

Case Report

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Suprarenal unicentric Castelman's disease: A rare case report**Mohammed Arafath Ali; Ramachandra C; Uday Karjol; Ravi Arjunan; Syed Altaf; Srinivas C; Akkamahadevi S Patil; Pavan Sugoora***

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Abstract

Castleman Disease (CD) is a rare non-clonal lymphoproliferative disorder. The clinical presentation of CD often overlaps with autoimmune, infectious, or other malignant diseases. The diagnosis is confirmed by a biopsy of the affected lymph-node tissue. A 39-year-old gentleman was evaluated for intractable abdominal pain fatigue of 2 months duration. CT scan showed a 4 X 4 X 5 cm right suprarenal mass. Laparotomy and near total excision was performed which showed reactive B cells expressing CD20 and PAX5, favoring CD. Patient received adjuvant 6 cycles of Rituximab and steroids. A follow up CT scan after 3 months showed a near complete resolution.

We herein report a rare case of retroperitoneal Castleman's disease.

Keywords: Castelman's disease; unicentric; multicentric; lymphoproliferative disease; rituximab.

Introduction

Castelman's disease (CD) was first documented and described by Dr. Benjamin Castelman as a rare lymphoproliferative disorder [1]. CD is a heterogeneous group of disorders and can present as single enlarged lymph node i.e. Unicentric or as a Multicentric lymphadenopathy affecting multiple lymph node stations.

Multicentric disease is usually associated with constitutional symptoms like fatigue, loss of weight, pyrexia, cytopenias and multiple organ dysfunction and is believed to be driven by IL-6 and many other cytokines [2]. However 50% of multicentric disease is caused by uncontrolled Human Herpes Virus – 8 (HHV-8) infection in immunocompromised patients whereas HHV-8 negative multicentric disease is considered idiopathic [3].

Unicentric disease presents as a solitary enlarged lymph node or as multiple enlarged lymph nodes in a single lymph node station [1]. The clinical course is generally indolent with slow and gradual enlargement of lymph nodes [4]. The average size of lymph node is larger in unicentric disease than multicentric disease [5].

Case presentation

A 39 year old gentleman presented to us with complaints of intractable abdominal pain, fatigue, lethargy and anorexia for 2 months. The patient had no comorbidities. Physical examination was unremarkable and on abdominal examination, mild tenderness was elicited in right hypochondrium. All the blood investigations and markers were in normal range. A CT scan showed a 3 X 4.2 X 5.2 cm homogeneously enhancing soft tissue mass in the right suprarenal location, in close relation to inferior vena cava (Figure 1-3).

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Laparotomy and near total excision of the lymph nodal mass was performed as image guided biopsy was technically challenging. Histopathological examination demonstrated lymph nodes with follicular hyperplasia admixed with numerous plasma cells and eosinophils, ateric lymphoid follicles with hyalinised blood vessels with nests of paraganglions showing the possibility of CD. Immunohistochemistry stains demonstrated reactive B cells expressing CD20 and PAX5 with absence of CD30 and LMPI expression, favouring CD (Figure 7-12).

A response assessment CT scan at the end of 6 cycles of rituximab and steroids showed near complete resolution (Figure 4-6) and a good clinical response.

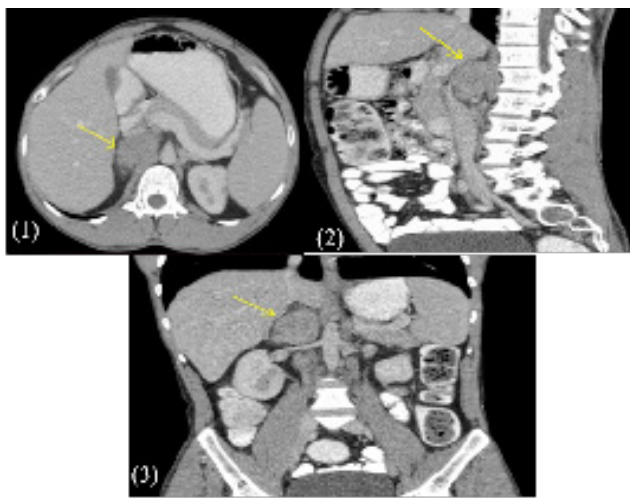


Figure 1-3: 3 X 4.2 X 5.2 cms homogenously enhancing soft tissue mass in right supra renal region at the level of L1, anteriorly displacing and compressing Inferior Vena Cava.

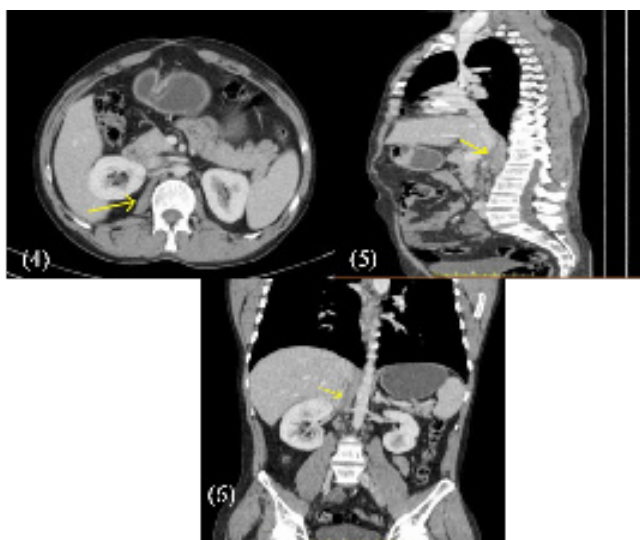


Figure 4-6: Near complete resolution of the mass.

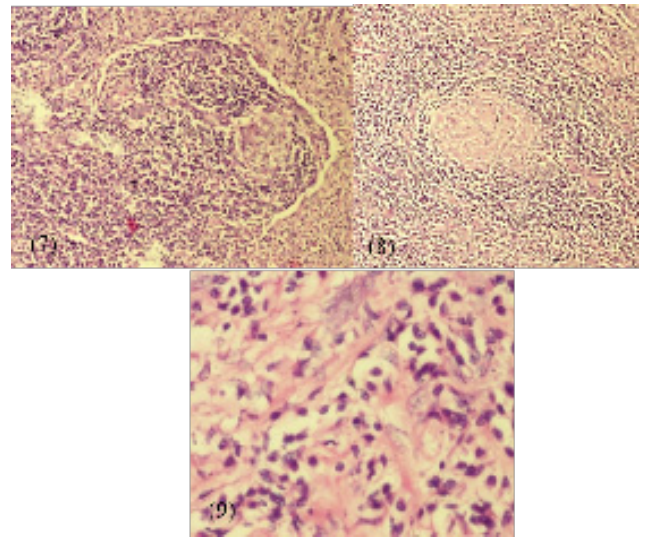


Figure 7-9: (7) Hyaline-vascular variant of Castleman disease hyaline vascular (lollipop) radially penetrated by a blood vessel with cracking artefact in the periphery of the follicle. (8) Hyaline-vascular variant of Castleman disease, the germinal centre is partially lymphocyte depleted and replaced by hyaline deposits and follicular dendritic cells. (9) Hyaline-vascular variant of Castleman disease, in the inter follicular region, blood vessels are lined by plump endothelial cells and are associated with small lymphocytes and plasma cells.

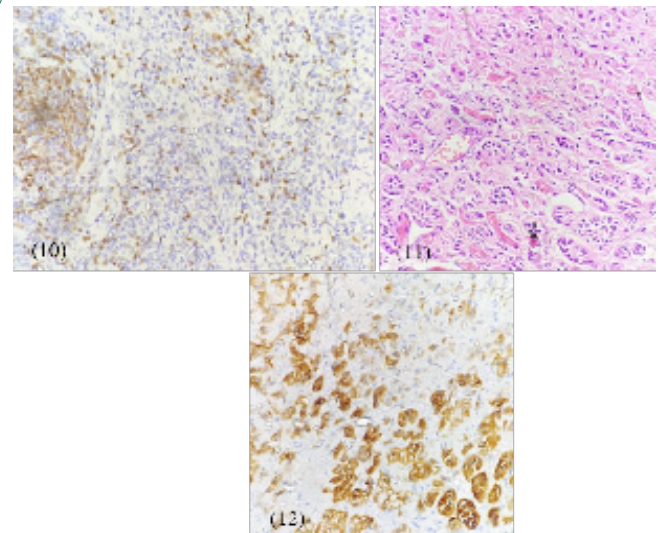


Figure 10-12: (10) PAX5 highlighting the pro- B cells in the marginal zone. (11) Cells arranged in nesting (Zellbalen pattern)with prominent vascular network. The cells are round to oval with abundant granular eosinophilic to basophilic granular cytoplasm. (12) PanCK showing membranous positivity in the paraganglion component.

Discussion

Unicentric disease is usually insidious in onset and involves a single lymph nodal station, it can be diagnosed at any age ranging from 2 – 84 years with a median age at diagnosis of 34 years. Unicentric disease has a slight female preponderance.

There are no known epidemiologic factors for the development of unicentric disease and are almost always HHV – 8 negative [6].

Due to the indolent course of unicentric disease its diagnosis is usually incidental as lymphadenopathy is often asymptomatic in nature, however the symptoms can arise if there is mass effect on adjacent structures from enlarged nodes for example on airways, ureters or adjacent neurovascular bundle.

Most common sites of involvement of unicentric CD are mediastinum (29%), neck (23%), abdomen (21%) and retroperitoneum (17%) and other uncommon sites like axilla, inguinal region, orbits, nasopharynx and small intestine [7].

Unicentric disease is divided into 3 subtypes based on histopathological features (i) Hyaline vascular subtype. (ii) Plasmacytic subtype. (iii) Mixed subtype which shows features of both. The hyaline vascular subtype is the most common subtype and is usually not associated with systemic symptoms.

However the patients with plasmacytic or mixed variant of unicentric CD may have iMCD like constitutional symptoms and deranged laboratory parameters like anemia, elevated ESR, or C reactive protein, hypoalbuminemia [8].

Management of unicentric disease is based on consideration of the following factors: (I) location of the enlarged lymph node, (II) resectability status of the involved lymph node or group of lymph nodes, (III) degree of compression of neighboring structures or systemic inflammatory syndrome.

Based on the above considerations the management differs.

- (i) Resectable UCD: The recommended first line treatment is surgical extirpation which eliminates any systemic symptomatology and laboratory abnormalities and is found to have high response rate [9]. Recurrence is uncommon after complete resection [10].
- (ii) Unresectable UCD: Generally encountered in the mediastinum adjacent to major blood vessels or bronchi. This category is further classified into:
 - (a) Asymptomatic unresectable UCD: As the UCD can remain stable or slowly grow over a long period of time it is best to wait and watch [11].
 - (b) Symptomatic unresectable UCD: Usually due to compression of adjacent vital structures and can be treated with partial resection in order to reduce the bulk of the disease without complete removal, it is associated with good results provided the patient is followed up to monitor the growth [10].
 - (c) Inflammation related unresectable UCD: It is driven by IL-6 and is rare in UCD; it is ideally treated with Rituximab as first line or Tocilizumab as second line before considering surgery [12].
 - (d) Persistent symptomatic unresectable UCD: Radiotherapy can be used as an alternative therapy in this subgroup with persistent symptoms and lymphadenopathy, which

persists despite treatment directed at relieving compression or inflammation related symptoms, another alternative approach is treatment with immunomodulators, immunosuppressant like corticosteroids, cyclosporine and sirolimus [13].

Follow-up of UCD is done 1 – 3 months after initial treatment and the response to the treatment given must be documented by history. Physical exam, laboratory values and imaging. Later UCD patients are followed annually with CT and laboratory studies. Annual imaging may be discontinued after 5 years.

Conclusion

UCD presents with a diagnostic dilemma as just lymphadenopathy may have a differential diagnosis of lymphoma or testicular tumors in males. Castlemans disease must be viewed as diagnosis of exclusion due to the rarity of the disease.

Complete surgical excision is the preferred intervention of UCD and the recurrence of the disease is rare and in patients with inflammation related symptoms IL-6 inhibitors like Rituximab along with steroids are feasible and effective options, as in our case due to difficulty in procurement of first line drugs.

Declarations

Funding: None

Conflict of interest: No potential conflict of interest relevant to this article

Ethics approval: Appropriate statements included

Consent to participate: All patients had consented for the proposed surgery. This is a retrospective analysis of prospectively maintained database which doesn't necessitate consenting for analysis

Consent for publications: All authors have consented for publication.

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