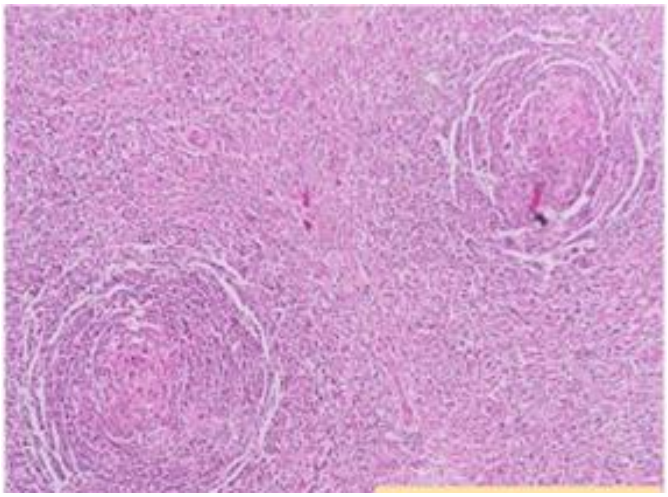

Fig 3: Photomicrography showing atretic centres trans versed by penetrating vessels (H&E, x100)

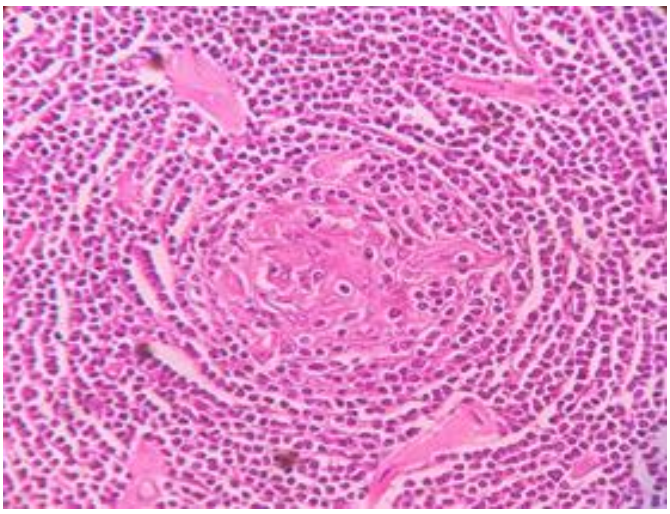

Fig 4: Photomicrography showing onion skin appearance (H&E, x400)

Discussion

CD was first reported by Symmers in 1921 and the pathology characterised by Benjamin Castleman in 1956 as a benign lymph node hyperplasia.[3] It rarely presents as retroperitoneal mass. It affects all age groups with no predilection for either gender and occurs anywhere along the lymphoid chain.[4]CD is found primarily in the mediastinum. Mesenteric, retroperitoneal, and pelvic tumours are described less frequently.[5]

Castleman disease based on their histological feature divided into hyaline vascular (HV) type and plasma cell

(PC) type. Clinically, CD can be divided into unicentric disease (localised form) and multicentric disease (MCD) (generalised form).MCD can be divided into Kaposi sarcoma herpesvirus (KSHV)-associated MCD(KSHV-MCD) and idiopathic MCD (iMCD), which is KSHV negative.[6] The majority of localised disease is hyaline vascular type.[7] It's manifestations are heterogenous, ranging from asymptomatic discrete lymphadenopathy to recurrent episodes of diffuse lymphadenopathy with severe systemic symptoms.[8]CD in retroperitoneal and abdominal location presents with mass effect symptoms such as anorexia, weight loss, vomiting, urinary retention, and abdominal pain.[9] The plasma cell (PC) type presents with systemic symptoms such as fever, anaemia, weight loss, elevated ESR levels, hyper gammaglobulinemia or hypoalbuminemia.[3]

The CD associated with chronic human herpes virus (HSV8) also known as Kaposi's sarcoma associated herpes virus (KSHV). Lesions are made of polyclonal proliferation confirming its non-neoplastic nature. It occurs as a result of chronic low grade inflammatory process triggered by latent infection with HSV8, which leads to lymphoid system hyperplasia and also stimulates secretion of IL-6.[3]

Radiologically, CD is seen as a homogeneously hypoechoic mass on USG and non- specific enhancing homogeneous mass with micro calcification on CT. But both of these are not diagnostic. Due to low specificity, preoperative fine needle aspiration cytology has no role in diagnosis. A definitive diagnosis can only be made by pathological examination of the biopsy tissue or resected specimen.[1]

Macroscopically CD appears as an encapsulated homogeneous mass with an orange yellowish colour. Microscopically, is characterised by follicular and interfollicular capillary proliferation with perivascular hyalinization and atretic germinal centres traversed by penetrating vessels (lollipop follicles). Mantle zones are thickened with lymphocytes arranged in layers giving it an onion skin appearance. Mantle zones may fuse and contain more than one germinal centre (twinning).[1]

The standard therapy for the localised form is en bloc surgical excision. Since these lesions are hypervascular, embolization before surgery could be of help in reducing blood loss during surgery. For the systemic form, corticosteroid therapy, immunosuppressive drugs, chemotherapy, and radiotherapy have been tried without any convincing results.[1]

Conclusion

Localised form, though a rare entity, should be considered in the differential diagnosis of retroperitoneal mass. A knowledge regarding this disease would help the surgeon to prevent unnecessarily extensive resection for this benign disease.

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