

Unicentric Castleman's disease presenting as retroperitoneum lump

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ABSTRACT

Castleman disease is a rare lymphoproliferative disorder, also known as giant lymph node hyperplasia or angiofollicular lymph node hyperplasia. Clinically, Castleman disease is of two types: localised/unicentric type and multicentric/systemic type. Unicentric or localised Castleman disease affect a single lymph node or group of lymph nodes. The multicentric type affects two or more groups of lymph node in different part of the body. It can also affect organs containing lymphoid tissue. Histologically it is classified as hyaline vascular variant, plasma cell variant and a mixed variant. Clinical symptoms may vary from asymptomatic to symptomatic lymphadenopathy accompanied by fever, anaemia fatigue, abdominal or thoracic pain and weight loss. There is no specific test to diagnose Castleman disease. We report a case of 16 years old male who presented with a painless lump in left lumbar region without any constitutional symptoms. CECT suggested a retroperitoneal lump with calcification. Laparotomy was done and complete excision of mass done. Histopathological examination of excised tissue suggested Castleman disease of hyaline vascular variant. After six month of follow up, the patient has no complain.

Keywords: POEMS, castleman's disease, hyaline vascular variant, lymphoproliferative disorder, lump, lymph node

Introduction

Castleman's disease or angiofollicular lymph node hyperplasia is a rare disorder characterised by benign proliferation of lymphoid tissue. It was first described by Benjamin Castleman in 1950. It may be localised / unicentric or disseminated /systemic/ multicentric. Histologically, it is classified as hyaline vascular type, plasma cell type and a mixed type. ^[1] The unicentric type can be treated and cured by surgery. To date, approximately 1000 cases of Castleman's disease have been reported in the literature. ^[2, 4] We report

Case Report

A 16 year old male presented with complain of painless lump in the left lumbar region of three months. The patient did not have any constitutional symptoms. On physical examination a

mobile lump of size 9.0 cm x 8.0 cm noted in the left lumbar region. The lump was mobile, had nodular surface and was firm in consistency. There was no family history of similar illness or cancer. Routine laboratory investigation and chest X-rays were normal. Abdominal sonography revealed a hetero echoic mass of size 9.0 cm x 8.0 cm with calcification in left lumbar region. CECT revealed hypodense enhancing mass measuring 9.0 cm x 8.0 cm x 6.5 cm with rim like calcification [Fig 1].

At laparotomy, the mass was found attached to the root of mesentery and complete resection of mass was undertaken. Macroscopically, the mass measured 9.0 cm x 8.0 cm x 6.0 cm. The tissue was grey white, capsulated and lobulated. (Fig.2) Cut section shows grey white hemorrhagic surface.

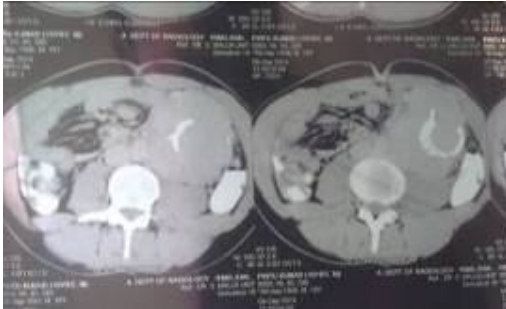


Fig.1 CECT showing retroperitoneal mass with calcification

Microscopic examination showed the tissue having few small follicular centres evenly distributed, surrounded by concentric layers of lymphocytes having prominent central vessels with hyalinized walls and proliferating endothelial cells. The interfollicular area shows proliferation of small blood vessels surrounded by a cuff of collagen with variable number of plasma cells and immunoblast. (Fig.3) A diagnosis of Castleman disease, hyaline vascular type was made. The patient had an uneventful postoperative course.



Fig. 2 Specimen

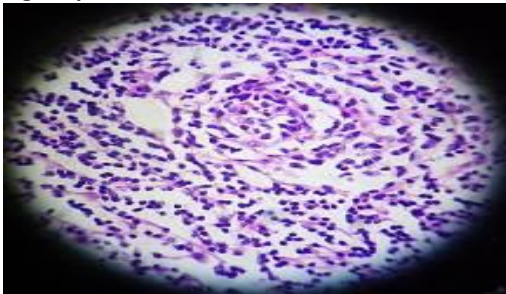


Fig. 2 HPE at 100x showing Angiofollicular Castleman disease (H & E)

Discussion

Castleman disease represents a morphologically distinct form of lymph

node hyperplasia. Clinically Castleman's disease has been divided into solitary/unicentric and multicentric/systemic form. Unicentric Castleman disease is the most common type and consists of an isolated lymphoproliferative disorder of young adults that is not associated with an HHV-8 infection and usually curable with surgical resection. The solitary form present as a mass located most commonly in the mediastinum and less commonly in the neck, lung, axilla, mesentery, broad ligament, retroperitoneum, soft tissue extremities.^[3] Majority of the patient are asymptomatic. Pre-operative diagnosis of hyaline vascular Castleman disease is difficult. The usual appearance of the disease on CT or MRI is that of a mass with or without calcification with enhancement on post contrast study.

The histological subtype of Castleman's disease is: (Table 1) hyaline vascular variant (unicentric in 72%); plasma cell variant (unicentric in 18% and multicentric in 10%); mixed variant and a plasmablastic variant of Castleman disease.^[4] Only 10% to 17% cases involve the abdomen, of which the majority of cases are retroperitoneal. The plasmablastic variant of the disease is commonly associated with HHV-8 and tends to have a poor prognosis. The multicentric and especially, HHV-8 positive types are risk factors for the development of Hodgkin lymphoma, non-Hodgkin lymphoma and Kaposi sarcoma. It has been hypothesized that HHV-8 induces the changes of Castleman disease through the production of interleukin-6. Multicentric Castleman disease may be seen in association with the POEMS syndrome.^[5] Most patient with multicentric Castleman disease die from progression of their disease, disseminated infection or related malignancies.

Table: 1 Overview of the histological variants and sex and age distribution of localized retroperitoneal Castleman's disease in the literature

Histological variants				Distribution of sex			Distribution of age (in years)				
HV	PC	Mixed	NK	M	F	NK	Age	M	F	NK	
148	9	5	37	69	79	51	0 to 9	2	0	52	
74.37%	4.52%	2.51%	18.59%	34.67%	39.7%	25.63%	10 to 19	9	6		
							20 to 29	7	21		
							30 to 39	16	18		
							40 to 49	14	13		
							50 to 59	7	7		
							60 to 69	12	3		
							70 to 79	1	0		
In total: 199 cases				M:F ratio: 1:1.14			Mean		38.25		

F=female, M=male, HV=hyaline vascular, PC=plasma cell, NK=not known

Laboratory studies with blood count, C-reactive protein, interleukin-6, HIV, HHV-8 testing should be done. CT and MRI is non diagnostic. Kim et al described two types of radiological manifestation in abdominal Castleman disease: a localised type and a disseminated type.^[6] The hyaline vascular variant shows high enhancement on CT whereas plasma cell variant shows low enhancement. In our case the mass showed a high degree of enhancement on post contrast CT scan. In a recently published study, 'rim like' enhancement, a predominantly left sided location in the retroperitoneum, and the presence of peritoneal thickening were described as relatively characteristic findings of the disease. Non-specific features like necrosis, fibrosis and calcifications can be observed. In our case, mass was located on left side with a rim like calcification.

A variety of treatments have been used for multicentric Castleman disease including surgery, radiation, steroids, antiviral agents, specific antibodies, inhibitors of cytokines activity and chemotherapy. Surgery generally does not

have a role in the treatment of multicentric Castleman disease. The unicentric type if resected completely is not associated with increased mortality and is known to have excellent prognosis. Therefore, radical surgical resection is considered to be gold standard therapy in these cases.^[5] Patient with unicentric disease should receive radiological follow up every 6 to 12 months.

Conclusion

Castleman's disease is a rare disorder that remains a diagnostic challenge. Radical surgical resection is considered to be gold standard for treating the unicentric variant. This type is not associated with increased mortality as long as resection is radical and complete.

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