

A rare case of retroperitoneal castleman disease

Nithin Diwagar^{1*}, Sri Devi M², Padma Priya³, Rashmi R⁴

^{1,4}Postgraduate, ²Associate Professor, ³Tutor, Dept. of Pathology, Saveetha Medical College, Thandalam, Chennai Tamil Nadu, India

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Abstract

Castleman disease is a rare hyperplastic disorder that can affect any group of lymph nodes. Clinically and radiologically, it can mimic a malignancy and can only be diagnosed by histopathological examination. Castleman disease should be considered as one of the differential diagnosis for a patient presenting with a retroperitoneal mass. Complete surgical excision is usually curative; a preoperative diagnosis by biopsy can avoid extensive surgery for this condition. In this case report, we present a 40 year old female with a retroperitoneal mass, which was clinically suspected to be a malignancy. Complete surgical excision was done and histopathological examination of the mass revealed a unicentric Hyaline vascular type of Castleman disease.

Introduction

Castleman disease represents a heterogeneous group of diseases with a similar histological picture but with varied etiological factors, clinical presentation and prognoses. It is a hyperplastic condition rather than a neoplasm.¹ It is classified into two types based on the microscopic appearance or the clinical presentation. Microscopically, it can be of Hyaline vascular type or Plasma cell type. Clinically, it can be unicentric or multicentric, depending on the number of nodes involved. Hereby, we discuss the case of a 40 year old female presenting with a retroperitoneal mass to the General Surgery OPD.

Clinical History

A 40 year old female, not a known diabetic or hypertensive, came with complaints of pain in the upper abdomen for the past 20 days. The pain was intermittent and there was no history of vomiting, constipation, loss of weight or loss of appetite. On examination of abdomen, there was a mass palpable in the left lumbar region, measuring 10X8 cms. It was mobile and ballotable. CECT Abdomen showed a well-defined, predominantly retroperitoneal, solid lesion that was epicentered in the left anterior para renal space with multiple internal cystic areas, suggesting a neoplastic lesion. Laparotomy and excision of the lesion was done and the specimen was sent for histopathological examination.

Gross

We received a globular yellowish soft tissue measuring 7x6.5x6 cm. External surface was smooth and cut surface

was grey-white with multiple cysts, largest measuring 0.5cm in diameter. (Fig. 1)

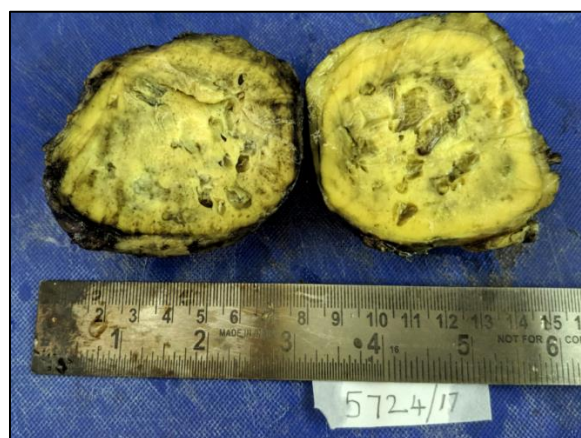


Fig. 1: Gross

Microscopy

Multiple sections studied showed a lesion composed of numerous lymphoid follicles with regressed germinal centers, which were mainly composed of follicular dendritic cells. Many follicles showed concentric rings of lymphocytes (onion skinning). Few follicle showed a single prominent sclerosed blood vessel traversing into the germinal center (lollipop follicle). The interfollicular areas showed obliteration of sinuses, increase in number of small blood vessels with hyalinized walls, few clusters of plasmacytoid dendritic cells, few plasma cells and immunoblasts. No evidence of necrosis.

*Corresponding Author: Nithin Diwagar, Postgraduate, Dept. of Pathology, Saveetha Medical College, Thandalam, Chennai Tamil Nadu, India

Email: drnithin27@gmail.com

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Based on the microscopic and clinical features, we diagnosed this case of retroperitoneal mass as Unicentric Hyaline vascular type of Castleman disease.

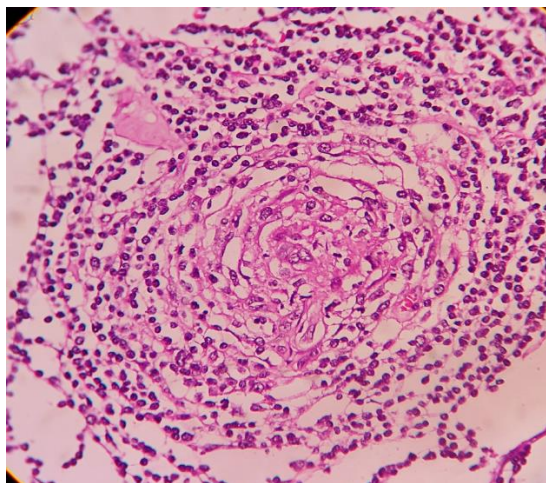


Fig. 2: H&E 40X- regressed germinal centers, which were mainly composed of follicular dendritic cells with surrounding concentric ring of lymphocytes

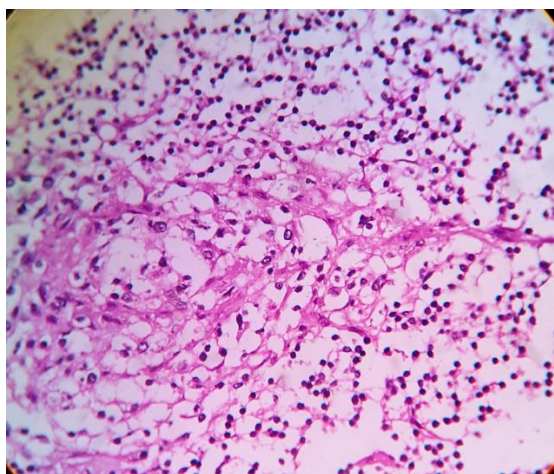


Fig. 3: H&E 10X - single prominent sclerosed blood vessel traversing in to the germinal center (lollipop follicle)

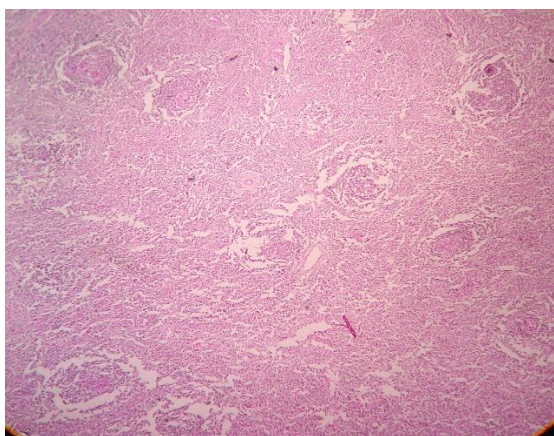


Fig. 4: H&E 4X – numerous follicles with regressed germinal centers

Discussion

Castleman disease does not represent a single disorder; rather it is a heterogeneous group of diseases with a similar histological picture and varied etiological factors, clinical presentation and prognoses. It is a hyperplastic condition rather than a neoplasm. It is also referred to by various other terms such as Angiofollicular lymph node hyperplasia, Giant lymph node hyperplasia, benign giant lymphoma and so on.

Castleman disease was named after Dr. Castleman,² who discovered and described it as a large, benign, asymptomatic mass involving the mediastinal node. In the subsequent years, it was classified into two types, based on either the microscopic appearance or the clinical presentation. Microscopically, it can be of Hyaline vascular type or Plasma cell type. Clinically, it can be unicentric or multicentric, depending on the number of nodes involved. Recently, HHV 8 has been recognized in a subset of multicentric cases. A specific cause for CD is still not known. However, HHV-8, also known as Kaposi sarcoma herpes virus, has been demonstrated in 50% of unicentric PC type cases and most of the multicentric forms.³ Much less is known about the etiopathogenesis of HV type of castleman disease. Patient may present with a mass involving a single node (unicentric form), or multiple nodes of different regions can be affected (multicentric form). The unicentric form can be either HV type or PC type histologically. Multicentric forms are almost always of plasma cell type.² Both males and females are affected equally.

HV type constitutes around 80 – 90% of the unicentric cases.^{2,4} It can occur over a broad age range, but it is usually seen in younger age groups. They are usually asymptomatic, often incidentally discovered during chest radiography. In some cases, symptoms are present due to compression of neighboring structures such as airway and major vessels.⁶ Systemic symptoms are rare in HV type. Mediastinal nodes are most commonly affected but it can affect any group of nodes. Complete surgical excision is curative.² In cases where complete excision is not possible, radiation therapy is used.⁷

PC type constitutes 10 – 20% of unicentric cases and nearly 100% of the multicentric cases.² It is usually seen in older age group. Mediastinal involvement is less frequent compared to HV type. They commonly present with systemic symptoms such as fever, night sweats, weight loss, malaise etc.⁸ Hepatomegaly and splenomegaly can be present. Multicentric CD may be associated with HIV and POEM's syndrome.⁹ Elevated erythrocyte sedimentation rate, anemia, thrombocytopenia, polyclonal hypergammaglobulinemia, and elevated serum levels of IL-6, LDH and C-reactive protein are commonly observed in MC castleman disease. Patients with multicentric CD are treated with chemotherapy and steroids.^{10,11}

Radiology and pre-operative FNAC are usually non diagnostic;¹² Histopathology remains the gold standard for diagnosis. Grossly, it presents as a firm mass involving any

lymph node chain, ranging in size from 2 – 20 cm.¹³ The cut surface is usually grey white with areas of calcification.

Histologic findings of HV type can be divided broadly into follicular changes and interfollicular changes. Follicular changes are characterized by numerous large follicles with regressed germinal centers composed of predominately follicular dendritic cells. Many follicles show concentric rings of mantle zone lymphocytes (known as onion skinning). A single prominent sclerosed blood vessel is usually seen traversing in to the germinal center in few follicles (known as lollipop follicle). Interfollicular changes are characterized by obliteration of sinuses, increase in number of high endothelial venules with hyalinized walls, clusters of plasmacytoid dendritic cells, plasma cells and immunoblasts but lower in count than PC type CD. PC type of castleman disease is characterized by large sheets of mature plasma cells with occasional binucleate forms in the interfollicular area. Lymph node sinuses are intact and distended, follicles are widely spaced and few follicles shows hyperplastic germinal center. Other features are usually similar to HV type. Immunostaining of HHV-8 is positive in majority of multicentric cases.

Prognosis of unicentric CD is good, rarely relapses can occur after surgical excision. Multicentric CD has a poor prognosis and they are prone to develop a variety of neoplasms of the reticulo-endothelial system.²

Conclusion

Castleman disease is a fairly benign condition but can mimic a malignant tumor clinically and radiologically. Preoperative FNAC and imaging studies is usually non diagnostic, Histopathological examination remains the gold standard for the diagnosis of CD. Complete surgical excision is curative for unicentric CD; a preoperative diagnosis by biopsy can be done to avoid extensive surgery.

Abbreviation

CD – Castleman disease, HV – Hyaline vascular type, PC – Plasma cell type, UC – Unicentric, MC – Multicentric.

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