Unicentric Castleman's Disease

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ABSTRACT

Castleman's disease (CD), a rare disease of lymph nodes and related tissues is an atypical lymphoproliferative disorder. It occurs in two forms unicentric and multicentric. Unicentric CD commonly occurs in the mediastinal region. Here we present a case of unicentric CD in a retroperitoneal lymph node.

Keywords: Castleman's disease, lymphoproliferative disorder, unicentric

Castleman's disease (CD) is a rare form of lymph node hyperplasia of unknown etiology.[1] It was first described in 1954, and subsequently better defined by Castleman in 1956.[2] CD is classified into two clinical subtypes: a localized and a multifocal subtype. CD may occur anywhere along the lymphatic system, although the most common location (70%) is the mediastinum. Extrathoracic sites have been reported in the neck, axilla, pelvis and retroperitoneum.[2]

Unicentric forms of CD have been reported as single, mediastinal masses with systemic symptoms that could be resolved after surgical excision. On the other hand, patients with multicentric CD, defined by the involvement of at least 2 noncontiguous, lymph node areas, were often refractory to treatment and show worse clinical outcomes.[3]

Surgery is the optimal therapeutic approach only in the localized form, while for unresectable or disseminated disease, partial surgical resection, steroids, chemotherapy and radiotherapy have been employed with some measurable success.[2] There are three major histological subtypes: hyaline-vascular CD (HV-CD), plasma cell CD (PC-CD) and a plasmablastic-variant associated with human herpesvirus 8 and human immunodeficiency virus. The first is much more frequent (91-96%). The majority (57-91%) of localized disease is hyaline-vascular.[2]

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CASE REPORT

A 31-year-old lady presented to outpatient department with 20 days history of pain upper abdomen radiating to back. Ultrasonography of abdomen and computed

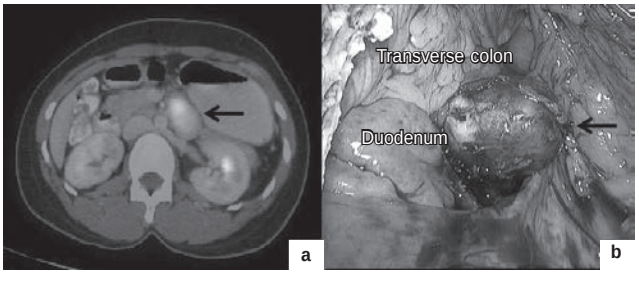

Figure 1. Hypermetabolic lesion of size 3.5 × 2.5 cm along lower border of pancreas (*arrow head*) **(a)**; well-circumscribed lymph node mass in retroperitoneum along lower border of pancreas *(arrow head)* **(b)**.

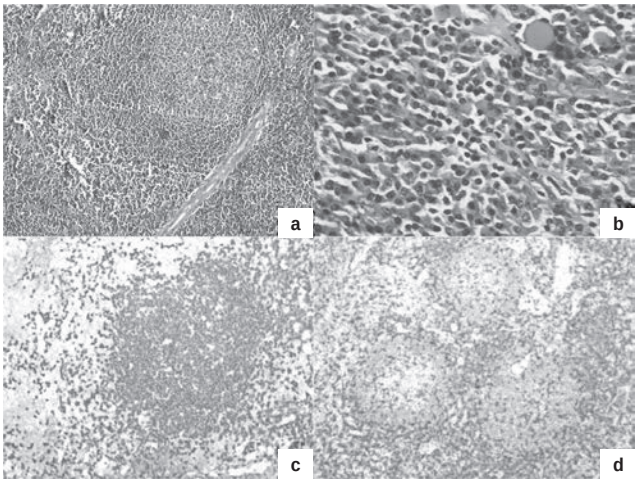

Figure 2. Atretic germinal center with prominent mantle zone and increased vascular proliferation **(a)**; interfollicular prominence of plasma cells **(b)**; CD20 diffuse positive in follicles **(c)**; CD3 highlighting interfollicular T cells **(d)**.

tomography (CT) of abdomen done elsewhere were suggestive of para-aortic lymph node mass along the lower border of pancreas. Guided fine needle aspiration cytology (FNAC) done from the lymph node mass was inconclusive. Positron emission tomography (PET) CT showed hypermetabolic lesion of size 3.5 × 2.5 cm in para-aortic region along inferior border of pancreas (Fig. 1a). Patient underwent laparoscopic excision of lymph node mass (Fig. 1b).

Histopathological examination (HPE) showed atretic germinal center with prominent mantle zone and increased vascular proliferation with interfollicular prominence of plasma cells (Fig. 2 a and b). Immunohistochemistry (IHC), showed CD20 diffuse positive in follicles and CD3 positivity highlighting interfollicular T cells (Fig. 2 c and d). Based upon aforementioned findings diagnosis of unicentric Castleman's disease (mixed variant) was made.

REFERENCES

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