

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Surgery Quiz – Case 9

A 40-year-old female was admitted for management of an asymptomatic left pararenal mass. Clinical examination and past medical history were unremarkable. Thoraco-abdominal computed tomography (CT) scan demonstrated a 7.6×5.3×12 cm large heterogeneous lesion adjacent to the left kidney. The mass was regularly limited and exhibited hypervascularity, without invading any adjacent structures. The rest of the abdominal cavity was unremarkable (fig. 1). Further investigation with magnetic resonance imaging (MRI) confirmed the lesion exhibiting both T1 and T2 sequence hyperintensity in a heterogeneous manner (figures 2a, 2b). The patient was operated via a left flank approach under general anesthesia. The mass was removed in its entirety. Postoperative course was uneventful and the patient was discharged on the 5th postoperative day.

Pathology report demonstrated a lymph node specimen constituted by small-sized follicles with atrophic germinal centers and hyperplastic follicular mantle region. The germinal centers were almost totally replaced by endothelial and dendritic cells. Mantle cells were arranged to concentric layers surrounding the atrophic germinal centers. The interfollicular area abounded in numerous post-capillary venules, small vessels, with concentric fibrosis or hyalinization of their wall, and multiclonal plasma cells, while the number of the lymphocytes was reduced. No

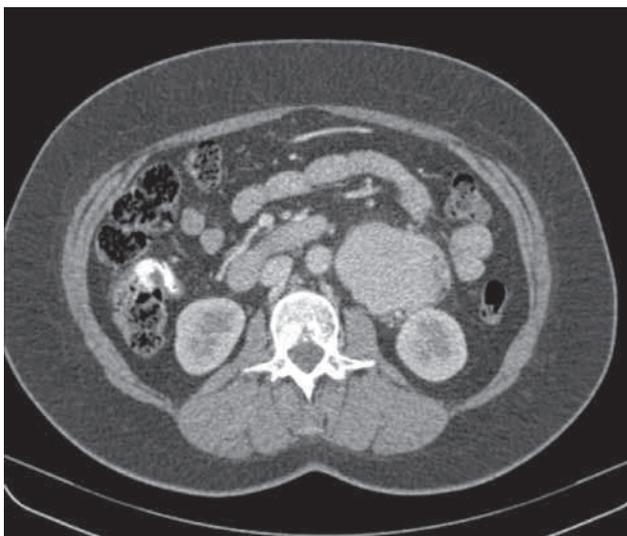


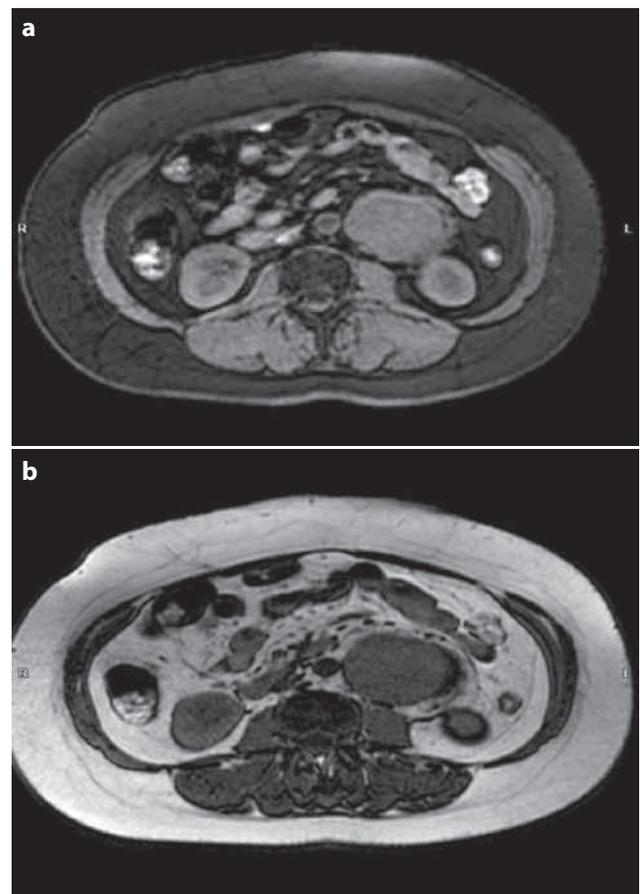
Figure 1

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2013, 30(1):119–120

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normal lymphoid tissue or lymph node sinus was recognized circumferentially. The immunohistochemical assays highlighted the CD20 expression of the mantle cells surrounding the hyaline-vascular germinal center follicles, the dendritic cell hyperplasia,



Figures 2a and 2b

prominent vascularity of the interfollicular area and reduced expression of Ki67 in the atrophic germinal centers. HHV-8 staining was negative.

Comment

Castleman's disease (hereafter depicted as CD), also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia or lymphoid hamartoma, is a non-clonal, non-neoplastic lymph node proliferation first described by Castleman and Town in 1954. CD has a peak incidence in females on the third and fourth decade of life.

Clinically, there are two distinct types of the disease: The unicentric and multicentric subtypes. It is of pivotal importance to identify unicentric as opposed to multicentric disease at a clinical level in a stepwise approach. Histologically, CD consists of three variants: hyaline-vascular (HV), plasma cell (PC) or mixed cellularity (mixed). The HV variant is described in 90% of CD cases and is characterized by the proliferation of capillary vessels in germinal centers of lymphatic follicles.

The distribution of localized CD has been reported as 65% in the mediastinum, 16% in the neck, 12% in the abdomen, and 3% in the axilla. Mediastinal CD can mimic thymoma, lymphoma, sarcoma, hemangiopericytoma, neural crest-derived neoplasms such as paraganglioma, neurofibroma, or schwannoma, and chest wall tumors.

Complete surgical excision is the treatment of choice for localized, unicentric lesions in any organ domain; cytoreduction of radiotherapy has also been advocated in cases where complete resection is not feasible. In a recent report from Memorial Sloan Kettering Cancer

Centre, complete resection of unicentric disease was curative for all patients regardless of histological subtype. Likewise, Keller et al, retrospectively, examined 61 patients with unicentric disease who were treated with surgery over a 20 year period. Their study demonstrated that for patients with unicentric HV-CD, complete resection offered the best chance for cure.

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