Castleman's Disease: A Rare Mass in the Pararenal Retroperitoneum that Mimics Other Tumors

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Castleman's disease (CD) is a rare lymphoproliferative syndrome typically involving a mediastinal mass. CD in the pararenal area should be distinguished from lymphoma diseases. We analyzed the clinical features of ten CD patients in the pararenal retroperitoneum that mimics other tumors to summarize some of our experiences and conduct a literature review.

Ten patients with pararenal CD who were admitted to Peking Union Medical College Hospital from August 2005 to July 2015 were retrospectively analyzed. Seven patients (7/10) were without any symptom. One (1/10) had a medical history of hypertension for the past 10 years. All patients' urinary catecholamine excretion, plasma renin activity, and plasma aldosterone concentration were within the normal range. Computed tomography (CT) imaging confirmed the presence of a solid, homogeneous, hypervascular, and well-delineated mass in all ten patients that showed enhancement with intravenous contrast [Figure 1]. Two cases were taken magnetic resonance imaging (MRI) before the operation. Six patients (6/10) were diagnosed with pheochromocytoma/paraganglioma. The longest diameter of the involved masses ranged from 3.5 to 9.4 cm. The lesions were located in the adrenal region (seven cases), renal hilum region (two cases), and the inferior pole of the kidney (one case). Six patients (6/10) were examined by 99mTc-octreotide scanning. ^{18}F-fluorodeoxyglucose positron emission tomography/CT was applied in two patients. Five patients (5/10) underwent iodine-131-metaiodobenzylguanidine scintigraphy. Follow-up visits were conducted via telephone or outpatient examination.

Nine patients (9/10) underwent surgical resection of the tumors, and 1 (1/10) underwent CHOP chemotherapy with a biopsy operation. Seven patients underwent mass excision through a retroperitoneal laparoscopic approach. Two tumors were removed by open laparotomy. The diagnosis of CD was confirmed in all patients by histopathological examination. All cases were of the hyaline-vascular (HV) type. Six patients (6/10) were misdiagnosed before the operation. The longest follow-up period was 3 years, and no recurrence was found.

CD is a rare, heterogeneous lymphoproliferative syndrome. CD mainly occurs in young people between 15 and 35 years old, and there are no gender differences in the incidence of CD.[1] The etiology of CD has not yet been identified, IL-6 and human herpes virus-8 may be associated factors. CD can be classified into two clinical types: unicentric CD (UCD) and multicentric CD (MCD). The subtypes, such as HV, plasma cell (PC), and mixed type (MV), are classified according to their histology. UCD is usually associated with the HV histological subtype, whereas MCD is often associated with the PC subtype or MV subtype.

CD can arise wherever lymph nodes are found, but the most involved site is the chest, particularly in the retroperitoneum,[2] as well as other areas such as the suprarenal area and lateral

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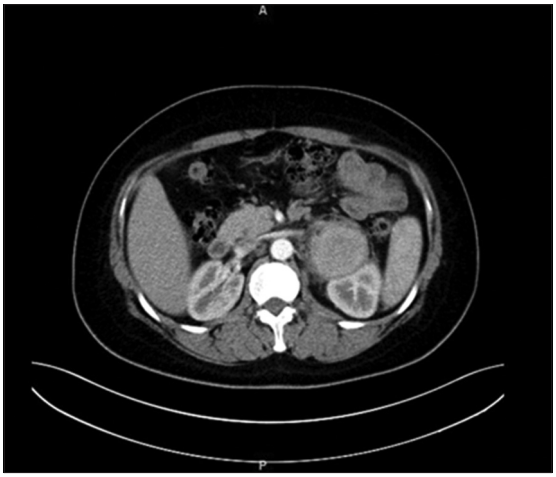

Figure 1: A computed tomography scan showed a big mass located in the anterior part of the left kidney.

to the kidney or abdominal or cervical lymphadenitis.[3] A relevant literature review in the PubMed database suggests that CD localized in the suprarenal area is rare and of ten mimics an adrenal tumor, as only a small number of cases have been reported.[2] In fact, CD is easily misdiagnosed as an adrenal neoplasm; thus, differential diagnosis is important due to the different treatments and prognosis for these different conditions.

CD or pararenal masses, such as pheochromocytoma or paraganglioma, can be detected with a physical examination. Pheochromocytoma or paraganglioma can cause hypertension due to renin secretion. Most UCD patients are asymptomatic or only experience symptoms due to local compression, such as lumbar or abdominal pain or vomiting. However, MCD is always associated with a systemic such as fever, weight loss, skin rash, anemia, and hypoalbuminemia. In this study, most of the patients were asymptomatic, and abdominal pain was the second most detected symptom. This is in agreement with the literature reports.

In CT imaging, an adrenal mass or lymphoma can be easily misdiagnosed as CD. MRI shows a solid mass that is slightly increased on T1-images compared with muscle and hyperintense on T2-images. Intralesional flow voids on T1 and T2 images and central linear hypointensity may also exist. Therefore, an MRI is necessary to help ensure the diagnosis of CD. The subtypes of CD can be differentiated through CT. The PC and HV types differ in enhancement on CT due to their pathologic characteristics. The HV type shows homogeneous high enhancement due to the dilation of capillary vessels and the abundance of blood vessels with abnormal proliferation. In contrast, the PC type exerts mild or absent homogeneous enhancement. However, PC type might also exhibit a remarkably high degree of enhancement, which is attributed to the many feeding arteries adjacent to the mass. As a result, the CT enhancement degree cannot differentiate the two pathologic types.

CD cannot be easily distinguished from other tumors through their manifestations or radiologic findings. Some lymphatic correlation diseases, such as acquired immunodeficiency syndrome, have various morphologic lymph node manifestations, which may be indistinguishable from CD. In particular, a differential diagnosis between lymphoma and disseminated CD requires a pathological examination, and the laparoscopic approach combined with lymph node biopsy may be more helpful. Some radiological signs, including remarkable enhancement, hemorrhage and a lack of enlarged lymph nodes, can also be useful for differential diagnosis. CT or ultrasound can only provide accurate tumor location. The differential diagnosis between these two conditions requires both imaging and the examination of tumor biomarkers, but the differential diagnosis between a renal mass and pancreatic carcinoma might be easier. The UCD type responds well to surgical resection, as the literature reported, the laparoscopic approach is an effective therapy method,[4] and there was no recurrence in our patients.

CD should be considered in the differential diagnosis of unclear pararenal masses that mimic other tumors. Clinical experience may help to insure the proper diagnosis through the clinical manifestations, but improving diagnostic examinations is still needed. We demonstrated that retroperitoneoscopic laparoscopic management is effective with a good prognosis following surgery.

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Conflicts of interest

There are no conflicts of interest.

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