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Original Research Paper

General Surgery

CASTLEMAN DISEASE - A RARE CASE REPORT

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ABSTRACT **Castleman disease,** also known as **angiofollicular lymph node hyperplasia** or **giant lymph node hyperplasia,** is an uncommon benign B-cell lymphoproliferative condition. . It can affect several regions of the body although commonly described as a solitary mediastinal mass.There are mainly 2 types (1) Unicentric and (2) Multicentric and two basic histopathologic pattern that are hyaline-vascular(HV) and plasma cell(PC) type.

KEYWORDS :

INTRODUCTION

Castleman disease was first described by Benjamin Castleman in 1914.

Castleman disease is unusual benign B-cell lymphoproliferative disorder that involves LN but extranodal sites can also be affected.

There are mainly 2 subtype
1) Unicentric Castleman disease
- involve single site
- Localized and locally treated
- Associated with good prognosis
- Cured by surgery

2) Multicentric Castleman disease –
- involves more than one sites
- Usually in context with HIV infection
- Caused by HHV-8
- Systemic disease and characterized by lymphadenopathy, splenomegaly and anemia
- Systemic inflammation symptoms such as fever, malaise and body ache present
- Poor prognosis and recurrence common after surgery

EPIDEMIOLOGY-

6500-7000 new cases diagnosed each year in USA. Common in 3rd and 4th decade and more common in female than male.

PATHOPHYSIOLOGY-

The disease is of unknown aetiology, but most widely accepted theory is that Castleman disease is a chronic low-grade inflammatory process. The interleukin-6 in Unicentric Castleman disease and both Interleukin-6 and HHV-8 in multicentric Castleman disease are demonstrated to play a role in pathogenesis and symptomatology of the disease.

PRESENTATION-

Unicentric Castleman disease- incidentally found

Multicentric Castleman disease- Due to systemic inflammation symptoms such as fever, night sweat, fatigue, weight loss are common. Also associated with genralised lymphadenopathy, hepatosplenomegaly and fluid retention.

INVESTIGATIONS-
- Hematologic investigation are non specific; anemia may be present.
- Radiological investigations are nonspecific and shows enlarged lymph node like mass lesion.

DIAGNOSIS –

Biopsy and histopathological examination are diagnostic modality of choice.

On pathological examination disease is characterized by hypervascular lymphoid hyperplasia. There are also subtype based on pathologic examination

Hyaline vascular:
- most common ~90%, corresponding to most cases of the unicentric Castleman disease
- most commonly found in the mediastinum
- classically appears as an avidly enhancing mediastinal mass at CT and MRI
- increased numbers of small hyalinised blood vessels within and between the lymphoid follicles, which show obliteration of the medullary sinuses
- "onion-skinning" arrangement of the small lymphocytes of the mantle zones, forming concentric rings around the germinal centre

Plasma cell
- often multicentric
- hyperplastic follicles of varying sizes, which show patent medullary sinuses
- less enhancing
- may be more symptomatic

HHV Associated Castleman disease

Multicentric Castleman disease not otherwise specified

Castleman disease is usually diagnosis of exclusion and diagnosed after histopathologic examination.

TREATMENT-

For Unicentric Castleman **disease** treatment is surgical, with good prognosis(can be curative)

Multicentric Castleman disease may be treated with any combination of surgery, chemotherapy and prednisolone. Antiviral agents (targeting HHV 8) and monoclonal antibody therapies targeting CD20 or IL-6 . prognosis is relatively poor.

CASE REPORT

A 29 year old female presented to civil hospital Ahmedabad with c/o left side upper abdominal dull aching pain since 1 year without any aggravating or reliving factor and no associated jocomplaints.

She was married since 7 year and was trying to get pregnant but was unable to conceive. She was started on clomiphene citrate for treatment of PCOD(polycystic ovarian disease). Otherwise her medical and surgical history was unremarkable.

She was vitally stable and her per abdomen examination was normal with no palpable organ or palpable lump.

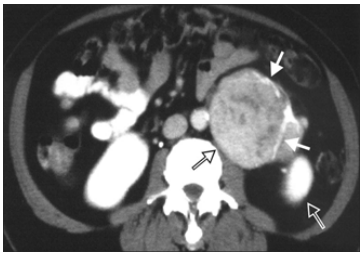
We investigated patient by USG (abdo+pelvis) which was s/o approx. 51*35 mm sized well defined homogenously hypoechoic solid lesion with internal vascularity is noted in left paraumbilical region without any communication with bowel loops p/o benign mesenteric mass lesion.

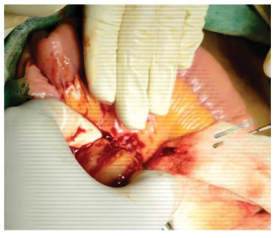
We did CECT(abdo+pelvis) of this patient which was showing well defined homogenously enhancing mass lesion arising from mesentry with p/o abdominal desmoid tumor.

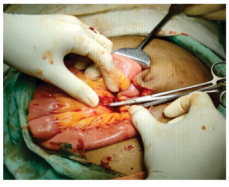
We operated this patient and found out approx. 5*4 cm size well defined lesion in mesentry of jejunum without any communication or adhesion with bowel and no connection with mesenteric vessels. Rest of the organs were normal and no other mass lesion found out on exploration.

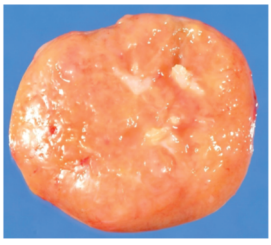
Biopsy report was s/o structure of lymph node with marked reactive follicular hyperplasia with evidence of plasma cell infiltration in parafollicular region p/o Castleman disease.

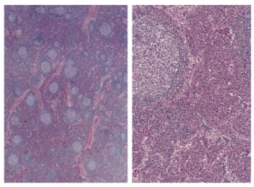
Patient had uneventful post-op course and was discharged on post op day 4.











CASTLEMAN'S DISEASE, PLASMA CELL TYPE: LYMPH NODE
Left: Reactive follicles are separated by an appreciable amount of vidaseous-staining interfollicular tissue.
Right: The interfollicular tissue comprises predominantly sheets of plasma cells.

DISCUSSION

Castleman disease is a rare diagnosis and extranodal involvement is even more rare as the usual site of occurrence is mediastinum, diagnosed on HPE and it is a diagnosis of exclusion.

In our patient radiological investigation was suggestive of intra-abdominal desmoid tumor but it turned out to be Castleman disease on HPE. This highlights that one should not rely on CECT finding so much; one needs to keep Desmoid tumor and GIST as other possible diagnosis for intraabdominal mesenteric mass lesion as post operative management is very different.

Patient's postoperative course was uneventful and for unicentric Castleman disease surgical excision is curative.