

Clinical Image

Retroperitoneal Castleman's Disease of the Hyaline-Vascular Type

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Clinical Image

A 41-year-old man presented with mild and fitful abdominal pain which had progressed over the preceding 2 weeks. His past medical history was normal. The physical examination was unremarkable during acute episodes. Complete blood count, electrolytes, immunoglobulins, and liver function tests were normal. Auto antibodies tested, viral detection, and neoplastic markers were negative. Abdominal Computed Tomography (CT) scanning was performed to further investigate the source of abdominal pain. CT revealed a homogeneous-solid mass in the retroperitoneal space without colliquation necrosis, measuring 4.2 cm × 3.2 cm, between the aortaventrals and the left adrenal gland, which was further defined by magnetic resonance imaging (Figure 1A and 1B). After admission, operative resection of the mass was performed. The resected specimen was covered with a capsule, measuring 4.0 cm × 2.5 cm × 2.5 cm with the rufous cut surface macroscopically (Figure 1C). Microscopically, the specimen was characterized by the presence of regressed atretic germinal centers with an onion-skin appearance, increased vascularity with hyalinization and radially penetrating blood vessels leading to a lollipop appearance (Figure 1D), which was further highlighted by the lesion staining positively immunoreactive for CD20, CD3, CD21 (representative of dysplastic follicular dendritic cells), BCL2 (except in the germinal centers), CD10 (only in the germinal centers), and a proliferation index Ki-67 (mainly in the germinal centers) by immunohistochemistry (Figure 2). The patient received the diagnosis of a stromal-rich variant of Castleman's disease of hyaline-vascular type on the basis of the above histologic findings. The differential diagnosis included autoimmune disease, lymph node draining carcinomas, and acquired immune deficiency syndrome. Castleman's Disease is a rare, benign, and lymphoproliferative

disorder, partially related to Epstein-Barr and cytomegalovirus infection, which often appears in the mediastinum [1]. However, retroperitoneal involvement is uncommon [2]. The hyaline vascular type is the most common type of Castleman's Disease distinguished by histological technique. The more common localized solitary form of Castleman's Disease usually reveals a benign behavior with recovery after surgical resection irrespective of the histopathology [1]. Surgery, corticosteroids, chemotherapy, monoclonal antibodies, and immune modulators are currently clinically applied [3]. The therapeutic approaches remain controversial and mainly depend on the different forms of Castleman's Disease according to localization. The less frequent multicentric form is considered to be associated with a poor prognosis and a more aggressive course [3]. After surgery, there was no tumor recurrence for this patient and no episodes were noted in the next several years.

Keywords

Castleman's Disease; Computed tomography; Chemotherapy; Hyaline-vascular type

Ethical Statement

The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Written informed consent was obtained from the patient for publication of this "GI Image".

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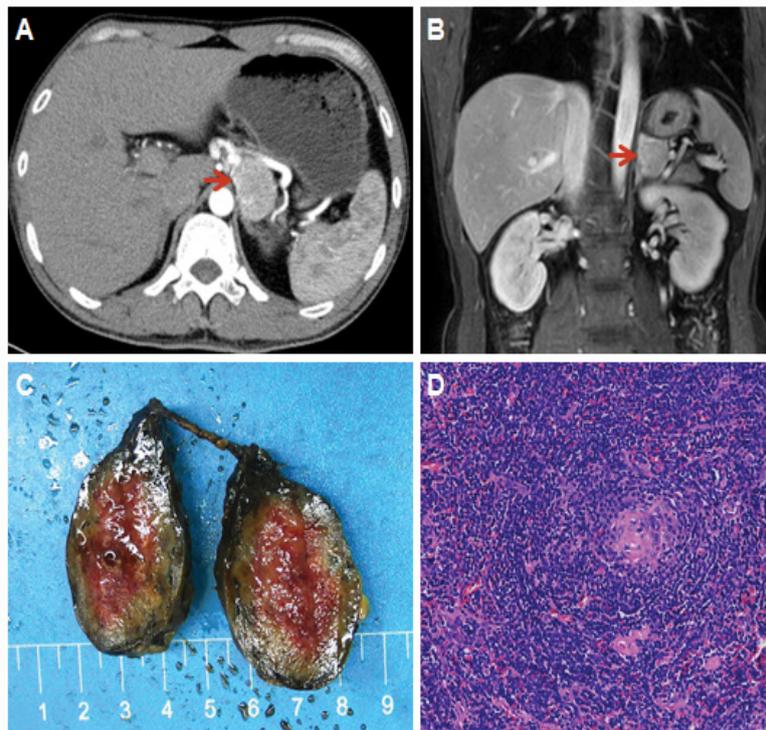


Figure 1: Retroperitoneal Castleman's Disease of the Hyaline-Vascular Type. A and B): A homogeneous-solid mass in the retroperitoneal space defined by computed tomography and magnetic resonance imaging. C): Operative resection of the mass. D): Hematoxylin and eosin staining of the lesion.

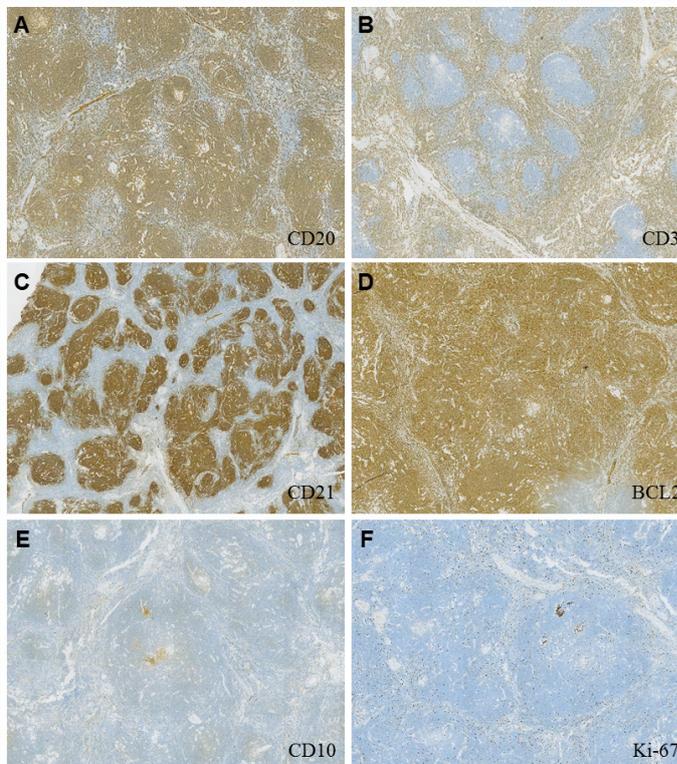


Figure 2: Retroperitoneal Castleman's Disease of the Hyaline-Vascular Type. A-F): Immunohistochemical analysis positive for CD20, CD3, CD21, BCL2, CD10 and Ki-67 in neoplastic cells.