Castleman's Disease: a Suprarenal Surprise!

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Abstract

Castleman's disease is a distinct form of lymph node hyperplasia. It commonly presents as a mediastinal mass and rarely as a solitary retroperitoneal mass. We narrate a case of Castleman's disease presenting as a right suprarenal mass emphasising the usefulness of robot-assisted retroperitoneoscopy in surgical management of retroperitoneal masses in close relation to vital structures.

Keywords Castleman disease · Retroperitoneal mass · Robot assisted Retroperitoneoscopy · Adrenal mass

Introduction

Castleman's disease (CD) is a form of giant lymphoid hyperplasia seen in young to middle aged people. It commonly presents as a mediastinal mass and less frequently as an abdominal mass (11%) [1]. We report a case of Castleman's disease in a 44-year-old lady presenting as a right suprarenal mass which was managed by robot-assisted lateral retroperitoneoscopic approach.

Case Report

A 44-year-old lady presented with vague abdominal discomfort and was found to have a right suprarenal mass on ultrasonography. There was no history of hypertension, palpitation, sweating or flushing. On further evaluation with contrast-enhanced computed tomogram (CT), a well-defined lobulated right suprarenal mass measuring 5.5 × 3.3 × 6.7 cm with heterogenous enhancement, closely abutting the right renal artery and extending posterior to IVC (inferior vena cava), was seen. The adrenal gland could not be identified

separately (Fig. 1a). Physical examination showed no weight gain, proximal myopathy or abdominal striae. Laboratory investigations showed normal renal function and electrolytes. Endocrinology evaluation showed normal cortisol, 24-h urine fractionated metanephrines and dehydroepiandrosterone sulphate levels. In view of the size and enhancing nature, surgical excision of the lesion was planned. Patient underwent robot-assisted retroperitoneoscopic excision of the mass. Intra-operatively, a 6 × 6-cm right suprarenal mass was seen, with extension behind the IVC. The right renal artery was posterior and the renal vein anterior to the mass. Adrenal was identified separately and was dissected away from the mass (Fig. 1b). Histopathology was suggestive of hyaline vascular variant of CD (Fig. 2). Immunohistochemistry (IHC) confirmed the diagnosis (CD3/CD20/BCL2 showed reactive pattern in the follicles. CD138 highlighted the inter-follicular plasma cells). Complete haematology workup was done to rule out other sites of involvement. Peripheral smear, kappa and lambda chain values were normal. Positron emission tomography scan was done, which did not show any evidence of systemic disease, thus ruling out multicentricity. Patient has been followed up after 6 months, with no evidence of disease.

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Discussion

CD was first described in 1956. CD also known as giant lymph node hyperplasia represents a morphologically distinct form of lymph node hyperplasia. The exact aetiology is not known, but it has been found to be associated with human herpes virus-8. There are two morphologic types, the hyaline vascular type

Fig. 1 **a** CT image showing 5.5 × 3.3 × 6.7 cm mass (asterisk) abutting right renal artery (RA), posterior to IVC. **b** Intra-operative. Single asterisk (*) shows mass. Double asterisk (**) shows right RV. Number sign (#) shows right RA

which is common and the plasma cell type that has systemic manifestations [2]. From the clinical perspective, it can present as a solitary form or a multicentric form. The usual presentation is an asymptomatic solitary lymphadenopathy in the mediastinum or the abdomen. Few patients may have fever, night sweats, weight loss and fatigue. Our patient was incidentally found to have a suprarenal mass on evaluation for vague abdominal discomfort. Multicentric disease is a systemic lymphoproliferative disorder and the patients can present with fever, lymphadenopathy, hepatosplenomegaly, overexpression of interleukin-6 and polyclonal hypergammaglobulinemia. Multicentric Castleman is associated with HIV infection, Kaposi sarcoma, non-Hodgkin lymphoma, Hodgkin lymphoma and POEMS (polyneuropathy, organomegaly, endocrinopathy, M-proteins and skin lesions) syndrome [3]. To our knowledge, the world literature has described only seven cases of CD in the upper urinary tract [4].

Treatment of a solitary CD is surgical excision, with excellent long-term outcomes. In this patient, the tumour had a

retrocaval extension, and therefore, robot-assisted retroperitoneoscopic approach facilitated manipulation of main renal vessels and IVC, which would have been difficult transperitoneally (open or laparoscopic). In cases of multicentric disease, treatment options include chemotherapy, steroids, monoclonal antibodies to CD20 (rituximab), monoclonal antibodies (siltuximab) to IL-6, anti-IL-6-receptor antibodies (tocilizumab), antiviral agents (Ganciclovir) and interferon- α . The long-term prognosis of systemic CD is poor.

Conclusion

CD should be included in the differential diagnosis of any solitary, heterogeneous and hypervascular retroperitoneal mass [5]. Robotic retroperitoneoscopic surgery is a useful technique for dealing with retroperitoneal masses. On diagnosing CD on histopathology, patient should be thoroughly evaluated for systemic CD and appropriate treatment must be initiated.

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Fig. 2 Haematoxylin and eosin photomicrograph (×100) showing lymphoid follicles with regressed germinal centres. Inter-follicular areas with proliferated vessels