

Ewing's Sarcoma: An Unexpected Case

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Abstract

Ewing's Sarcoma represents 6% of all malignant bone tumours, with an annual incidence of 1/1000000 people. It is a characteristic since the childhood until the 3rd decade of life, to affect primarily the long bones, pelvis, and possibly the soft tissues. Recognizing this fact, it's essential to value certain recurrent and in specific symptoms.

Keywords: Bone tumor; Ewing sarcoma; B-cell lymphoma

Background and Aim

Currently, the malignant neoplasms assume the 1st cause of child mortality. Recognizing this fact, it is essential for the practitioner to value certain recurrent symptoms. The Ewing's Sarcoma (ES) is an undifferentiated neoplasm that is part of the Ewing's family of Tumours, derived from embryonic cells that migrated from the neural crest. This represents 6% of all malignant bone tumours, with an annual incidence of 1/1000000 people. It is a characteristic since the childhood until the 3rd decade of life, to affect primarily the long bones and the pelvis, and possibly the soft tissues [1-3].

Methods

Interview/ Family evaluation with patient.

Results

Female, 24-year-old, Caucasian, single. No smoking, alcohol or drug abuse. Up-to-date vaccination. No sexual activity. Irrelevant personal and family history. Visited the family doctor on the 07/05/2014 with amenorrhea complaints lasting for 3 months and metrorrhagia in the preceding month. Mentioned asthenia, migratory myalgia and lost 8 kg in 3 months. Weight loss was justifiable by the onset of regular physical exercise. Had been admitted to the emergency room twice in the preceding month following pain in the left shoulder. Analgesics and anti-inflammatory drugs were prescribed with partial improvement.

The physical exam showed pain in the right iliac region. An analytical study was requested to evaluate blood count, renal, liver and thyroid performance, sedimentation rate, sex hormones dosing and gynaecological ultrasound. Returned to family doctor on 14/05/2014 with the results. Showed normocytic normochromic anaemia (Hb: 10.8 g/dL), sedimentation rate of 108 mm/h with no impaired renal, liver, thyroid or ovarian performance. Gynaecological ultrasound showed 'complex cyst of 4.8 cm in the left adnexal region, suggestive of hemorrhagic cyst (...) in the right iliac region, a heterogeneous mass can be identified, apparent retroperitoneal, with at least 7.7 cm of larger diameter (...)'. With the results described, an abdominal-pelvic CT was requested, as well as proteinogram and tumor biomarkers.

On the 20/05/2014 the patient came back with the results. CT showed a mass in the right iliac fossa with 11.6 × 9.3 × 5 cm, and another mass of 9.6 cm × 5.2 cm in the gluteal region, with slight erosion of the external cortical bone of the adjacent ilium. The patient was referred to a general surgery appointment with urgency. On the 28/05/2014 the patient had the first general surgery consultation where an abdominal-pelvic MRI, thorax CT, preoperative study and tumor biomarkers were requested.

The exams showed extensive neoplasia of about 140 mm × 75 mm in the right iliac region, multiple lytic lesions and pleural injury of

about 42 mm × 52 mm. After biopsy and cytogenetic study, Ewing's Sarcoma was diagnosed. Began chemotherapy on the 11/07/2014 with cyclophosphamide, vincristine and doxorubicin. Unfortunately the chemotherapy wasn't effective and the patient died on 05/2015.

Conclusion

There is no specific cause, however, in 90% of this type of tumors there is specific translocation that requires the EWSR1 gene (22q12.2), especially the translocation t (11;22) [4]. Early metastization (10% to 35%) is seen at the time of diagnosis. The 5 year-survival rate is approximately 75% if the disease is localized. In the case of disseminated disease, the prognosis is poor, with survival rates of 25%. It's extremely rare in the Asian and Afro-descendent populations, with a slight predominance in males [5]. Early symptoms are usually tumoration or bone pain in the areas compromised by the tumor. Other symptoms such as fever or local heat can also happen, rendering differential diagnosis difficult due to osteomyelitis. Other differential diagnoses that include small rounded-cell tumors are neuroblastoma, rhabdomyosarcoma, Hodgkin's lymphoma and non-mesenchymal chondrosarcoma [6]. The treatment comprises three branches: chemotherapy, locoregional surgery if possible, and radiotherapy [7].

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