CASE REPORT

Retroperitoneal Castleman's disease mimicking soft tissue tumour

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Abstract

Castleman's disease is a type of non-neoplastic lymphoproliferative disease having lymph nodal hyperplasia. It has two distinct microscopic types: hyaline-vascular type and plasma cell type. Clinically, it may present either as a solitary mass, most commonly in the mediastinum, or as a multicentric form whose features are generalized lymphadenopathy, splenomegaly and involvement of other organs like the lungs and kidneys. Here we report a case of isolated retroperitoneal Castleman's disease, which presented as a lump in the iliac fossa in a young female. A clinico-radiological diagnosis of retroperitoneal soft tissue tumour was made and the patient underwent complete surgical excision. The exact diagnosis was only obtained at histopathology and there is no evidence of recurrence at six months follow-up.

Keywords Retroperitoneal · Castleman's disease · Isolated · Hyaline vascular

Introduction

Castleman's disease is a rare benign disease of lymphocytes of obscure aetiology which may present clinically as a solitary, asymptomatic mass or as multisystem involvement with accompanying fever, anaemia and hypoalbuminaemia. Most solitary masses occur in the mediastinum but have been reported in all parts of the body, including the retroperitoneum. Here we present a young female who presented with abdominal lump which mimicked a retroperitoneal soft tissue tumour, which after excision was found to be Castleman's disease.

Case Report

A 24-year-old, HIV negative female presented with a slowly growing, painful mass in the left lower abdomen of 6 months duration. There were no associated bowel or bladder disturbances. Her menstrual cycles were regular. On examination, there was a solid, firm, mobile, smooth surfaced, non-tender lump in the left iliac and hypogastric region. Ultrasonography of the abdomen revealed the lump to be a solid, highly vascular mass of size 7cms above and medial to the left psoas, without any lymphadenopathy or other abnormality. Contrast CT reported that the mass was a retro-peritoneal soft tissue tumour displacing the left ureter laterally, probably arising from the psoas. The patient underwent complete surgical excision of the tumour. During operation, the mass was found to be a greyish, smooth, highly vascular tumour behind the mesentery of sigmoid colon. Histopathology of the specimen revealed a diagnosis of hyaline vascular Castleman's disease. We found no clinico-radiological evidence of multicentric disease. At six months follow up, the patient has not shown any signs of recurrence.

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Fig. 1 CT scan (oral contrast) showing retroperitoneal tumour above left psoas.

Discussion

Castleman's disease is a morphologically distinct form of lymph node hyperplasia as compared to lymphoid malignancies. There are two microscopic categories – hyaline-vascular type and plasma cell type [1]. It is a disease mainly of the young occurring most commonly in mediastinum but it has been reported in the head [2] neck [3], pleura [4] and retroperitoneum [5]. The retroperitoneum is an uncommon site for Castleman's disease but it has been reported in this site where it mimics a soft tissue tumour, both clinically and radiologically [6, 7] and give rise to diagnostic confusion. There may be two types of clinical presentations, a solitary form without systemic manifestations and a multicentric form which is often associated with HIV infection and accompanied by anaemia and hypergammaglobulinaemia. The mass is characteristically hypervascular and may occasionally show an arborizing calcification pattern which is useful for diagnosis [8]. The localized form is amenable to surgical resection while the disseminated form generally needs systemic therapy like corticosteroids [9] or cytotoxic therapy [10]. Our patient had a solitary form of Castleman's disease without any evidence of systemic spread.

Because of the rarity of the condition, it was not entertained in the differential diagnosis during clinical examination or imaging studies, and the diagnosis was only obtained on histopathology. However, in a patient with retroperitoneal solid tumour which is hyper-vascular, Castleman's disease should be kept as a possible diagnosis, specially if the patient is also known to have concurrent HIV infection.

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