Radiología 64 (2022) 490–493

RADIOLOGÍA

www.elsevier.es/rx

SCIENTIFIC LETTER

Retroperitoneal Ewing Sarcoma: a challenging diagnosis☆

Sarcoma de Ewing retroperitoneal: un reto diagnóstico

P. Costa[a,*], **C. Costa**[b]

[a] *Departamento de Diagnóstico por la Imagen, Unidade Local de Saúde de Matosinhos, Matosinhos, Portugal*
[b] *Departamento de Endocrinología, Instituto Português de Oncologia do Porto, Porto, Portugal*

Ewing Sarcoma (ES) is an aggressive neoplasm, typically arising from bones and presenting in pediatric age.[1] Extraskeletal origin is rare and its presentation in age > 30 years is even rarer, although most cases of ES in this population arise from soft tissue.[1–3]

We present herein a case of a primary retroperitoneal ES aiming to highlight potential clues to its recognition by radiologists.

A 38-year-old female patient presented to a primary care facility with complaints of abdominal pain centered to the left upper quadrant and asthenia for a week. Physical examination was unremarkable. An abdominal ultrasound was performed, showing a large predominantly solid hypovascular mass at the left upper abdomen.

Computed tomography (CT) was then performed, confirming a 14 cm well-defined soft-tissue mass in the left retroperitoneum (Fig. 1A), which displaced and deformed all adjacent organs, without clear signs of invasion (Fig. 1B-F). The adrenal gland is only clearly depicted in the sagittal and coronal reconstructions, which may have hampered its recognition in the first interpretation of the images, resulting in a presumptive diagnosis of adrenal tumour.

The patient was then referred to our institution, a tertiary cancer centre, where a complete hormonal panel was requested, with no reported abnormalities.

Magnetic Resonance Imaging (MRI) was requested for preoperative planning. The lesion showed restricted diffusion (Fig. 2A-B) and areas of high T2 signal (Fig. 2C), suggesting necrosis. Enhancement was heterogeneous and progressive with an enhancing rim resembling a pseudo-capsule (Fig. 2D). MRI also confirmed close contact with adjacent organs, without unequivocal signs of invasion. Positron emission tomography (PET)/CT demonstrated a Fluorine-18-fluorodeoxyglucose (^{18}F-FDG)-avid mass, without evidence of metastatic disease (Fig. 2E-F).

Given the feasibility of complete excision, an extended resection was conducted. Pathology showed a tumour with a fibrous pseudo-capsule, adherent to the kidney, spleen, pancreas, and duodenum, without invading them. The left adrenal gland was contained within the pseudocapsule, with signs of microscopic invasion. The tumour comprised undifferentiated small round cells, with areas of necrosis. Immunohistochemistry showed CD99 expression and genetic testing demonstrated EWSR1 gene rearrangement, confirming the diagnosis of ES. Resection margins were tumour-free.

Another PET/CT was performed two months after surgery showing recurrent disease. The patient was treated with palliative chemotherapy.

* Corresponding author.
E-mail address: patriciamdc24@gmail.com (P. Costa).

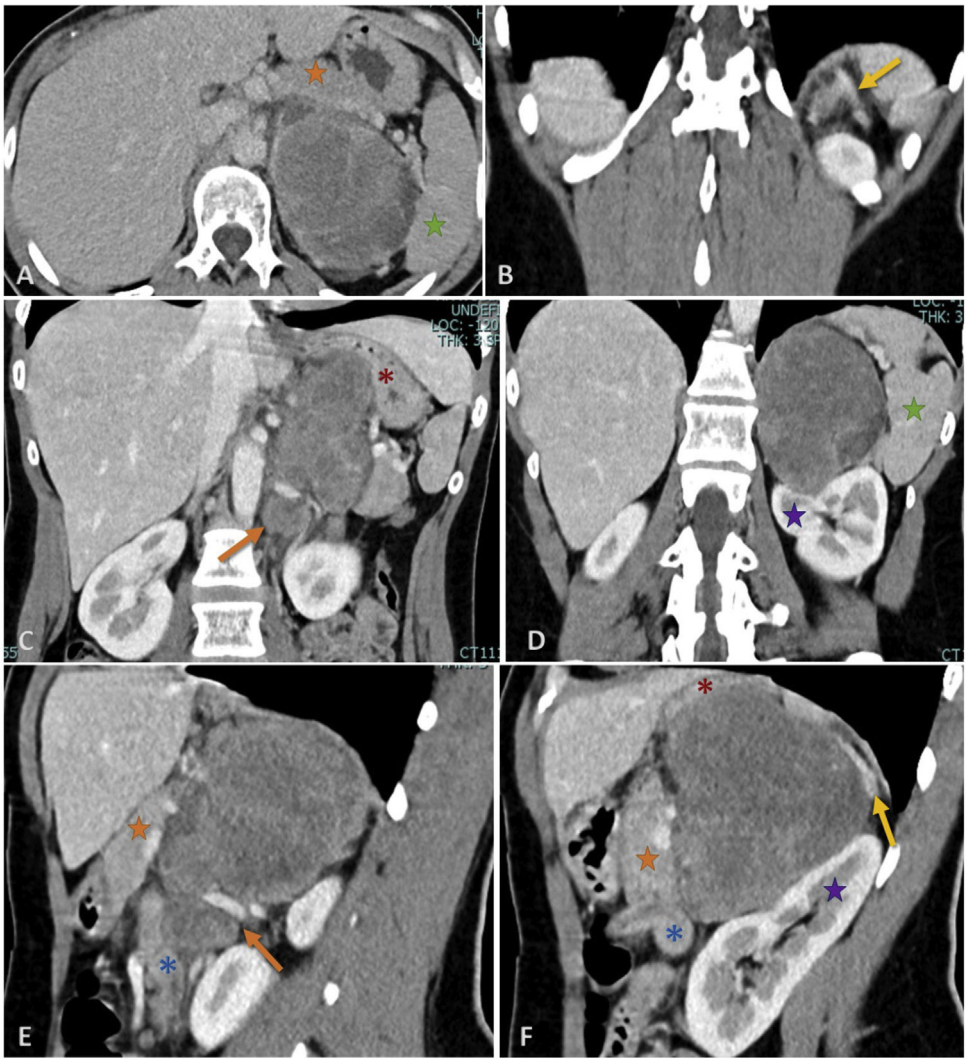

Figure 1 Contrast-enhanced abdominopelvic CT scan, soft tissue window (axial plane – A, coronal reformatted plane – B, C and D, sagittal reformatted plane – E and F) confirms the presence of an heterogeneous enhancing mass in the left upper retroperitoneum, displacing and deforming all adjacent organs (stomach – red asterisk, duodenum – blue asterisk, spleen – green star, pancreas – orange star, adrenal gland – yellow arrow, and kidney – purple star), with loss of fatty plane between these organs and the lesion. There are no clear signs of invasion of adjacent organs, however the mass is seen protruding into the renal sinus (orange arrow).

Evaluation of retroperitoneal masses should start by assessment of tumour location.[4] Once a tumour is located to the retroperitoneum, efforts should be made to identify the organ of origin, as most retroperitoneal masses arise from solid organs.[5] The ''phantom organ sign'' (concealment of a small organ by a large mass arising from it) should be used with care,[4] as large masses can turn small adjacent organs (as the adrenal gland) difficult to outline, as illustrated in the first CT evaluation (Fig. 1B and 1F). A negative ''embedded organ sign'' (when all adjacent organs are compressed and deformed into a crescent shape, without signs of invasion) allows to identify a mass as a primary retroperitoneal neoplasm[4] (Fig. 1).

A predominantly solid composition, as presented here, excludes more common cystic neoplasms, as lymphangiomas and mucinous cystic tumors.[4,5] The absence of fat and calcifications (Fig. 1) makes liposarcoma (the most common sarcoma of the retroperitoneum) and teratoma less likely. Necrosis (Fig. 2C-D) favours high-grade malignancies, namely sarcomas.[4] ES, in particular, is typically heterogeneous when large, due to necrosis and haemorrhage.[2] Extraskeletal ES can be differentiated from lymphoma based on its expansile pattern of growth (lymphoma usually spreads between normal structures, crossing the midline)[5] and absence of lymphadenopathy, a rare finding in ES.[3] Compression of adjacent organs can result in a pseudocapsule,[5] as identified in this case (Fig. 2D).

Local recurrence, as seen here, and metastatic disease are frequent, contributing to the poor prognosis.[2,3,5]

In conclusion, retroperitoneal ES should be considered whenever a young adult presents with a large unilateral heterogeneous mass centered to the retroperitoneum, not crossing the midline,[3] with an expansile growth leading to compression of adjacent organs, although some degree of infiltration may be present.[2] Presence of necrosis along with absence of

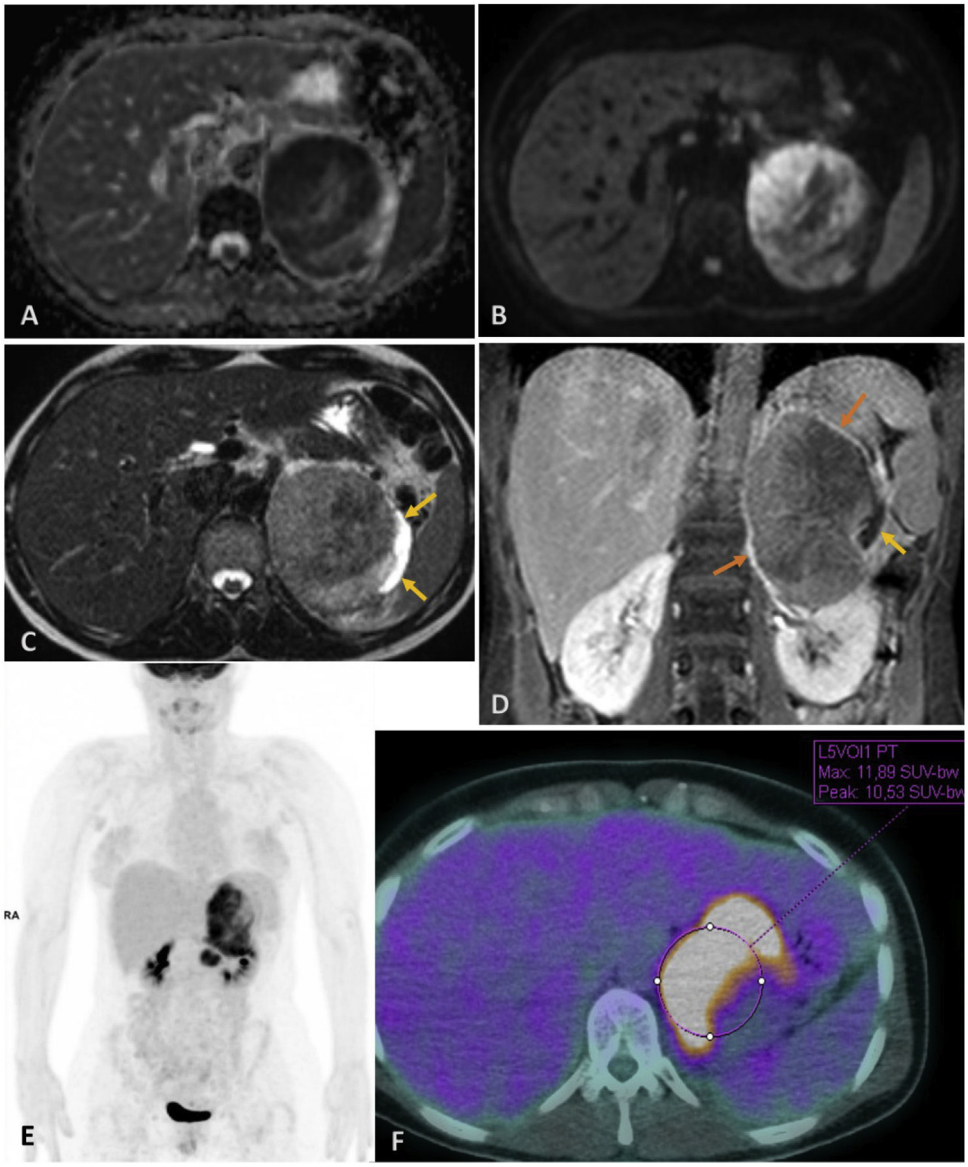

Figure 2 Abdominal MRI, ADC map (A), diffusion (B), axial T2-weighted images (WI) (C) and coronal T1WI with fat suppression after gadolinium administration, delayed phase (D) presents a mass with restricted diffusion and predominantly intermediate signal on T2WI, with an eccentric cystic component suspicious of necrosis (yellow arrow). Post-contrast images show delayed mild enhancement, with a thin enhancing rim, resembling a pseudo-capsule (orange arrow). PET/CT demonstrates that the mass is ^{18}F-FDG-avid, with a high uptake value and the areas of greater avidness corresponding to the areas of restricted diffusion on MRI. PET/CT also confirms that disease is localized to the left upper retroperitoneum, with no signs of distant metastases.

calcifications and lymphadenopathy also favour this diagnosis.[2,3]

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflicts of interest

The authors declare that they have no conflicts of interest.

References

1. Javalgi A, Karigoudar M, Palur K. Blue Cell Tumour at Unusual Site: Retropritoneal Ewings Sarcoma. J Clin Diagnostic Res. 2016;10:ED19.
2. Somarouthu BS, Shinagare AB, Rosenthal MH, Tirumani H, Hornick JL, Ramaiya NH, et al. Multimodality imaging features, metastatic pattern and clinical outcome in adult extraskeletal Ewing sarcoma: experience in 26 patients. Br J Radiol. 2014;87:20140123, http://dx.doi.org/10.1259/bjr.20140123.
3. Javery O, Krajewski K, O'Regan K, Kis B, Giardino A, Jagannathan J, et al. A to Z of extraskeletal Ewing sarcoma family of tumors in adults: imaging features of primary disease, metastatic

patterns, and treatment responses. AJR Am J Roentgenol. 2011;197:W1015–22, http://dx.doi.org/10.2214/AJR.11.6667.
4. Nishino M, Hayakawa K, Minami M, Yamamoto A, Ueda H, Takasu K. Primary Retroperitoneal Neoplasms: CT and MR Imaging Findings with Anatomic and Pathologic Diagnostic Clues. RadioGraphics. 2003;23:45–57, http://dx.doi.org/10.1148/rg.231025037.
5. Rajiah P, Sinha R, Cuevas C, Dubinsky TJ, Bush WH, Kolokythas O. Imaging of Uncommon Retroperitoneal Masses. RadioGraphics. 2011;31:949–76, http://dx.doi.org/10.1148/rg.314095132.