Castleman disease mimicking an adrenal tumor: A case report

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ABSTRACT

Castleman disease is a benign complex lymphoproliferative disease. The most common site is the mediastinum. In this paper, we present a case of Castleman disease in the adrenal gland, as a very rare region.

A 29-year-old woman was referred to our clinic due to a well-defined right adrenal mass. She underwent laparoscopic adrenalectomy with the transperitoneal approach. Microscopic histopathology confirmed the hyaline vascular type of Castleman disease.

In conclusion, Castleman’s disease, as a rare disorder, must be considered in the differential diagnosis of an adrenal mass especially in cases with an enhancing well-defined, nonfunctional solid adrenal mass with lymphadenopathy.

1. Introduction

Angiofollicular lymph node hyperplasia or giant lymph node hyperplasia, a benign complex lymphoproliferative disease, was first described by Castleman in 1956 at Massachusetts General Hospital. Castleman disease (CD) is an infrequent and poorly understood entity with unknown etiology. CD may occur anywhere in the lymphatic tissue; despite the mediastinum being the most commonly affected site, extrathoracic sites have been reported in the neck, larynx, axilla, mesentry, groin and retroperitoneum.

In this paper, we present a rare case of adrenal mass which was diagnosed as Castleman disease following surgical resection.

2. Presentation of case

A 29-year-old Iranian woman was referred to our clinic with an adrenal mass. She underwent abdominal ultrasonography due to her non-specific abdominal pain. Abdominal ultrasound imaging showed a right adrenal mass measuring 45 × 40 mm in size. On physical examination, the patient had normal blood pressure and no other abnormalities were noted. The results of laboratory studies including hemoglobin, hematocrit, serum electrolyte and renal function tests were within normal limits. Hormonal examination including serum catecholamines, cortisol, aldosterone, 24-h urinary excretion of metanephrine and normetanephrine, and plasma renin activity were also normal.

Abdominal computed tomography revealed a 50 × 45 mm well-defined mass in the right adrenal. No calcification and contrast enhancement were observed in this mass (Fig. 1).

She underwent laparoscopic adrenalectomy with the transperitoneal approach. The operation was uneventful and the patient was discharged after two days with no complications. Microscopic histopathology revealed hyaline vascular type of Castleman’s disease (Fig. 2). The patient was referred to the hematology clinic postoperatively.

3. Discussion

Castleman disease (lymphoid hamartoma or follicular lymphoreticuloma), is a rare benign lymphoproliferative disorder. The etiology of CD is not yet discovered. There are certain theories regarding its pathogenesis including the abnormal expression of Interleukin (IL)-6 and viral stimulation and angiogenesis due to the high expression of vascular endothelial growth factor (VEGF).

CD has three histological patterns and heterogeneous clinical manifestations. The histological patterns consist of the hyaline vascular type, plasma cell type and the mixed type. The hyaline vascular type is the most common pattern and accounts for up to 90% of CD cases. This type is diagnosed by abnormal lymph node follicles and vascular proliferation in the interfollicular region. Patients with the hyaline vascular type are usually asymptomatic. The plasma cell type is less common, and characterized by sheets of polyclonal plasma cells and a few vessels. Patients who suffer from the plasma cell type CD manifest symptoms such as anemia, fever, fatigue, hyperglobulinemia and...
hypoalbuminemia.²

From the clinical aspect, CD has been categorized into unicentric (UCD) and multicentric (MCD) variants. UCD is the most frequent form of CD and manifests as a well circumscribed or infiltrative solitary mass in young ages.²,³ The most common site of UCD is the mediastinum and retroperitoneal involvement has been documented in 7% of the cases. Based on our literature review, only seventeen cases of CD of the adrenal gland have reported.⁴,⁵ UCD is treated by radical surgery for removing the solitary mass and is associated with an excellent prognosis in long-term. The 5-year survival rate after complete mass excision is about 100%.⁴ In the present case, we performed laparoscopic adrenalectomy for resecting the adrenal mass, as the diagnosis of CD was confirmed after surgery. Nevertheless, adrenalectomy is the treatment of choice for this patient and therefore she did not require any additional therapeutic measures.

MCD is the more aggressive form of CD which is usually diagnosed in the sixth decade. Some clinicians believe that MCD is a potentially malignant disease that manifests with systemic symptoms including constitutional symptoms, autoimmune presentations, peripheral polyneuropathy, organomegaly, endocrinopathy and skin lesions.⁵ The paraclinical findings of MCD include anemia, hypoalbuminemia, polyclonal gammopathy, elevated erythrocyte sedimentation rate or C-reactive protein concentration and proteinuria.⁶ Although, the best treatment is not yet determined, clinicians use the combination of chemotherapy, systemic steroid therapy and radiotherapy in this respect. Despite the various treatment modalities, its mortality rate is still very high and around 50%.²,⁴

Fig. 1. Abdominal computed tomography revealed a well-defined mass in the right adrenal.

Fig. 2. Microscopic histopathology revealed hyaline vascular type of Castleman’s disease.
4. Conclusion

Castleman’s disease, as a rare disorder, must be considered in the differential diagnosis of a nonfunctional adrenal mass especially in cases with an enhancing well-defined solid adrenal mass with lymphadenopathy.

Author disclosure statement

No competing financial interests exist.

Declaration of competing interest

Authors have no conflict of interest.

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