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RETROPERITONEAL UNICENTRIC CASTLEMANS DISEASE – A RARE CASE REPORT

Pathology

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ABSTRACT

Castleman disease is characterized by proliferation of lymphoid tissue. Clinically it presents in two forms either a unicentric or multricentric. Most common location is mediastinum, neck, axilla and pelvis. Unicentric retroperitoneal location is rare. Three general histologic patterns are seen in Castleman disease: (1) Hyaline vascular (2) hypervascular (3) Plasma cell rich, mainly seen in patients with multicentric disease. **Case report** 49 year female came to our hospital with complaint of generalized abdominal pain since 1.5 months. Physical examination was unremarkable. No history of any oswellings in any part of her body. HIV, HBsAg and Anti-HCV antibodies were negative. Other investigations including hematological and biochemical tests were normal. The chest X-ray did not reveal any mediastinal mass. The CT IVP findings are suggestive of a well defined intensely homogenously enhancing soft tissue density lesion in anterior para renal space on left side with adjacent fat stranding and enlarged lymph nodes. Possibility of 1) arteriovenous malformation 2) Reactive lymphoproliferative disease 3) Castleman disease. Complete surgical excision was done. Pathological findings showed localized hyaline vascular type of Castleman's disease.

KEYWORDS

Castleman's disease, unicentric , retroperitoneum, hyaline vascular

INTRODUCTION

Castleman disease represents a very rare cause for development of retroperitoneal mass. CD is more common in young adults without predilection of sex, although age varies from 8 to 66 years. CD may occur anywhere along with the lymphoid chain[2]. Castlemans disease was described in 1954 at first time and known as an unusual nonmalignant lymphoproliferative disease. Most commonly Castlemans disease presented with enlarged hyperplastic lymphnode tissue[3]. It has been described mainly in case reports and small series, thus the incidence and prevalence of castleman disease is difficult to ascertain [5]. The reason is unidentified, and it is considered by the growth of neoplastic masses of lymphoid organ. Castlemans disease OR angiofollicular lymph node hyperplasia, which involves hyperactivation of the immune system with multiple organ system dysfunctions[1]. Two forms are unicentric (UC) and multicentric (MC), based on histology it is further subclassified into a hyaline vascular (HV) type and a plasma cell (PC) type. Unicentric castlemans disease affects mainly mediastinum and retroperitoneal location is rare[4].

DISCUSSION:

Castleman's disease is characterized by an angiofollicular lymphnode hyperplasia. There are two presentations of Castleman's disease according to the number of lymphnodes involved 1) unicentric CD and 2) multicentric CD [6]. Unicentric Castleman's disease is a localized disease affecting a single enlarged lymph node or a group of adjacent nodes. It is the most frequent type of castlemans disease (90% of cases) [7]. This variant is usually asymptomatic or pausisymptomatic. In unicentric castlemans disease most common site is mediastinum[8]. The abdomen is rarely affected and only few cases have been reported. Clinical spectrum of castleman disease is variable, ranging from generalized lymphadenopathy to nonspecific complaints, this heterogeneity of various clinical features depends mainly on location and whether it is unicentric and multicentric type, microscopic appearance like hyaline vascular or plasma cell type[1].

Gross examination:

On gross examination it was single yellowish brownish incapsulated mass, total measuring 12.7 x 8.3 x 6 cm. On cut section there is presence of brownish well circumscribed firm mass measuring 5 x 5 x 4.5 cm.

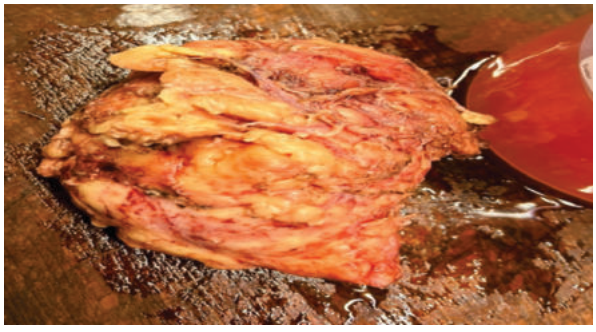

Figure 1 : Macroscopic findings : An encapsulated mass measuring 12.7 x 8.6 x 6 cm with yellowish brownish color.

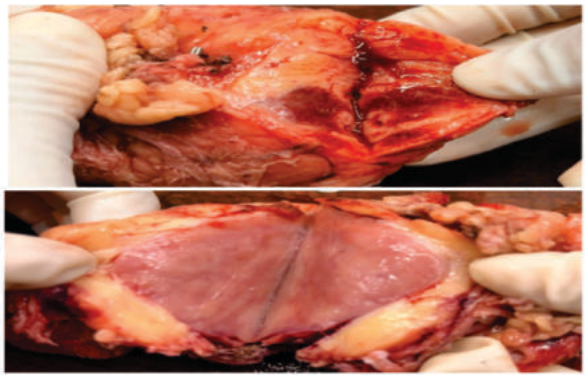

Figure 2: Cut section of para renal mass showing brownish well circumscribed firm mass.

Microscopic examination:
Microscopic examination showed diffusely arranged lymphoid follicles with germinal centers at variants stages of regression. It showed marked vascular proliferation and hyalinization of their abnormal germinal centers. Hyalinized germinal centers showed hyalinized areas, dendritic cells and a mantle zone forming onion skin arrangement. The interfollicular stroma is prominent with numerous hyperplastic vessels of post capillaries venules type along with admixture of plasma cells and immunoblasts. Dendritic cells are also seen.

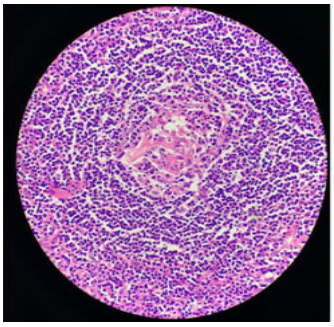

Figure 3: Lymphoid follicles shows features of onion skin appearance with interfollicular areas showed extensive hyaline vascular changes.

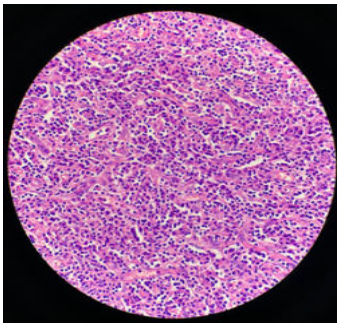

Figure 4: Interfollicular areas show extensive vascular proliferation

CONCLUSION:

Castleman disease is a real diagnostic challenge for all, for a clinician, radiologist and pathologists. As this disease comprise of a wide range of signs and symptoms and no leading signs on radiological imaging, all these features attribute to its difficulty in diagnosis. There is no any specific biochemistry test and blood investigations are currently present to help in diagnosis of Castlemans disease. A histopathological tissue diagnosis is definitely helps in diagnosing but routinely it is not used. Hyaline vascular type is most common than plasma cell type of castlemans disease and has a good prognosis. Surgery is the mainstay of treatment [1].

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