

Retroperitoneal Castleman's Disease in the Perinephric Space—Imaging Appearance: A Case Report and a Review of the Literature

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Abstract

Introduction: Castleman's disease (CD) is a rare lymphoid tumour usually found in the mediastinum. Extrathoracic sites are uncommon. Its radiological findings may be similar to other retroperitoneal tumours, making diagnosis difficult. **Clinical Picture:** A 54-year-old female was found to have an incidental hypoechoic mass in the left posterior perinephric space on routine ultrasound. Abdominal computed tomography (CT) scan demonstrated an isodense mass which enhanced brightly with intravenous contrast. Angiogram confirmed a hypervascular mass. **Treatment:** The retroperitoneal mass was excised. **Outcome:** Histology revealed CD of hyaline-vascular type. **Conclusion:** CD should be considered in the differential diagnosis of a retroperitoneal mass, which demonstrates homogeneous and intense enhancement.

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Introduction

Castleman's disease (CD) was first proposed by Castleman in 1954.¹ It is usually found in the mediastinum, but other sites have been described.^{2,3} It is classified into two histological subtypes, the hyaline-vascular (HV) and plasma-cell (PC) types.⁴ It can appear as a solitary localised disease or as multifocal form.⁵ The radiological characteristics may be similar to other retroperitoneal tumours making diagnosis difficult. To our knowledge, CD presenting in the perinephric space is rare, as only a handful of cases have been described in the literature.³ We report a case of retroperitoneal CD presenting in the perirenal region and discuss its radiological features that may be useful in the diagnosis of the disease.

Case Report

A 54-year-old female with dyspepsia of a few months was referred to our hospital for evaluation and treatment. Gastroscopy revealed gastric heterotopia. However, routine duplex ultrasound showed an incidental hypoechoic mass with increased vascularity adjacent to the left kidney (Fig. 1). A non-enhanced computed tomography (CT) scan of the abdomen demonstrated an isodense mass in the left posterior perinephric space, which enhanced brightly with

intravenous contrast (Figs. 2a & 2b). A few small foci of low-attenuation areas, which showed no significant enhancement, were noted within the mass. No calcified foci were noted. Angiogram showed a hypervascular mass, supplied by the left L3 lumbar artery and the capsular branch of the left renal artery (Figs. 3a & 3b). A dense blush was also demonstrated in the capillary and venous phase. Subsequent CT thorax showed no tumour metastasis or other areas of involvement. Urea and electrolytes were normal. Full blood count was also obtained which showed normal red and white cells parameters. Excision of the mass was made and a tan-coloured fleshy tumour, 4.5 cm in greatest diameter, with fibrin adhesion was excised from the retroperitoneum. Histology of the mass revealed CD of HV type.

Discussion

CD was first described in 1954 by Castleman and Towne as soft tissue masses in the mediastinum, clinically mimicking thymomas. These were thought to be hyperplastic lymphoid tumours, believed to be the result of chronic, low-grade, non-specific inflammation.^{1,6} Histologically, this was described as localised hyperplastic lymphoid process, characterised by Hassal body-like

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Fig. 1. Transverse abdominal sonogram. This shows a round homogeneous left posterior perinephric space mass with uniform hypoechogenicity.

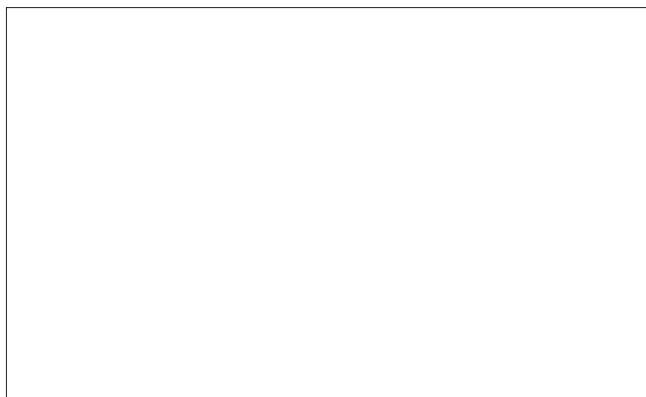


Fig. 2b. Contrast-enhanced scan. The mass reveals homogeneous enhancement reflecting the hypervascular nature of the lesion. A few low attenuation foci are also present. These may represent areas of fibrosis.

germinal centres with marked vascular proliferation. Others, who suggested a hamartomatous origin, later challenged Castleman's inflammatory response theory.⁷ Since the original description, many synonyms, such as benign lymphoma, follicular lymphoreticuloma, lymphoid nodal hamartoma,⁷ angiomatous lymphoid hamartoma,² and angiofollicular lymph node hyperplasia, have been used to describe this disease, reflecting the uncertain aetiology. In 1969, a new morphological type (plasma cell type) characterised by sheets of mature plasma cells associated with large hyperplastic follicles was described by Flendrig.⁸ In 1972, Keller et al⁴ proposed two histologic subtypes, the HV and PC types. During the last decade, numerous reports of an aggressive multicentric form of CD with clinical similarities to other lympho-proliferative disorders have resulted in the clinical distinction between localised and multicentric CD.⁵ Localised CD appears mostly in young patients. The patients' age at diagnosis ranges from 2 to 85 years with a median age of 28 years.^{4,5,9} There appears to be no racial or sexual predominance.⁴ The commonest site of CD is the mediastinum (in 70% of cases) with another 20% occurring in many other sites, including

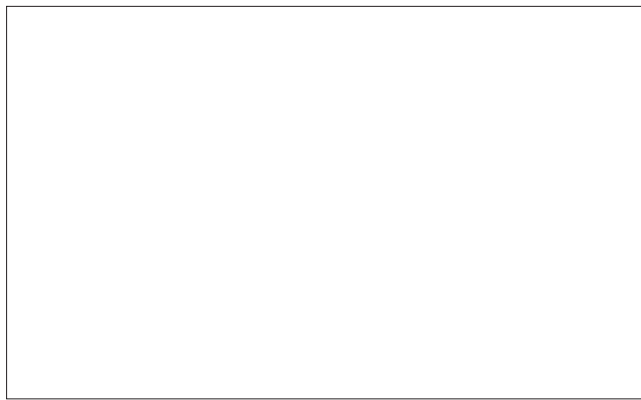


Fig. 2. Computed tomographic images.

Fig. 2a. Plain scan. A well-defined homogeneous mass is demonstrated in the left perinephric space posteriorly.

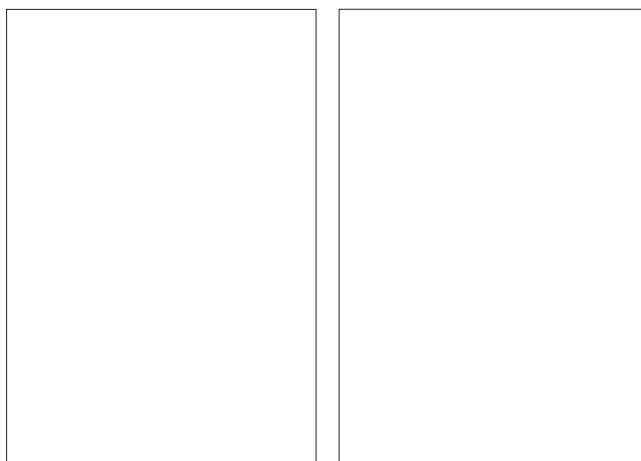


Fig. 3a. Aortogram.

Fig. 3b. Selective left renal angiogram. The tumour is hypervascular, being supplied by the lumbar artery and the capsular branch of the left renal artery. It shows capillary blush and persistent contrast staining.

superficial nodal groups such as the cervical, axillary, inguinal, shoulder and vulvar regions. Less commonly, this neoplasm is found in intraperitoneal and retroperitoneal locations.^{2,9} In a review of 315 cases of CD, 21 (7%) retroperitoneal tumours were described, with 6 (2%) having a pararenal location.³

Macroscopically, localised masses of CD are homogeneous, capsulated, dark yellow and often attached to the neighbouring structures.⁹ The masses are highly vascular, so dissection may produce considerable bleeding.¹⁰ The HV type constitutes 90% of cases of CD and usually presents as a large solitary mass without signs or symptoms. Symptoms occur in only 3% of the cases, usually due to compression of adjacent structures. The PC type appears in 10% of localised CD.⁴ It is symptomatic in 50% of cases.¹¹ Most of patients affected with the PC type have constitutional symptoms and haematological abnormalities, such as fever, sweats, fatigue, anaemia, elevated erythrocyte sedimentation rate, polyclonal hypergammaglobulinaemia,

and bone marrow plasmacytosis.⁵ The treatment for localised CD is surgical resection, which gives good results with the disappearance of related symptoms.¹²

More recently, CD has been reported as a systemic and multifocal disease. It occurs in elderly patients with a median age incidence of 57 years. This form usually is the PC subtype.⁵ Generalised lymphadenopathy is common, usually peripheral in location and involves multiple sites. Most patients with multifocal CD have constitutional symptoms and haematological abnormalities. Approximately 30% of them also predispose to the development of malignancy. POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, m protein and skin changes) or amyloidosis may be associated, and Kaposi's sarcoma, non-Hodgkin's lymphoma and monoclonal gammopathies may appear.¹¹ The course of the disease may be chronic, with recurrent exacerbations and remissions, or it may have an aggressive course with a poor prognosis and often-fatal clinical outcome despite treatment with corticosteroid and chemotherapy.⁵

A form with intermediate morphological features, suggesting a spectrum of abnormalities linking the varying histological subtypes, was also described and at times, both types are found in the same patient in separate places or in successive examinations.⁵

Retroperitoneal CD is radiographically not specific and indistinguishable from malignant disease. An intravenous pyelogram may demonstrate an abnormal renal axis or ureteral deviation with or without extrinsic obstruction from the retroperitoneal mass. Ultrasonography usually shows a homogeneous low-echo mass.^{13,14} Typically, the HV type appears on CT as a homogeneous mass, which enhances intensely after intravenous contrast medium. Punctate or arborising calcification may be seen.¹⁵ Although the mediastinum tumours are not commonly calcified, dense calcification has been reported in almost a third of cases of abdominal CD, more frequently with retroperitoneal CD.⁹ Radial or star-shaped calcification is said to be characteristic of CD. Calcification may be present in some retroperitoneal masses, such as fibrous malignant histiocytoma, smooth muscle tumours, neurogenic tumours and others. However, star-shaped calcification is seldom found in these neoplasms.¹¹ Three-phase dynamic CT is useful for differentiating fibrotic lesions. Fibrous tissue usually shows a very slow enhancement pattern, with little or no enhancement in the early (arterial) phase and more intense enhancement in the delayed phase.¹⁶ The non-enhancement on contrast-enhanced CT in our patient is most likely a result of timing related to the phase of the enhancement in the fibrotic area of the lesion. Keller et al examined 74 cases of CD and reported that fibrosis was not infrequently seen particularly around central vessels in the

HV type. Central fibrosis has now been recognised as an important feature of HVCD.^{4,17} Calcification sometimes occurs within the fibrous tissue.^{4,15}

The MRI findings are also non-specific, with low signal intensity on T1 weighted images and high on T2 weighted images.^{9,15} Recent reports have described hypointense streaks or stellate areas within HVCD masses on T2 weighted images attributed in these cases to fibrosis, calcification or vessels.^{15,17} The characteristic enhancement pattern seen in three-phase dynamic CT is also expected on MRI.

On angiography, the tumour appears hypervascular with dilated feeding arteries and showed capillary blush.^{3,11,18} In some cases, early venous return is noted.¹⁸ Hence, a solid and hypervascular tumour with capillary blush should include Castleman's tumour in the differential diagnosis under the appropriate clinical setting. Prominent vascular proliferation is the reason for the malignant appearance of this benign tumour. Encasement or amputation of vessels has been described. However, vascular encasement of the tumour should raise the suspicion of sarcoma. Nevertheless, there are some differences between CD and retroperitoneal sarcomas. A retroperitoneal sarcoma is generally a large tumour with necrosis that usually occurs in elderly patients. A sarcoma has heterogeneous high signal intensity on T2 weighted MRI.¹⁴

Radionuclide appearance of CD is rarely reported in the literature. It appears to be non-specific. A case report has described focal increased uptake of technetium 99m hexakis-2-methoxyisobutylisonitrile (99mTc-MIBI) in a patient with CD located in the mediastinum.¹⁹

Preoperative diagnosis of CD is difficult. However, CD should be considered in all retroperitoneal or mesenteric masses, especially when accompanied by anaemia and hypergammaglobulinaemia. Retroperitoneal tumours include various types of benign and malignant neoplasms. Benign retroperitoneal tumours are uncommon, comprising about 20% of all primary retroperitoneal neoplasms. The differential diagnosis includes lipomas, leiomyomas, lymphangiomas, xanthogranulomas, ganglioneuromas, neurofibromas, extra-adrenal pheochromocytomas, retroperitoneal cysts and urogenital ridge remnants.^{2,9} When located around the pancreas, it may be confused with a pancreatic neoplasm.²⁰

Surgical removal is considered the best treatment for the localised CD. Symptoms related to compression of adjacent structures or the systemic malaise seen with the plasma cell variants resolve after surgery. Even with partial excision of unresectable lesions, recurrence or progression of localised disease is not expected.⁹

Multifocal CD has a poor prognosis. Corticosteroids and

chemotherapy such as cyclophosphamide and azathioprine may improve the outcome, but total remission is difficult to achieve.^{4,5}

Conclusion

CD is a rare lymphoid neoplasm occasionally presenting as a retroperitoneal mass. This neoplasm is radiographically and operatively indistinguishable from other retroperitoneal malignancy.

CD should be included in the differential diagnosis of a retroperitoneal mass, which shows homogeneous and intense enhancement, especially when there is associated punctate or star-shaped calcification and central fibrosis.

Localised retroperitoneal neoplasms are indolent and surgical excision including partial resection is usually curative. Multicentric disease with retroperitoneal involvement usually present as an aggressive disease, often with poor prognosis, and systemic therapy is usually indicated.

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