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Retroperitoneal castleman's disease: A rare case report

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Abstract
Introduction: Castleman's disease is a rare lymphoproliferative disorder which may be confused with other cause of lymphadenopathy.
Case report: Here, we report a case of unicentric Castleman's disease presenting with retroperitoneal lymphadenopathy. This case has been reported for its rarity.
Conclusion: Though castleman's disease is a relatively rare entity. This case report brings to light the importance of obtaining definitive histological diagnosis in patients presenting with lymphadenopathy and systemic symptoms. This case emphasis the importance of histology to seal the diagnosis from the other lymphoproliferative conditions.

Keywords: castleman's disease, retroperitoneal, lymph node

Introduction
Castleman's disease (CD) is a heterogeneous group of lymphoproliferative disorders of uncertain cause [1] presenting with lymphadenopathy. It is histologically and prognostically distinct from malignant lymph node hyperplasia. It was first described in a group of patients with benign localized hyperplastic lymph nodes in 1956 by Castleman *et al.* [2].

Case presentation
History
A 25 years old female presented with complaints of right side abdominal pain on & off along with the generalized weakness. She had no past medical history.
Sonography was showing mesenteric mass measuring 10.9 x 1 x 7.2 cm. Computerized tomography revealed enhancing mass in retroperitoneal region posterior to ascending colon measuring 8.3 x 6.3 x 10.2 cm, suggestive of Angiosarcoma. Patient underwent exploratory laparotomy excision of retroperitoneal tumor and this was reported as Castleman disease.

On gross examination
Single grey/ brown capsulated mass 10 x 5.5 x 5.5 cm was received at laboratory in Inamdar hospital. External surface unremarkable. C/s was grayish/ yellow, firm. Relevant sections from the mass were given and formalin fixed, paraffin embedded blocks prepared from them.

Fig 1: Photograph showing retroperitoneal mass, external surface unremarkable. Cut surface showing grayish yellow in colour with firm consistency.

On microscopic findings

Haematoxylin & Eosin stained sections showed lymph nodal mass with partially effaced architecture. Vascular proliferation with perivascular hyalinization noted. Mantle zone shows onion skin appearance. Lymphoid and sinusoidal hyperplasia noted.

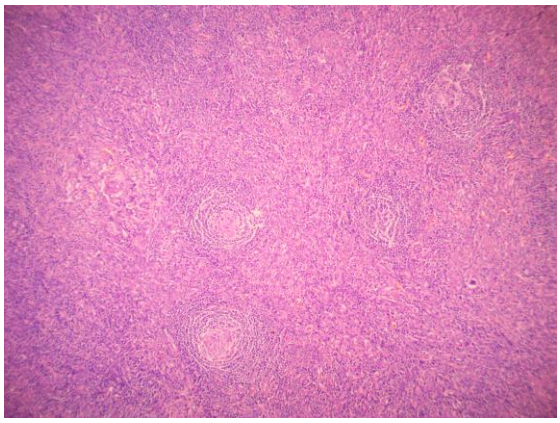

Fig 2: Micrograph showing lymph node mass with partially effaced architecture. Vascular proliferation and perivascular hyalinization noted. Mantle zone shows onion skin appearance.

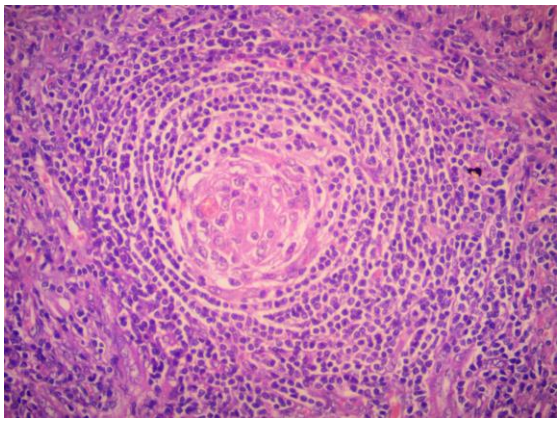

Fig 3: Micrograph showing vascular proliferation and perivascular hyalinization with onion skin appearance in mantle zone.

Discussion

Castleman's disease (CD) is lymphoproliferative disorder which is histologically characterized by angiofollicular lymph-node hypertrophy [3]. It may be borne in mind in the differential diagnosis of localized/ diffuse lymphadenopathy with or without systemic manifestations. This case report, together with a review of medical literature, attempts to provide new insight into this rather rare and benign disorder, though mimicking mass clinically, varies from the latter histologically [4].

There are 2 different clinical entities: the unicentric type which only one anatomic lymph node affected and the multicentric type characterized by generalized lymphadenopathy, constitutional symptoms and more aggressive clinical course [5].

Localised CD is localized to one site by definition. It features lymphoid hyperplasia associated with excessive angiogenesis [1]. It is asymptomatic in over 50% of patients[6] and is often discovered incidentally. Histological diagnosis requires lymph node biopsy [4].

Multicentric CD is characterized by a predominantly lymphadenopathy presentation consistently involving peripheral lymph nodes and manifestations of multisystem involvement. It is considered as a systemic B cell lymphoproliferation, probably arising in immunoregulatory deficit, and resulting in the outgrowth of clonal B-cell populations [1]. It is always symptomatic. Symptoms, primarily a consequence of elevated interleukin-6 (IL-6) production, are asthenia (65%), weight loss (67%) and fever (69%) [3]. Polyadenopathy is common (84%) with a mean of four sites involved and is often associated with hepatosplenomegaly[3]. Histological diagnosis is made upon biopsy of an excised lymph node [4].

Histologically the main types are:

- The hyaline vascular type characterized by lymphoid follicular proliferation at different levels of maturity, often forming a layered or 'onionskin' pattern surrounding a hyalinised vessel at center of the follicle. These vessels are often prominent and reactive. This is more commonly seen in the localized form of the disease.
- The plasma cell variant has sheets of mature plasma cells within the interfollicular tissues surrounding larger germinal centers and has significantly less vascularity. The multicentric form of the disease is nearly always associated with this variant.
- A third histological variant showing a mixed picture can also been in MCD [7].

Localised CD is treated by surgical excision which allows full recovery without relapse in almost all cases. However, no therapeutic consensus exists for MCD and diverse treatments (surgery/ corticotherapy/ chemotherapy) are used, often in combination [3]. Anti-interleukin-6 antibody has also been successfully tried in the alleviation of systemic manifestations [8]. The five year survival rate in MCD is 82 % [3] and this prognosis appears to be far better than that encountered with malignant lymphomas [4].

Conclusion

Though castleman's disease is a relatively rare entity. This case report brings to light the importance of obtaining definitive histological diagnosis in patients presenting with lymphadenopathy and systemic symptoms. Unicentric Castleman's disease is a relatively uncommon cause for such a presentation. Though clinically synonymous with lymphoma. It is an entity that is distinct from malignant lymphoproliferative disorders histologically and prognostically.

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