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Retroperitoneal inflammatory myofibroblastic tumor: A case report

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ABSTRACT

Inflammatory myofibroblastic tumors (IMT) were previously included in the "inflammatory pseudotumors" family, but have emerged as a distinct entity recently. The management of IMT may be challenging due to its intermediate malignant potential. We present a case of a young patient with an asymptomatic retroperitoneal mass with some imaging findings that could point to the IMT diagnosis pre-operatively. After two failed attempts to fully characterize the lesion by percutaneous and surgical biopsy, surgical resection was conducted and the final diagnosis was achieved.

1. Introduction

In recent years, the inflammatory myofibroblastic tumor has emerged as a distinct entity with characteristic clinical, pathological and molecular features, being previously included in the "inflammatory pseudotumors" family [1,2]. These lesions are found mainly in children and young adults and can mimic a true malignancy [3]. Their management is not well defined and can be challenging, and most of the time, surgical diagnosis is needed [2–4]. We present the case of a patient with a retroperitoneal IMT that was surgically removed.

2. Case report

A 21-year-old female was admitted to the Emergency Room of our hospital with abdominal pain, mainly in the right iliac fossa, over the last two days. She denied nausea, vomiting or anorexia. The medical history was unremarkable. On the physical examination, she displayed pain with palpation of the right abdominal quadrants. The basic lab work was also unremarkable. It was requested an abdominal ultrasound (Fig. 1) that showed the existence of a heterogeneous mass, mostly isoechoic to muscle with some hypoechoic areas, between the right kidney and the vertebral body, posteriorly to the renal artery and vein. It measured about 5, 5 × 4, 7 cm and it seemed to displace the regional anatomy, more than invading it. Moreover, the mass was vascularized on Doppler ultrasound. Regarding the lack of specificity of the ultrasound findings, an abdominal CT was suggested.

The abdominal CT was performed two days later, showing a solid and heterogeneous mass in the retroperitoneum, with lobulated contours, resulting in lateral shift of the right kidney and anterior shift of the renal vasculature (Fig. 2). There were some linear calcifications. In the enhanced scans, it showed very high uptake of iodinated contrast, suggesting an hypervascular mass, with a hypodense central area, that was assumed as being due to tumor necrosis.

There was some fat stranding around the mass, without signs of invasion of the nearby organs. There were no signs of metastatic disease in the abdomen.

At this point, our differential diagnosis consisted in paraganglioma or another neurogenic tumor or a sarcoma. Paraganglioma was considered given the location of the mass (being closely related to the sympathetic nervous system) and suitable CT appearance (although not specific for this diagnosis, paragangliomas can show high uptake of iodinated contrast and central necrosis as well as punctuate calcifications[5]). We suggested an abdominal MRI to further characterize the lesion as well as a MIBG I123 scintigraphic scan. The scintigraphic scan didn't show any uptake by the lesion, virtually excluding a neurogenic origin. It was also performed a SPECT/CT examination for better anatomical correlation with the scintigraphic scan (Fig. 3).

Although abdominal MRI didn't contribute much in terms of lesion characterization, it reinforced the probable necrotic center and showed high diffusion restriction in the hypervascular areas (Fig. 4). The diagnosis of sarcoma (and given the age of the patient

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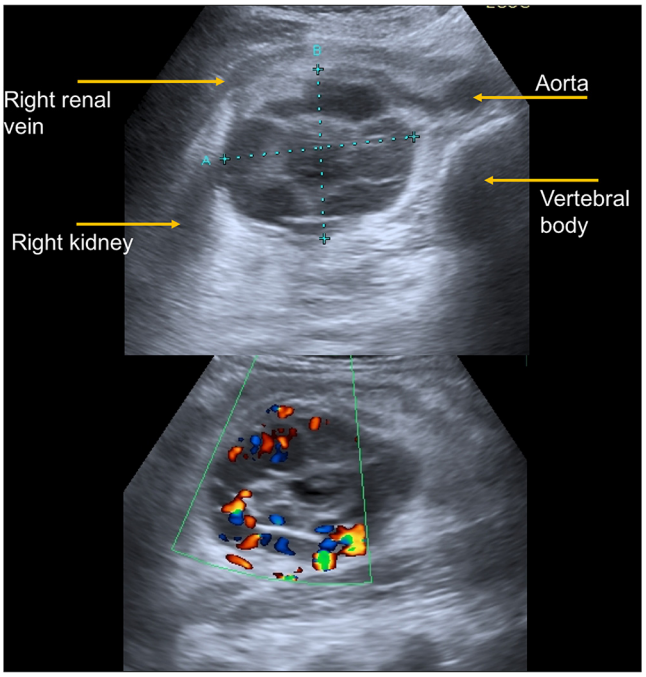

Fig. 1. Ultrasound study, both in B and Color-Doppler modes.

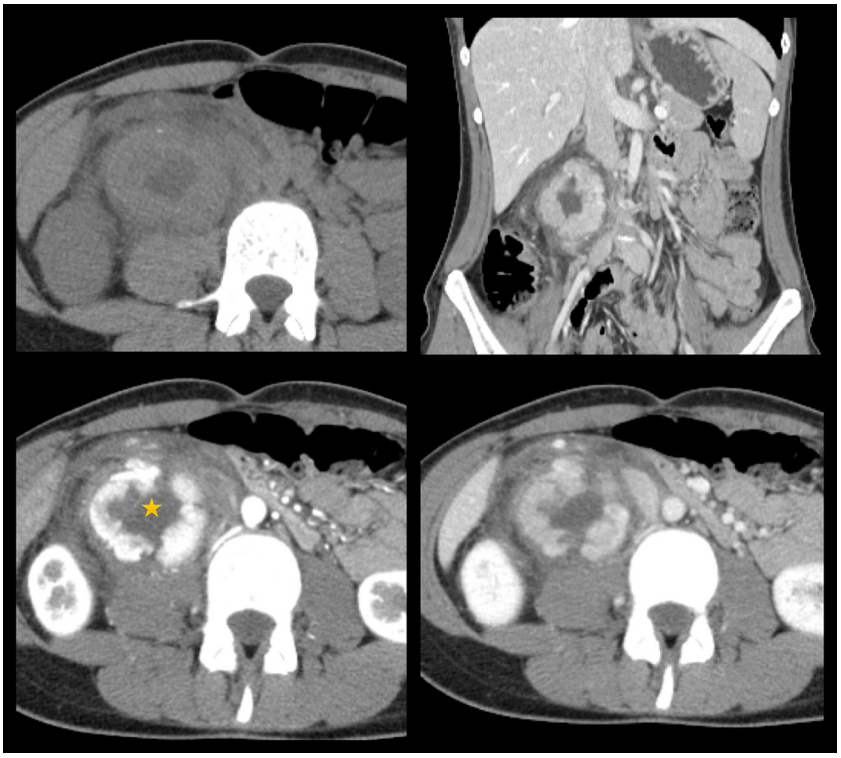

Fig. 2. Abdominal CT, from left to right, top to bottom: unenhanced scan, contrast-enhanced scan in portal phase on the coronal plane, arterial phase, portal phase. A hypervascular mass is seen displacing the right kidney, with a hypodense center (*), compatible with necrosis.

and the image findings, particularly the hypervascularity and center necrosis, a leiomyiosarcoma was the most probable [6,7]) was maintained in the differential, as could not be ruled out completely, even more as the scintigraphic scan was negative.

A biopsy was requested, that was performed with CT guidance. Pathology reported that there were some lymphoid structures with such architectural changes that resembled a lymphoproliferative disease, such as Hodgkin lymphoma. On a multidisciplinary meeting, it was decided to perform a surgical biopsy, which reported it as being part of a lymph node with reactive changes. It was then decided to surgically excise the mass.

Pathology reported the mass as having a capsule and some fusiform cells, as well as diffuse inflammatory infiltration. No Hodgkin or Sternberg-Reed cells were found. The immunochemistry tests showed reaction to actin in the fusiform cells and lymphoid markers (Fig. 5). IMT's are composed predominantly of