Article published online: 2021-03-31

Pelvic Ewing Sarcoma: The Great Mimicker
Sarcoma de Ewing pélvico: O grande imitador

Francisco Lima-Bernardes[1] Diogo Manuel Soares[1] Joana Monteiro Pereira[1] Isabel Catarino[1] Sofia Esteves Vieira[1] João das Dores Carvalho[1]

[1] Orthopaedic and traumatology doctor, Department of Orthopedics and Traumatology, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal

Address for correspondence Francisco Manuel Flores Lima-Bernardes, MD, Department of Orthopedics and Traumatology, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal (e-mail: franciscolimabernardes@gmail.com).

Rev Bras Ortop 2023;58(5):e822–e825.

Abstract

Ewing sarcoma is the most common malignant bone tumor of the pelvis in children and young adults. Even with aggressive treatment, its survival rate is amongst the poorest. Classical presentation may not be the rule. It may simulate clinically, imagiologically and histopathologically other nonmalignant entities. Therefore, its suspicion should not be overlooked. We report two cases of pelvic Ewing sarcoma: the first mimicking eosinophilic granuloma, and the second mimicking osteomyelitis. In the latter, we also report an atypical finding of its natural history: an initial response to antibiotic and anti-inflammatory treatment. In both cases, we highlight the possibility of an inconclusive percutaneous bone biopsy and the importance of immunochemistry and cytogenetics for the definitive diagnosis.

Keywords
- sarcoma, Ewing
- pelvic neoplasms
- eosinophilic granuloma
- osteomyelitis
- anti-onflammatory agents, non-steroidal

Resumo

O sarcoma de Ewing é o tumor ósseo maligno da pelve mais comum em crianças e adultos jovens. Mesmo com tratamento agressivo, sua taxa de sobrevivência está entre as piores. A apresentação clássica pode não ser a regra. Ele pode simular clinicamente, imaginologicamente e histopatologicamente outras entidades não malignas. Portanto, sua suspeita não deve ser negligenciada. Relatamos dois casos de sarcoma pélvico: o primeiro imitando granuloma eosinofílico e o segundo imitando osteomielite. Neste último, também relatamos um achado atípico de sua história natural: uma resposta inicial ao antibiótico e ao tratamento anti-inflamatório. Em ambos os casos, destacamos a possibilidade de uma biópsia óssea percutânea inconclusiva e a importância da imunoquímica e da citogenética para o diagnóstico definitivo.

Palavras-chave
- sarcoma de Ewing
- neoplasias pélvicas
- granuloma eosinofílico
- osteomielite
- anti-inflamatórios não esteroides

Introduction

Ewing sarcoma (ES) is a primary bone tumor. In > 50% of cases, a soft tissue component is associated.[1] Ewing sarcoma is rare, with an annual incidence of 2.93 per million individuals.[2,3] Amongst the bone tumors, it is one of the most lethal, with high propensity for recurrence and distant metastasis (predominantly to the lungs). Generally, it affects the diaphysis and metaphysis of long bones.[4,5] It is rare in patients > 30 years old, with 90% of the cases affecting individuals < 20 years old.[4] In this age group, it is the most common

Work developed at Orthopaedics and Traumatology Department, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal.

received
July 18, 2020
accepted
September 17, 2020

DOI https://doi.org/10.1055/s-0040-1722582.
ISSN 0102-3616.

Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

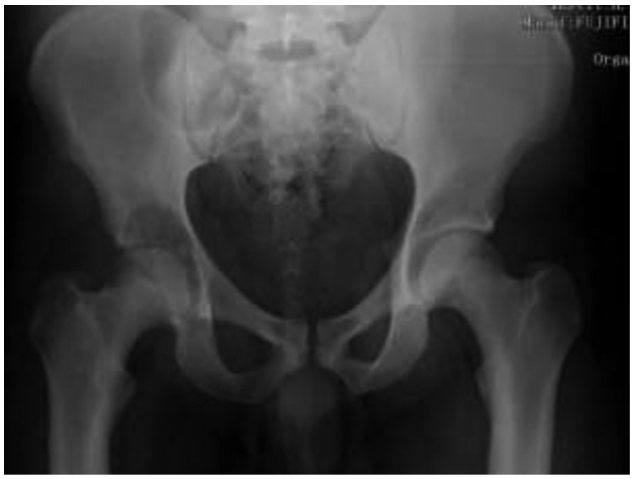

Fig. 1 Radiograph on admission showing a radiolucent lesion of the right acetabulum.

pelvic tumor. The prognosis of pelvic ES is poor, with a 5-year survival rate of 50%.[6] Its most common symptoms are night pain (90%) and swelling (70%), the latter of difficult assessment in pelvic ES. Fever, weight loss, anemia and increased inflammatory markers are uncommon unspecific manifestations.[1,7,8] Radiographs feature lytic destruction and subperiosteal bone formation similar to "onion skinning."[1,4,5,8] These features are most easily detected in long bones in comparison with the pelvis. The value of radiographs in the pelvis is compromised by the anatomy of the iliac bone and of overlying structures.[1,8] Computed tomography (CT) allows a better assessment of bone destruction and may show a soft tissue component.[1,5] Early magnetic resonance imaging (MRI) for soft tissue characterization and, to a lesser extent, identification of a "sharp transition zone", is useful.[3,7] These nonspecific clinical and imagiological findings may be present in conditions such as metastatic carcinoma, malignant lymphoma, osteomyelitis,[4] fibrous dysplasia[5] or eosinophilic granuloma (EG),[9] making its early diagnosis challenging.[1–9] The definitive diagnosis is made by open biopsy and histological examination combined with immunochemistry and cytogenetics.[1,4] The lack or insufficiency of histologic specimens by percutaneous biopsy may lead to its misdiagnosis.[2,4] We report two cases of ES that attended our department in the last 3 years, the first mimicking EG and the second osteomyelitis. Our purpose is to highlight pitfalls on the diagnosis of pelvic ES and to report an atypical finding of its natural history: an initial response to antibiotic and anti-inflammatory treatment, only described once in the literature.[4]

Case Report 1

A 20-year-old man presented to our emergency department with a 6-month history of both mechanical and inflammatory right hip pain and recurrent fever. Blood tests were normal. Radiographs revealed a radiolucent lesion affecting the right acetabulum (►**Fig. 1**). A CT scan showed an osteolytic lesion involving the posterior acetabular rim, with cortical disruption (►**Fig. 2**). A pelvic MRI showed an associated soft tissue component (►**Fig. 3**). This lesion was consistent with ES, not excluding EG. Histopathological examination following image-guided percutaneous bone biopsy was consistent with EG. Due to the risk of fracture, the patient was submitted to curettage and autograft placement. Histopathological examination of new samples was consistent with ES, later confirmed by immunochemistry and cytogenetics. The patient was transferred to a specialized center.

Case Report 2

A 30-year-old man, with former history of syphilis and drug addiction, presented to our emergency department with a 1-month history of right inguinal pain without constitutional symptoms. Blood tests demonstrated an elevated white cell count (15.9×10^9 cells/L) and increased C-reactive-protein

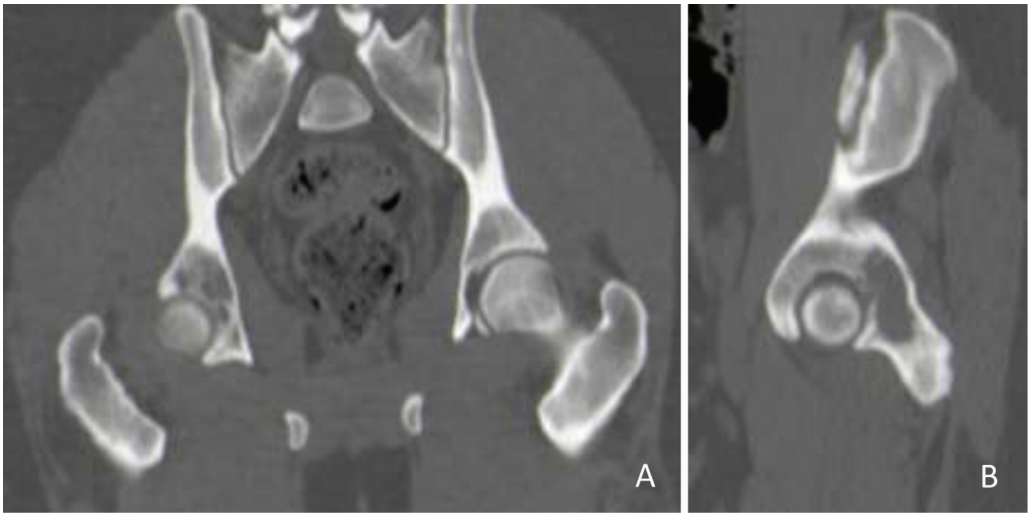

Fig. 2 Computed tomography on admission showing an osteolytic lesion of the right posterior acetabular rim with cortical disruption. Coronal (**A**) and sagittal (**B**) views.

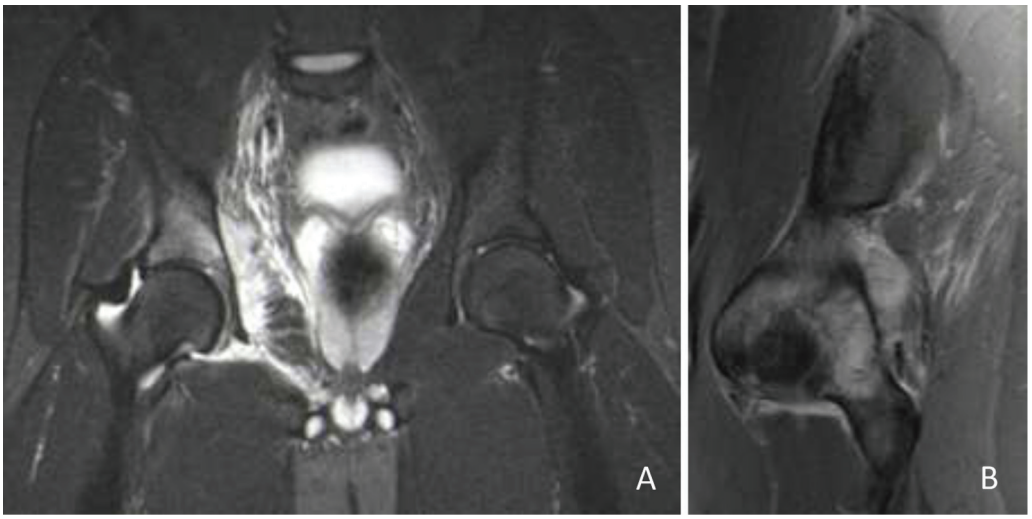

Fig. 3 Magnetic resonance imaging showing an associated soft tissue mass. Coronal (**A**) and sagittal (**B**) views.

(104 mg/L). Hemoglobin and erythrocyte sedimentation rate were normal. Radiographs were inconclusive (►**Fig. 4A**). Computed tomography and MRI findings were consistent with an inflammatory lesion involving the right ischiopubic ramus and obturator muscles, with an associated abscess, making the diagnosis of osteomyelitis most likely (►**Figs. 4B** and **5A**). Due to fever onset and increasing inflammatory markers, empiric antibiotic therapy was started (intravenous ceftriaxone, 2 g/day). Prior to the antibiotic therapy, blood cultures were collected and a percutaneous drainage was performed, but no agent was isolated. The patient responded well to the antibiotic and anti-inflammatory treatment with fever and pain resolution. Following 15 days of treatment, there was a considerable decrease on white cell count (11.5×10^9 cells/L) and C-reactive-protein (18.0 mg/L). However, an MRI showed persistency of the lesion (►**Fig. 5B**). The patient was submitted to an excisional biopsy. The histopathological examination was consistent with ES, and was later confirmed by immunochemistry and cytogenetics. The patient was transferred to a specialized center.

Discussion

Ewing sarcoma may raise suspicion for malignancy in young adults when it involves long bones in a typical location with associated pain and palpable soft tissue mass.[5] In the pelvis, the latter is difficult and, therefore, constitutional symptoms should not be overlooked. On the other hand, EG, a benign tumor-like condition, commonly affects flat bones. Radiologically, acute phase EG shows osteolysis with poorly defined margins similar to ES. On MRI, bone marrow involvement and an associated soft tissue component may also be present. Since imagiological features are similar, lesion biopsy is indispensable for the correct diagnosis.[9] In case 1, samples collected by percutaneous biopsy were not representative, leading to the misdiagnosis of EG. Therefore, when clinical and imagiological features are consistent with ES, a negative percutaneous biopsy should not rule it out, and an excisional biopsy should be performed. To minimize diagnostic errors and contamination of biopsy pathways, biopsies should be performed by surgeons with expertise in bone tumors.[10]

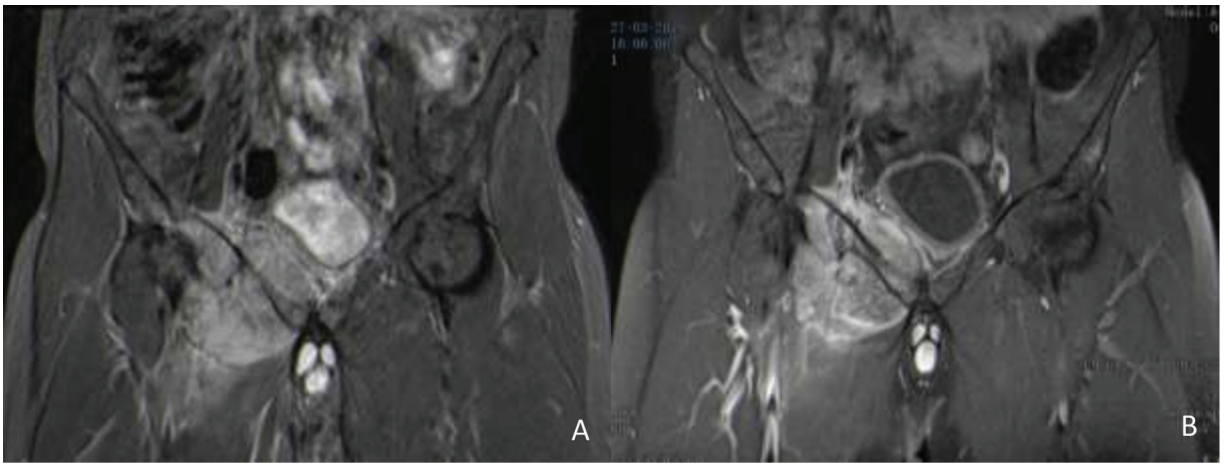

Fig. 4 Inconclusive radiograph on admission (**A**) and computed tomography scan showing enlargement of the right obturator muscles (**B**).

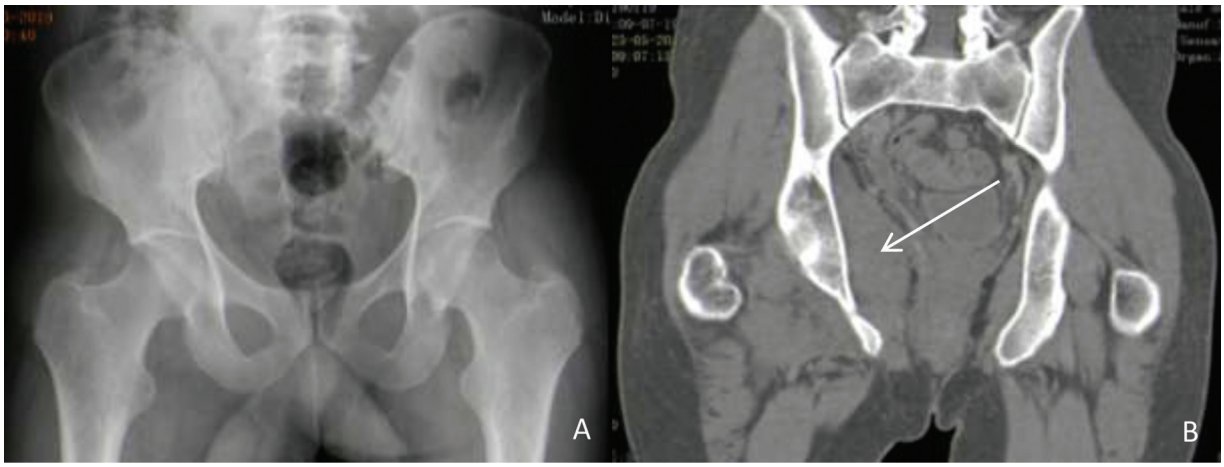

Fig. 5 Initial magnetic resonance imaging showing a lesion involving the right ischiopubic ramus and obturator muscles (**A**) and magnetic resonance imaging following antibiotic treatment showing persistency of the lesion (**B**).

In case 2, the presentation was atypical. The age, previous medical history, imagiological and biochemical findings were consistent with osteomyelitis. The sterility of the biopsy and blood cultures did not exclude its diagnosis, since as many as 26% of cases of osteomyelitis are diagnosed without culture confirmation and are supported retrospectively by their resolution following antibiotic therapy.[4] Moreover, our patient responded to the antibiotic and anti-inflammatory treatment. This response may be explained by the association of elevated levels of arachidonic acid metabolites and prostaglandins to malignancies. Inhibition of the arachidonic acid pathway by antibiotic and nonsteroidal anti-inflammatory agents may have modulated the clinical symptoms and tumor growth.[4]

Conclusions

The diagnosis of ES should not be overlooked in the assessment of pelvic lesions, and exclusion diagnosis should be the rule. If clinical and imagiological features are consistent with its diagnosis, a negative percutaneous biopsy should not rule it out, and an excisional biopsy should be performed. If clinical and imagiological features are consistent with osteomyelitis, an initial response to antibiotic and anti-inflammatory therapy does not allow its exclusion, with watchful waiting being recommended.

Financial Support
There was no financial support from public, commercial or non-profit sources.

Conflict of Interests
The authors have no conflict of interests to declare.

References

1 Halwai MA, Mir BA, Wani MM, Bashir A, Hussain A. Ewing's sarcoma of the ilium mimicking inflammatory arthritis of the hip: a case report. Cases J 2009;2:6487
2 McCarville MB, Chen JY, Coleman JL, et al. Distinguishing osteomyelitis from Ewing sarcoma on radiography and MRI. AJR Am J Roentgenol 2015;205(03):640–650
3 Kasalak Ö, Overbosch J, Adams HJ, et al. Diagnostic value of MRI signs in differentiating Ewing sarcoma from osteomyelitis. Acta Radiol 2019;60(02):204–212
4 Tow BP, Tan MH. Delayed diagnosis of Ewing's sarcoma of the right humerus initially treated as chronic osteomyelitis: a case report. J Orthop Surg (Hong Kong) 2005;13(01):88–92
5 Flores M, Caram A, Derrick E, Reith JD, Bancroft L, Scherer K. Ewing sarcoma of the pelvis with an atypical radiographic appearance: a mimicker of non-malignant etiologies. Cureus 2016;8(09):e787
6 Natarajan MV, Sameer MM, Bose JC, Dheep K. Surgical management of pelvic Ewing's sarcoma. Indian J Orthop 2010;44(04): 397–401
7 Ray P, Girach J, Sanghrajka AP. Ewing's sarcoma of the pelvis: an unusual, but not to be missed, cause of an irritable hip. BMJ Case Rep 2016;2016:bcr2015213782
8 Alshaya OS, Abbasher MI, Wani MM. Ewing's sarcoma of ilium presenting as right lower quadrant pain. Case Rep Orthop 2015; 2015:629601
9 Ando A, Hatori M, Hosaka M, Hagiwara Y, Kita A, Itoi E. Eosinophilic granuloma arising from the pelvis in children: A report of three cases. Ups J Med Sci 2008;113(02):209–216
10 Garcia JG, Marques DS, Viola DCM, Petrilli MT, Alves MTS, Jesus-Garcia Filho R. Biopsy Path Contamination in Primary Bone Sarcomas. Rev Bras Ortop (Sao Paulo) 2019;54(01):33–36