Retroperitoneal Pararenal Castleman’s Disease
OP Michail1, P Tsirkinidis2, A Androulaki3, C Georgiou1, M Angelopoulou2, J Griniatsos1

ABSTRACT
A 51-year old male patient with a three-month history of constant and dull left flank pain was investigated by ultrasonography, computed tomography (CT) scan and magnetic resonance imaging (MRI) of the abdomen, which disclosed a 8 x 7 x 6 cm retroperitoneal pararenal mass with heterogeneous imaging characteristics and bright enhancement following intravenous contrast injection. Based on the hypervascularity of the mass and the lack of specific signs in the imaging investigation, lymphoma, sarcoma or vascular tumour were considered as probable diagnoses and the patient underwent an exploratory laparotomy. The histologic examination of the surgically resected specimen disclosed “a hyaline type of Castleman’s disease”. Further evaluation of the patient with antibody testing for HIV 1 and 2, as well as viral load by PCR for Herpes Virus-8 (HHV-8) were negative. Bone marrow aspiration, biopsy and immunophenotypic study did not disclose any evidence of lymphoma. Molecular study of the bone marrow for immunoglobulin heavy chain rearrangement showed a polyclonal pattern; serum protein electrophoresis did not show any evidence of hypergammaglobulinaemia and serum immunofixation electrophoresis did not show any monoclonal protein. A diagnosis of localized – unicentric type of Castleman’s disease was made.

Castleman’s Disease should be included in the differential diagnosis of any solitary, heterogeneous and hypervascular retroperitoneal mass. Discovery of Castleman’s disease at any area of the body should be followed by a thorough imaging and laboratory work-up in order to exclude the multicentric type of the disease and the co-existence of lymphoma.

Enfermedad de Castleman de Localización Retroperitoneal Pararenal
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RESUMEN
Un paciente varón de 51 años con una historia de tres meses de dolor constante y sordo en el costado izquierdo, fue sometido a investigación mediante ultrasonografía, tomografía axial computarizada (TAC) e imagen por resonancia magnética (IRM) del abdomen. La investigación reveló una masa retroperitoneal pararenal de 8 x 7 x 6 cm, con imagen de características heterogéneas y aumento de la luminosidad tras la inyección intravenosa de contraste. Sobre la base de hipervascularidad de la masa y la falta de signos específicos en la investigación por imágenes, el linfoma, el sarcoma o el tumor vascular fueron considerados como diagnósticos probables y el paciente fue sometido a una laparotomía exploratoria. El examen histológico del espécimen resecado quirúrgicamente reveló “un tipo hialino de la enfermedad de Castleman.” La evaluación ulterior del paciente con prueba de anticuerpos de VIH 1 y 2, así como la carga viral por PCR para la detección del virus herpes humano tipo 8, dio resultados negativos. La aspiración de médula ósea, la biopsia y el estudio inmunofenotípico no mostraron ninguna evidencia de linfoma. El estudio molecular de la médula ósea para el reordenamiento de las cadeNA pesada de inmunoglobulina mostró un patrón policlonal. La electroforesis de la proteína en suero no mostró evidencia alguna de hipergammaglobulinemia y la
INTRODUCTION

In 1956, Benjamin Castleman introduced the term angiofollicular lymph node hyperplasia which described a rare atypical lymphoproliferative disorder. Clinically, the disease is classified in the localized – unicentric (LCD) and the generalized – multicentric (MCD) type, due to major differences in pathogenesis, clinical presentation, treatment and prognosis (1, 2). Histopathologically, the disease is divided into the hyaline vascular type which is more frequent in the LCD type and the plasma cell type which is more common in the MCD type (1). The localized type usually presents as an asymptomatic solitary mass whereas the multicentric type frequently shows generalized symptoms, organomegaly and lymphadenopathy and is mostly located in the peripheral lymph nodes (1, 2).

The LCD type predominantly arises in the mediastinum or the lung hilum (60–75%) and the neck (20%). Development in the retroperitoneum accounts for 10% of the reported cases while development in extremely rare locations, such as the parotid gland, has also been reported (3–6%) (3). In a recent review (4), a total of 122 retroperitoneal CD cases were reported and among them, 24 cases (20%) were located in the pararenal region. In this report, we describe a relatively rare case of LCD development in the pararenal region.

CASE REPORT

A 51-year old male patient presented with a three-month history of constant and dull left flank pain. He was initially evaluated by an internist. No abnormal clinical findings were recorded. He underwent ultrasonography, computed tomography (CT) scan and magnetic resonance imaging (MRI) of the abdomen which disclosed a 8 x 7 x 6 cm retroperitoneal pararenal mass with heterogeneous imaging characteristics and bright enhancement following intravenous contrast injection (Fig. 1. a–c).

Based on the above imaging findings, the patient was referred for further evaluation and treatment. The patient was completely asymptomatic except for the left flank pain. His previous medical history was significant for fatty liver with intermittent elevated transaminases. Two years before presentation to us, he was subjected to a CT scan of the abdomen which disclosed a 2 x 3 x 2 cm retroperitoneal mass was found then, for which no further evaluation was undertaken. His physical examination was completely normal with no lymph node enlargement or organomegaly. His complete blood counts and blood chemistries were within normal limits apart from slightly elevated transaminases while erythrocyte sedimentation rate (ESR) was normal. For staging purpose, a cervical and thoracic CT scan was performed but did not disclose any enlarged lymph nodes or any other abnormal findings.

Since no other involved site was found, the patient was subjected to an exploratory laparotomy through a standard midline incision. At laparotomy, an encapsulated, elliptical yellow tumour with diffuse and abundant vascularity was found in the left pararenal region. Dissection of the tumour was difficult due to thick vascular fibrous adhesions between the capsule of the tumour and the adjacent retroperitoneal structures. The surgically resected specimen measured 6.5 x 5.5 x 5 cm and weighed 180 g (Fig. 1d) The patient had an uneventful postoperative course and was discharged on the seventh postoperative day.

The histologic examination of the surgically resected specimen disclosed “a hyaline type of CD” (Fig. 2). In detail, the tumour represented lymph node tissue with follicles that consisted of broad mantle zones surrounding small germinal centres with follicular dendritic cells, giving a characteristic “onion peel” appearance. Follicles were penetrated by hyalinized venules. Interfollicular areas contained a large num-

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