



Retroperitoneal inflammatory myofibroblastic tumor: A case report

Carlos Oliveira*, Rui Costa, Amélia Estêvão, Filipe Caseiro-Alves

Medical Imaging Department, Coimbra University Hospital, Praceta Mota Pinto/Av. Bissaya-Barreto, 3000-075 Coimbra, Portugal

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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.

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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 calcifications⁵). 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

* Corresponding author.

E-mail addresses: carlosmigoliveira@gmail.com (C. Oliveira), alvescosta87@gmail.com (R. Costa), ameliaestevao50@gmail.com (A. Estêvão), caseiroalves@gmail.com (F. Caseiro-Alves).

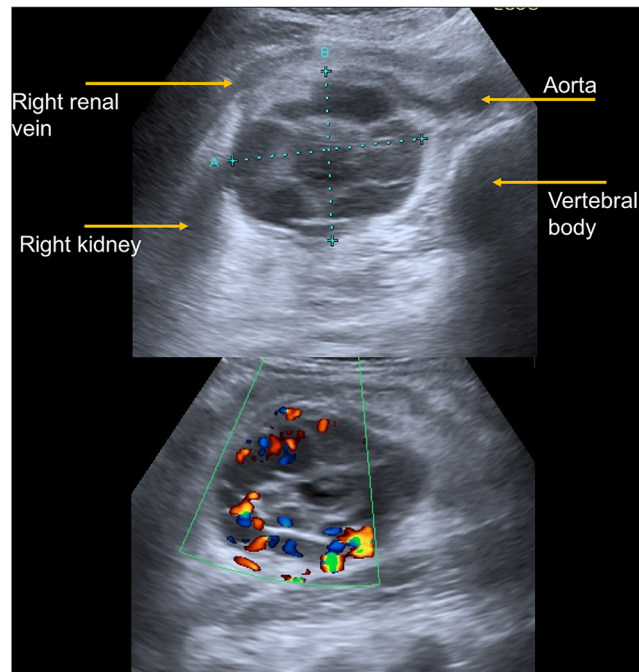


Fig. 1. Ultrasound study, both in B and Color-Doppler modes.



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 leiomyosarcoma 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 meet-

ing, 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 immunohistochemistry tests showed reaction to actin in the fusiform cells and lymphoid markers (Fig. 5). IMT's are composed predominantly of

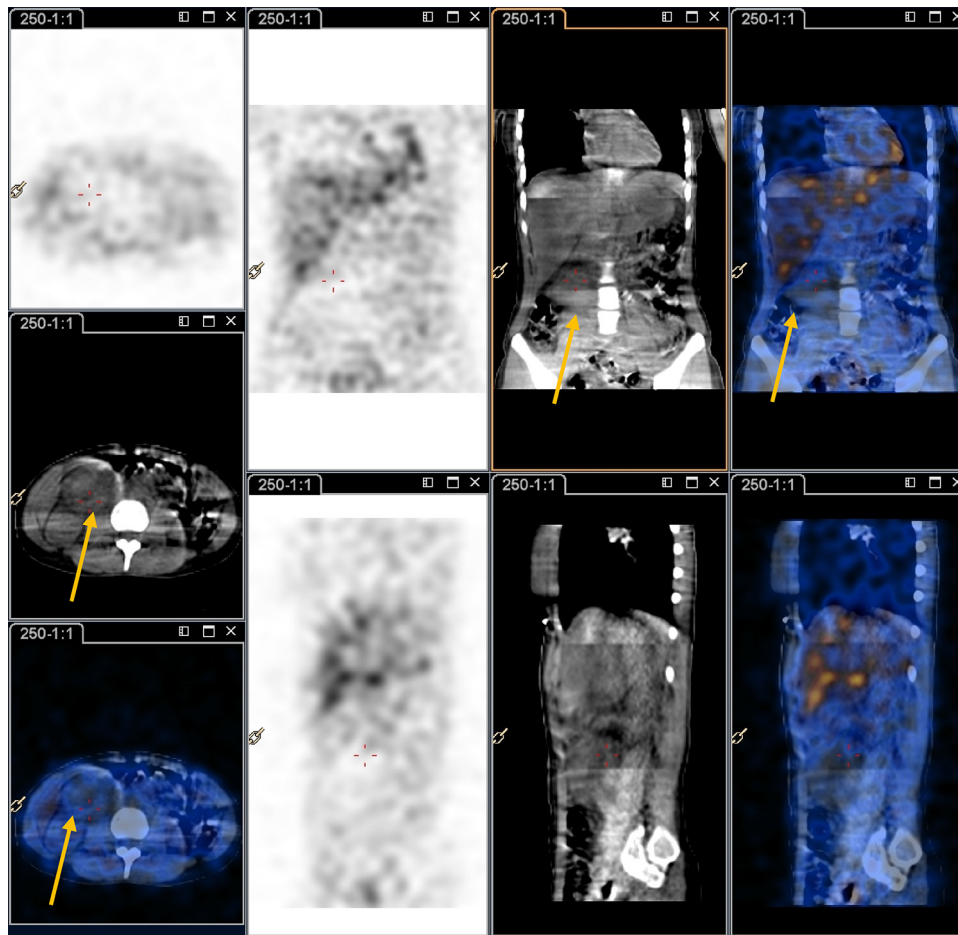


Fig. 3. MIBG I^{123} scintigraphic scan with SPECT-CT. The lesion is marked with the yellow arrow, not showing any kind of uptake of MIBG I^{123} .

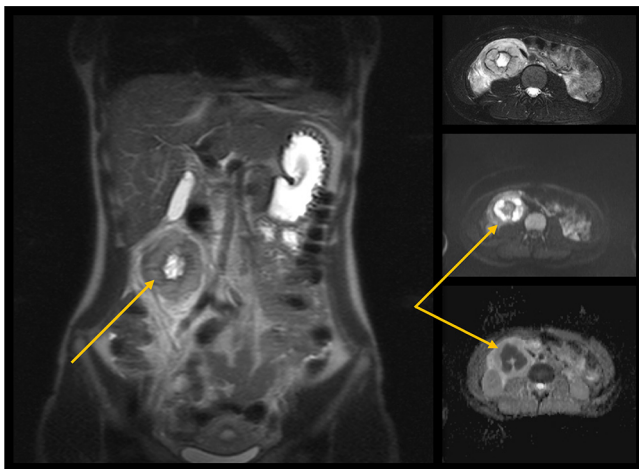


Fig. 4. MR study, on the left T2WI acquired in singleshot, on the right top to bottom: T2 with fat saturation, DWI-b1000, ADC map. As with the CT images, a mass is seen displacing the right kidney, with very hyperintense center (necrosis) and very high diffusion restriction in the solid portion of the mass.

myofibroblasts, whose positivity for actin reaches 90% of the cases [1]. At the same time, there is usually a prominent inflammatory infiltrate of polyclonal origin in these tumors, susceptible of being mistakenly associated with lymphoproliferative diseases, as suggested in the first histological diagnosis of this case [1]. These two factors were of utmost importance in the final diagnosis of an inflammatory myofibroblastic tumor.

3. Discussion

Inflammatory myofibroblastic tumors are difficult to diagnose and there is no consensus regarding their treatment [2]. The symptoms of IMT depend on the site of tumor origin [5]. The majority of IMT are found in the abdominopelvic region, lung and retroperitoneum, although they can appear in a myriad of locations [1]. Most abdominal IMT are usually asymptomatic and, for the most part, found incidentally. In this very patient, the lesion only became symptomatic when reached considerable dimensions and exerted mass effect on the remaining abdomen organs. On imaging, these tumors can show a wide range of findings, regarding their size, location or appearance [3,5]. The literature shows that they can have different appearances regarding the organ of origin. However, even on retroperitoneal tumors, there are no specific findings that can assure the diagnosis without histologic analysis [3,5]. Nonetheless, large lesions may present with nonenhancing central necrotic areas and calcifications as well as early prominent enhancement that persists at delayed imaging [5].

In our patient, the mass showed early and profuse enhancement, with a necrotic center, matching the commonly reported findings. Moreover, there was some fat stranding around the mass, a feature typically thought as of inflammatory origin, which also occurs in invading malignancies. The first biopsy result reported a possible Hodgkin lymphoma but the CT and MRI findings were atypical for lymphoma, as it usually does not show calcifications before the start of treatment or presents as an hypervascular mass [7]. Moreover, lymphoma usually shows a type of growth that “encases” the retroperitoneal structures, more than displaces them [6,7]. No

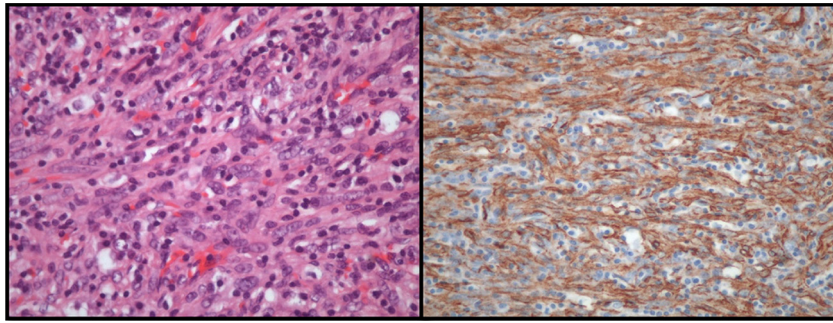


Fig. 5. Left: Hematoxylin-Eosin: multiple polymorphous cells showing no atypia; right: Intense reaction to actin.

adenopathy was reported both on CT and MRI. This posed as an intersection in the patient management chain (chemotherapy vs. surgical resection) and a surgical biopsy was warranted, that only showed some lymphoid tissue without atypia. This ultimately led to the surgical resection and the final diagnosis.

Surgical excision is the treatment of choice for patients with IMT [4]. Although there are reports of partial response with chemotherapy [2,8], that can be used mainly in case of unresectable tumors. Recurrence is seen in about 25% of cases and distant metastases to the lungs, brain, liver, and bone are possible but unlikely [4,5]. Our patient remains disease-free two years after surgery.

4. Conclusion

Inflammatory myofibroblastic tumor is a rare tumor of childhood and young adults with a wide spectrum of clinical and radiologic findings because of the variability of location.

These lesions can be challenging to diagnose to clinicians, radiologists or even pathologists, requiring a multi-disciplinary approach to better evaluate each case individually.

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