

CASE REPORT

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Unusual case of retrorectal tumor: A case report and review of the literature

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ABSTRACT

Ewing's sarcoma is a variety of sarcoma of reticuloendothelial origin. There are atypical forms of Ewing's sarcoma, encountered in 20% of cases, where the tumor grows in the soft tissue without any skeletal connection. The imaging of extra-skeletal Ewing's sarcoma (EES) has been revolutionized by the advent of modern techniques with magnetic resonance imaging (MRI) at the forefront. Magnetic resonance imaging is an essential aid for determining target volumes for radiotherapy and for monitoring patients during chemotherapy to assess its effectiveness and for the diagnosis of local recurrences. The diagnosis is mainly based on cytogenetics and immunohistochemistry. Their prognosis is a priori poor, with the possibility of local recurrence or late metastases. We report a particular case of EES located at the retrorectal space in a 15-year-old girl revealed by gluteal pain.

Keywords: Ewing's sarcoma, Extra skeletal, MRI, Retrorectal

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INTRODUCTION

Extra-skeletal Ewing's sarcoma is a rare pathology with a sadly poor prognosis, which affects young adults [1]. Its diagnosis is difficult even on the anatomopathological level. It is currently considered the most undifferentiated side of peripheral neuroectodermal tumors (PNETs) [1].

However, knowing how to evoke it in front of a retrorectal tumor is essential because it points toward a systematic biopsy and can modify therapeutic management in order to offer the best chances of survival. We report a case of EES localized in the retrorectal space.

CASE REPORT

A 15-year-old healthy girl presented with a four week history of back pain without radiation, treated with general and local analgesics. Despite the use of non-steroidal anti-inflammatory medications and physical therapy, the pain had become unbearable with a deterioration of the general state and a 4 kg weight loss in two weeks.

The clinical examination revealed two painful gluteal swellings, on either side of the intergluteal cleft, with no inflammatory signs, movable relative to the superficial plane and adhering to the deep plane, measuring approximately 5 cm from the long axis in the left and 9 cm long in the right.

The two masses converged in depth toward the intergluteal cleft. Rectal examination found a bulging mass on the posterior surface of the rectum. Besides, the abdominal, neurological, and lymph node examinations were unremarkable.

The biological check-up was normal, and serum alpha-fetoprotein (AFP) and beta-HCG levels measured respectively 0.5 ng/mL and 0.24 UI/L. Initial ultrasound imaging showed a retrorectal space mass, well limited, oval, with heterogeneous echogenicity, containing tissue

and cystic areas and a calcification, measuring 101×63 mm (Figure 1).

The computed tomography (CT) scan revealed a medial retrorectal mass extended to the perineal region, roughly oval, with cystic, calcium, and soft tissue components, measuring 98×97×83 mm, pushing forward the rectum and the anal canal. This mass included the 4th and 5th sacral vertebrae and the coccyx without bone lysis, with a doubt on a periosteal reaction of the coccyx (Figure 2).

Additional evaluation with MRI of the pelvis revealed a complex pre-coccygeal mass with lobulated contours, an heterogeneous signal on the T1 and T2 sequences, a hyper-signal diffusion, containing necrotic, hemorrhagic areas, and calcification, measuring 112×91×102 mm. Anteriorly, this mass infiltrates the rectum with no clear visible interface, behind and below it infiltrates the gemelli muscles and the gluteus maximus. Laterally, it infiltrates the ischiorectal fossa and comes in contact with the obturator internus muscles without a clear fatty border of separation (Figure 3).

Thus, we concluded to an aggressive tumor lesion centered on the coccyx with rectal infiltration. At surgical exploration, the tumor was in close contact with gluteal muscles and no cleavage planes were encountered. The decision was to do a biopsy and postpone surgical excision.

The biopsy found a massive tumor proliferation with round cells and the immunohistochemical markers were negative for CD56, leukocyte common antigen (LCA), synaptophysin, desmin, myogenin, and chromogranin and positive for CD99 and vimentin, which corresponds to an EES (Figure 4).

The initial staging work-up revealed the presence of pulmonary metastases, and chemotherapy protocol was started. After five sessions of chemotherapy, the clinical condition of the patient improved markedly and ultrasound control showed a reduction in tumor volume. Surgery is scheduled after completion of chemotherapy treatment.

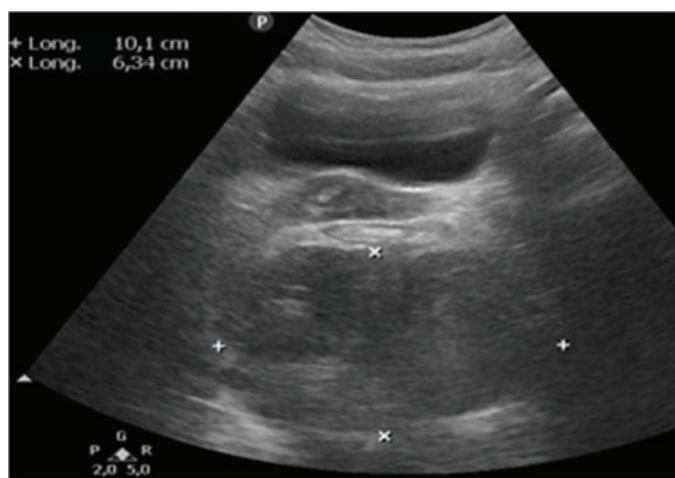


Figure 1: Pelvic ultrasound shows a mass of the retrorectal space, well limited, oval, of heterogeneous echogenicity, containing tissue and cystic areas and a calcification, measuring 101×63 mm.

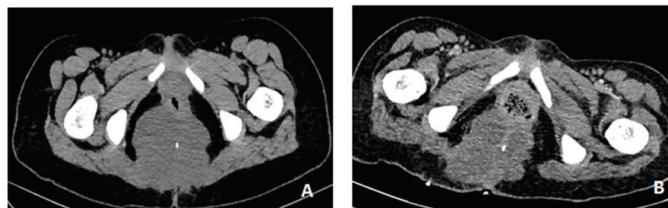


Figure 2: Pelvic CT axial view with (B) and without contrast (A): Retrorectal mass with cystic, calcium, and soft tissue components, extended to the perineal region, pushing forward the rectum and the anal canal.

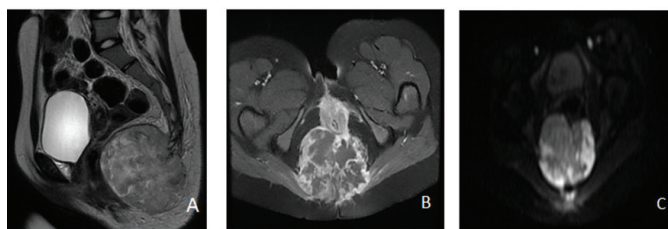


Figure 3: MRI of the pelvis: Sagittal T2 (A), axial T1 with gadolinium (B), axial diffusion (C). A complex retrorectal mass with lobulated contours, of heterogeneous signal on the T1 and T2 sequences, in hyper-signal diffusion, containing necrotic, hemorrhagic areas, and calcification, measuring 112×91×102 mm (H×AP×T). Infiltrating the rectum, the gemelli muscles, the gluteus maximus, the ischiorectal fossa and comes in contact with the obturator internus muscles without a clear fatty border of separation.

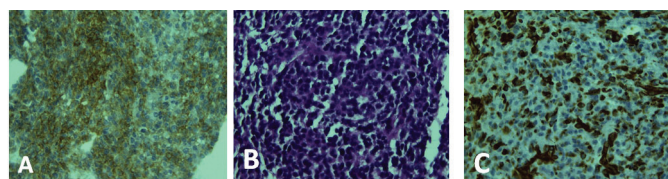


Figure 4: Histologic and immunohistochemical features of tissue biopsy. (A) The tumor cells show positive and strong membranous immunostaining for CD99. (B) HE staining showing proliferation of undifferentiated round cells. (C) Intense vimentin staining.

DISCUSSION

Extra-skeletal Ewing's sarcoma is a rare pathology, which affects young adults, difficult to diagnose even on the anatomopathological level [2]. Unlike the osseous form there is no apparent sex predilection, and patients are older with a median age of about 20 years [3]. The most frequent primary tumor sites were the extremities, followed by the pelvis [4].

Pain occurs only in 1/3 of cases, by involvement of the peripheral nerves or bone marrow with the possibility of motor and sensory disturbances or by spontaneous hemorrhage [5]. Remote metastatic extension is hematogenous, and mainly affects the lung, 30% of patients with soft tissue sarcomas have lung metastases [6]. Therefore, CT examination of the lungs should be part of the initial staging procedure provided metastatic disease has not already been revealed by conventional radiography [3].

In standard imaging, the lesions are often non-specific; it appears as a well-limited tumor, polylobulated, with the appearance of a pseudo-capsule, rarely, the tumor may contain calcifications [7].

The ultrasound associated with the color flow Doppler is a simple examination, of first intention, in front of any discovery of a mass of the soft tissue. The tumor often appears as a well-circumscribed mass, hypoechoic, and well vascularized. Ultrasound examination is sensitive but non-specific to make the diagnosis. The danger of ultrasound is to give a wrong diagnosis, misunderstanding a heterogeneous, necrotic sarcoma, with a hematoma, which can lead to a misdiagnosis or lead to inappropriate management. It also allows performing diagnostic procedures, such as ultrasound guided biopsy [3].

Computed tomography is still used because of its superiority over MRI in the analysis of bone and cartilage structures. Its limits lie in the poor precision of tumor limits and in soft tissue artefact. Computed tomography angiography is very useful in the analysis of vascular structures in the vicinity of the tumor. The tumor is hypodense compared to the muscles, rarely contains calcifications, often hyper vascular, therefore hyperdense after injection of the contrast medium. Computed tomography remains primarily indicated in the staging work-up to determine pulmonary involvement, in addition to the chest X-ray [3, 8].

Magnetic resonance imaging allows an exhaustive work-up for the lesion and therapeutic follow-up. It is an essential aid for determining target volumes for radiotherapy and for monitoring patients during chemotherapy to assess its effectiveness and to diagnose local recurrences [5].

However, spontaneous tumor hemorrhage may alter the appearances with any of these imaging modalities [3].

The positron emission tomography place (PET) has not been demonstrated yet; it cannot be considered as a reliable technique for determining the malignancy of a sarcomatous tumor due to the ambiguous relationship of the fixation of the tracer.

The morphological assessment is essential but the diagnosis of certainty is provided by the anatomopathological examination [2]. Histological diagnosis by biopsy is essential before continuing treatment especially before considering any surgical procedure for excision [7, 9]. Differential diagnosis is sometimes difficult and depends essentially on the age of the patient and the location of the tumor [2, 3].

In our patient, differential diagnosis is essentially with the other tumors arising in the retrorectal space, namely the sacrococcygeal teratoma in its pure pre-sacral form called type 4, often diagnosed late with malignant potential, which has been ruled out in front of a low AFP level.

Moreover, pelvic localization of neuroblastoma remains rare and represents between 2% and 5% of all neuroblastoma localizations. About 95% of cases are diagnosed in the first decade of life [3].

On the other hand, primary involvement of the retrorectal space with rhabdomyosarcoma is uncommon. It is more often a secondary extension. The first peak of incidence is between 2 and 6 years of age, and the second between 14 and 18 years of age [3].

Rhabdomyosarcoma and neuroblastoma were eliminated by immunohistochemistry. Additionally, the chordoma, which is exceptional in children, has been eliminated because of the absence of sacral lysis on CT and the absence of intracanal extension on MRI.

The management of EES must be done in a reference center and involves a multidisciplinary committee. It combines surgery, polychemotherapy and radiotherapy [6].

Surgery is first in small, easily resectable tumors, preceded by biopsy and chemotherapy in other cases [6]. The cytogenetic similarity of PNETs and soft tissue Ewing's sarcomas includes them in the same therapeutic protocol [7]. Survival rates with EES are still poor and both local recurrence and distant metastases develop frequently [3].

Patients generally have a poor prognosis with possibility of local recurrence or late metastases (delay of 3 months to 2 years). We found that patients with localized EES have an unfavorable prognosis prior to two years from initial diagnosis, but then the outcomes for EES are significantly better [10].

CONCLUSION

Extra-skeletal Ewing's sarcoma is a rare tumor with a sad prognosis. Differential diagnosis is sometimes difficult and depends essentially on the age of the patient and the location of the tumor. A biopsy is necessary to confirm the diagnosis.

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Author Contributions

Amalik Sanae – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Imrani Kaoutar – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Allali Nazik – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor of Submission

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Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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