CASE REPORT

Pelvic Castleman's Disease Presenting as an Ovarian Mass

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Introduction

Castleman's disease is an uncommon disease with poorly understood etiopathogenesis. It was described by Benjamin Castleman in 1954. It commonly occurs in the mediastinal nodes. Virtually any lymphnodal area can be affected. Extranodal involvement is rare and occurs in the lung, pancreas, breast, adrenals, and muscle. Castleman's disease rarely occurs in the pelvic cavity and only 18 such cases are described. There are very few case reports of pelvic Castleman's disease in the Indian literature. The present case is Castleman's disease in the broad ligament mimicking as an ovarian mass.

Case History

A 26-year-old woman presented with complaints of vague pelvic pain. It was on the left side not associated with urinary or bowel symptoms. There was no history of fever. Her menstrual history was regular. She was a nulliparous woman. On examination, she was a moderately built

female. There was no pallor or jaundice and no peripheral lymphadenopathy. There was no organomegaly. Per vaginal examination revealed a mass in the left adnexal region. An ultrasound of the pelvis revealed a 6.0×5.0 cm hypoechoic homogenous mass in the left broad ligament with regular borders. No other masses were seen. Her chest X-ray was normal. Her routine hematological and biochemical investigations were within normal limits. Her retroviral and hepatitis B screening was negative. With a provisional diagnosis of ovarian mass, she was taken for surgery. At surgery, a soft to firm mass was seen in the broad ligament, not attached to the ovary. The mass was moderately vascular. The mass could be easily separated from surrounding structures. It was excised in toto and submitted for histopathology.

On gross examination, it was an oval mass 6.0×5.0 cms. The surface was smooth. Cut section was homogenous gray white and well circumscribed.

Microscopic examination

Routine Hematoxylin and Eosin sections revealed lymphoid tissue with follicles. There was an expansion of the mantle zone with many germinal centers. There was vascular proliferation and hyalinization within the germinal centers (Fig. 1). The interfollicular area revealed proliferation of post-capillary venules with an admixture of plasma cells and eosinophils. Cells forming the mantle zone expressed CD20. Bcl 2 was negative. A diagnosis of

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Fig. 1 **a** Lymphocytes arranged in onion skin pattern around hyalinized vessel. H&E $\times 200$. **b** CD20 immunostain showing positive cells in mantle zone $\times 200$

Castleman's disease hyaline vascular type in the broad ligament was made. The patient had an uneventful post-operative period and is symptom free on follow up.

Discussion

Castleman's disease is a rare disorder characterized by proliferation of lymphoid tissue. It is also called angiofollicular lymphoid hyperplasia. It was first reported by Symmers in 1921. The pathology was later described by Castleman in 1954 as a benign lymphnodal tissue resembling thymoma. Keller et al. in 1972 identified two histologic types, the Hyaline vascular type and the Plasma cell type. Two clinical subtypes are described, a localized or unicentric type and a multicentric type. Castleman's disease can occur in almost any area where lymphoid tissue is seen. In one large series, 70 % involved the thorax, 40 % in the neck region, 12 % in the abdomen, and 4 % in the axilla. Rare sites are the lung, broad ligament, leptomeninges, soft tissues of extremities [1], and parotids. Castleman's disease can occur at any age ranging from 8 to 66 years. Males and females are equally affected. Etiology is not clearly known. HSV8 is implicated in the systemic form. It is suggested as a low-grade inflammatory process triggered by HSV8, which leads to increased production of IL-6, leading to hyperplasia of the lymphoid tissue. The localized form is seen in immunocompetent patients, whereas the systemic form is seen in immunosuppressed patients. Localized lesions cause few symptoms, whereas the systemic form is associated with anemia, thrombocytopenia, increased erythrocyte sedimentation rate, and hypergammaglobulinemia. Clinically, patients may present with generalized lymphadenopathy, hepatomegaly, splenomegaly, impaired renal function, or polyneuropathy.

On rare occasions, POEMS syndrome is associated with Castleman's disease. It is also associated with glomeruloid hemangioma, amyloid deposits, and follicular dendritic cell sarcoma. Differential diagnosis includes Hodgkin's lymphoma, viral lymphadenitis. The lesion was completely excised with no local recurrence two and half years after surgery.

Castleman's disease is a rare disorder. Its occurrence in extranodal sites and in the pelvis is even rarer. Only few cases have been described in the literature [2]. In the Indian literature, one case of broad ligament Castleman's disease was described by Mondal et al. The localized form has no specific symptoms, hence preoperative diagnosis is difficult.

Conclusions

Although very rare, it is important to keep Castleman's disease in the differential diagnosis of pelvic masses whenever we encounter lymphoid hyperplasia in rare sites. Castleman's disease should be considered in the differential diagnosis and differentiated from malignant lymphoid proliferation.

References

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