

Case Report

Retroperitoneal Castleman Disease with paravertebral localization mimicking nephrolithiasis: A rare case

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Abstract. Castleman's Disease (CD), is a rare condition of uncertain etiology, involves a massive proliferation of lymphoid tissues and typically presents as mediastinal masses. In general, CD is an incidentally diagnosed disease with an asymptomatic course in most cases. We aimed to present a 48-year old woman who presented with left flank pain and had calcifications at the localization of left kidney on abdomen x-ray. The retroperitoneal mass with paravertebral localization was evaluated, because the size of mass in our case was larger than traditional sizes seen in Castleman Disease and calcifications and localization of the mass on KUB x-ray can mimic renal stone disease.

Key words: Castleman disease, retroperitoneal mass, nephrolithiasis

1. Introduction

Castleman Disease (CD) was first described by Benjamin Castleman in 1954. It is an extremely rare disorder with unknown etiology and incidence, which progress with lymph node hyperplasia (1, 2, 3). Castleman Disease can occur at any site in lymphatic chain. It is mostly seen at mediastinum (70%). It was also reported at regions of neck, axilla, pelvis and retroperitoneum.

Localized Castleman Disease is usually asymptomatic. Here, we presented a case with giant, localized, hyaline-vascular type Castleman Disease which mimicked nephrolithiasis by referred symptoms typical macroscopic calcification observed on KUB x-ray and symptoms.

2. Case report

As calcifications were detected on the abdominal x-ray in 48-year old woman patient with flank pain at the left side, further imaging studies were performed, which revealed a solitary mass at left retroperitoneum (Figure 1). In laboratory evaluations, complete blood count, liver and renal



Fig. 1. KUB x-ray: Macroscopic calcification compatible with the location of left kidney.

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function tests, acute phase reactants and urine analysis were within normal range. Serologic markers including HIV and HHV-8 were found to be negative. On computerized tomography (CT) scan, an ill-defined soft tissue mass (10x6.5x6.5 cm in size) with lobulated contours and left paravertebral localization within mesenteric fat tissue. The mass which displayed a popcorn calcification pattern and extended from the level of renal vein to 3th lumbar vertebral body L3 level cranio-caudally was observed. Left renal vein was shifted to the left secondary to mass and surrounding mesenteric fatty tissue planes were heterogeneous (Figure 2). Non-malignant lymphoid tissues were detected in the ultrasound-guided biopsy.

The patient underwent surgery via subcostal incision. During surgery, it was seen that the mass was adhered to vertebral column and aorta at left paravertebral area. The mass was successfully released from surrounding tissue and removed completely. The mass extracted to in size and in weight 16x10x16 cm, 343 g. (Figure 3). The histopathological evaluation of the specimen was reported as hyaline-vascular type Castleman Disease (Figure 4). No complication occurred after surgery. Also, no recurrence was observed in the following six month. Patient is still being considered under follow-up of our outpatient clinic.

3. Discussion

20% of all primary retroperitoneal masses are benign (4). Castleman Disease, which can also cause a benign retroperitoneal mass, is observed in mediastinum in 70% of the cases. While superficial nodal groups comprise 20%, retroperitoneal and pararenal localizations comprise 7% and 2% of the cases, respectively (4).

Although it is most frequently seen in early decades of life, it can be seen between 8 and 65 years of age and no sex predominance is present (5). Although several immunological mechanisms including infection with herpes virus type 8 and excessive production of interleukin 6 have been proposed, the etiology of the disease has not been fully elucidated (6).

There are two histological types of the disease (7). The most frequent type is the hyaline-vascular type (85%-90% of the cases), which is characterized by abnormal lymphoid follicles, numerous vessels, and wide fibrous septa. The disease is usually asymptomatic. The plasma-cell type is less frequent (10%-15% of the cases); and characterized by large follicles with intervening sheets of plasma cells and few vessels.

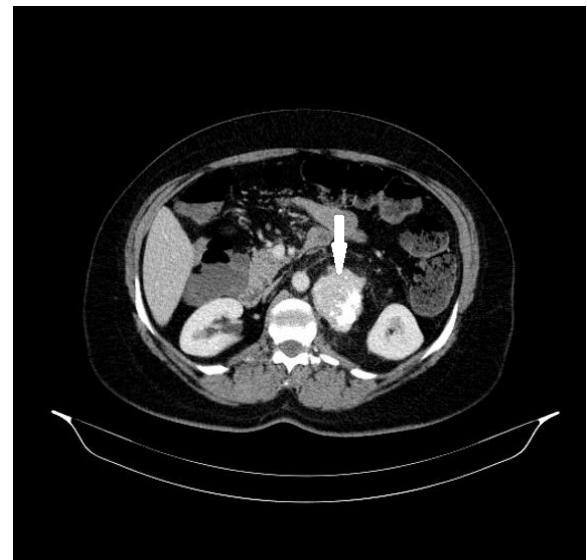


Fig. 2. CT scan: An ill-defined soft tissue mass with lobulated contours and left paravertebral localization which displayed popcorn-like macro-calcification.

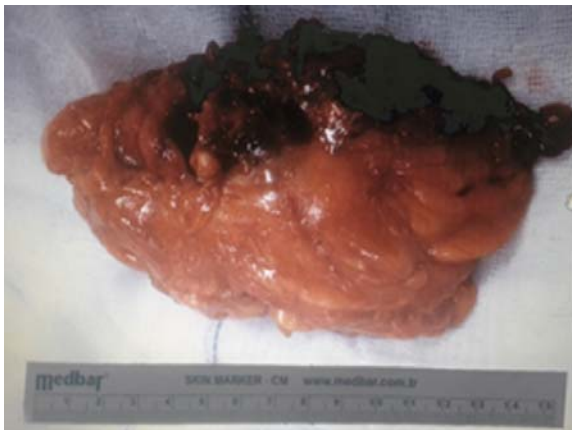


Fig. 3. The encapsulated mass lesion was 16x10x6 cm in size and 343 g in weight.

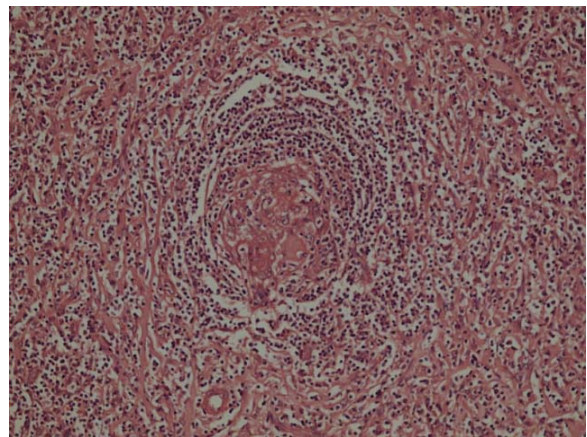


Fig. 4. Histopathology: Castleman Disease, hyaline-vascular variant; presence of hyaline deposit in the germinal center (H&E x200).

Localized form of Castleman Disease is unilateral and lesion size varies from 1 to 12 cm. Almost half of the patients are asymptomatic and unrelated to HHV-8. They are generally detected during routine physical examination, chest radiography or abdominal sonography. Symptoms, if any, include sensation of fullness at thorax and abdomen, low-grade fever, anemia, fatigue, weight loss, excessive sweating and skin eruptions (5,8,9,10,11). As localized mass lesions of Castleman Disease normally have no symptom, we assumed that left flank pain in our case resulted due to the excessive volume of the mass. The resected mass was 343 g in weight and 16x10x6 cm in size. It was rather large when compared to reported cases so far.

Although imaging techniques such as sonography, CT or magnetic resonance (MR) imaging are used for diagnostic purposes, these techniques do not provide definitive diagnosis as there is no specific marker for Castleman Disease, but they provide critical information regarding the localization of tumor (12). On CT scan, Castleman Disease can manifest as a mass lesion with homogenous or heterogeneous density relative to soft tissue (3). Central calcification is the typical finding in Castleman Disease and it is usually coarse in nature (8). In our case, there was coarse calcification on abdomen x-ray, suggesting nephrolithiasis due to its localization.

In the management of localized Castleman Disease, surgical resection is associated with good outcomes in the long-term, but radiotherapy is an effective therapeutic option that should be considered in cases in which complete resection is not feasible or surgical treatment is not appropriate (6). The prognosis and treatment of the multicentric type depends on its subtypes, while patients with multicentric CD and those presenting with systemic symptoms tend to have a worse prognosis (13). Currently, humanized anti-interleukin-6 receptor antibody has been proved to be an effective treatment for multicentric CD patients (14). Our case was unicentric, hyaline-vascular type and an appropriate candidate for surgery. No radiotherapy was given as complete resection could be achieved.

Malign transformation has been reported in both multicentric and unicentric type CD. Patients with CD should be regularly followed-up in order to early detection of malign lesions (15). In our case, no recurrence or malign transformation was detected in the short-term follow-up.

In conclusion, the Castleman Disease should be considered in the differential diagnosis of the cases with calcification in abdomen x-ray and that these cases should be properly managed by making accurate diagnosis with advanced imaging modalities.

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