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Ewing's Sarcoma of the Orbit: About a Case

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Abstract

Introduction:
Ewing's sarcoma is seen primarily in children and young adults, more common in boys. Tumors developed from the bones of the face represent less than 5% of all cases of Ewing's sarcoma.

Observation:
A 16-year-old patient with no notable pathological history, who consulted for headache, a binocular diplopia.

Radiological exploration by magnetic resonance imaging objective a lesional process centered on the large wing of the sphenoid and the upper wall of the right orbit, the bony origin is primarily a sarcoma.

The patient received a biopsy, which pathological examination with immunohistochemistry confirmed the diagnosis of orbital Ewing sarcoma.
The interest of this file lies in the unusual location of the tumor.

Conclusion:
Ewing's sarcoma requires management depending on the different locations.
Curative radiotherapy is widely used in this location but cannot compensate for limited surgery.

Keywords: Ewing-like sarcoma, EWS rearrangement, Chemotherapy, radiotherapy

Introduction

Ewing's sarcoma is a rare bone tumor corresponding to the undifferentiated form of primary and peripheral neuroectodermal tumors. It is the second most common malignant bone tumor after osteosarcoma in adolescents and young adults and affects around 80 to 100 new patients per year in France. The majority of cases (70%) occur in young people aged 5 to 25 years. However, these tumors can occur before 5 years and after 30 years. The preferred sites are the long bones and pelvis. The locations in the head and neck are exceptional (2 to 6% of cases).

Their low incidence and the difficulty of accessing this anatomical region pose diagnostic and therapeutic problems.

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This group of tumors, despite their chemosensitivity and radiosensitivity, remains of poor prognosis with particular histopathological and molecular criteria, the specific translocation EWS-Fli1 [1]. There are no randomized studies or published series in the literature dealing with orbital PNET.

Patient and Observation

This was a 16-year-old patient with no significant family history. Having presented two months before the consultation with progressive headaches and a binocular diplopia reason for his consultation for management

Initial WHO clinical examination 1, Glasgow score of 15 complaining of intense headache, binocular diplopia with slight exophthalmos, no feeling-motor deficit

A cerebral computed tomography objective a tumor process of the right orbital apex invading the optic canal and the superior orbital fissure, the appearance of which suggests a primary or secondary lesion (Figure 1) supplemented by magnetic resonance imaging making it a extraaxiallesional process centered on the large wing of the right sphenoid and on the upper wall of the orbit 74x48x37 mm in transverse diameter, anteroposterior and in height respectively, filling in front the optic hole, invading the optic nerve, ophthalmic artery and the upper right and upper oblique oculomotor muscles, pushing back the frontal parenchyma as well as the right frontal horn with a mass effect on the neighboring parenchyma and discreet peri-lesional edema.

Extension assessment, no distance anomaly.

The patient underwent a biopsy, the anatomopathological study of which had revealed a tumor proliferation arranged in a diffuse sheet. It is made up of small, round cells with a rounded nucleus, fine chromatin, and a scanty basophilic cytoplasm. Figures of mitosis are noted. Presence of foci of tumor necrosis (Figure 2)

In immunohistochemical studies, tumor cells express CD99 (intense and diffuse membrane labeling) and synaptophysin. They do not express neurofilament, CD45, desmin, or GFAP.

Molecular complement (FISH) was requested: absence of EWS rearrangement.

Given the histological description oriented towards an Ewing sarcoma, given the absence of EWS rearrangement, it was necessary to complete with molecular biology that we do not have to confirm whether it is Ewing Like sarcoma in our center.

We retained the diagnosis of Ewing's sarcoma.

The patient received neoadjuvant chemotherapy with a protocol: VAC IE (Vincristine 2 mg total dose, Doxorubicin 75 mg / m2, Cyclosphosphamide 1200 mg / m2, Ifosfamide 1800 mg / m2 and Etoposide 100 mg / m2 x 5 days).

He benefited from 3 cycles

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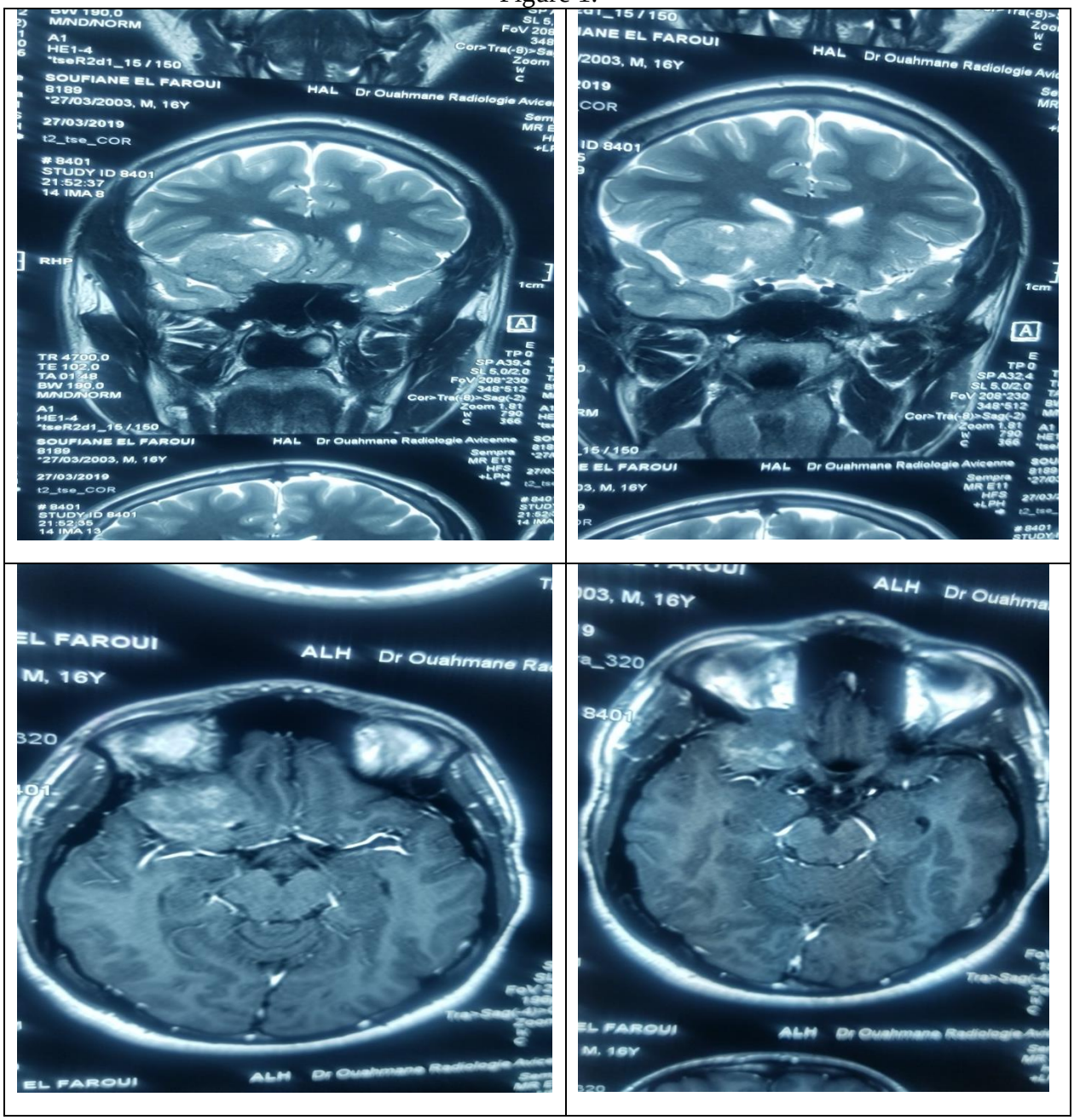
Evaluation Craniofacial MRI showed a decrease in size of the tumor process of the locally advanced skull base keeping the same ratio (Figure 3)

Surgery discussed in the SPC but given the inaccessibility of the tumor, he received curative radiotherapy followed by adjuvant chemotherapy.

Figure 1.



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Discussion

It was in 1921 that James Ewing first described the tumor that will bear his name to this day, distinguishing it from lymphomas and other types of cancer known at that time. The term "sarcoma" is in fact improper, it is a primary neuroectodermal tumor (primary neuroectodermal tumor or pNET) [2]. It represents common characteristics with this group of tumors in 88% of cases. These are phenotypic (small round cell tumors), genetic (t (11; 22)) [3] and immunohistochemical similarities (diffuse membrane labeling by the anti-CD99 antibody present in more than 90% of cases). . It is the most common primary malignant neoplasm of the bone in young subjects after osteosarcoma. Its peak incidence is in the second decade with a median age of 14 years. 90% of affected patients are less than 20 years old. The frequency of the disease is 1 to 3 per million per year with a slight male predominance. It is six times more common in Caucasian children than black children. No risk factor has been identified. The most commonly found pathophysiological mechanism of Ewing's sarcoma is a t (11; 22) [1] translocation responsible for the appearance of a fusion gene called EWS-FLIC. This abnormality is found in 85% of these tumors. In 10 to 15%, it is a t (21; 22) translocation giving rise to the synthesis of an EWS-Erg fusion gene [4]. In the remaining 1 to 5% of cases, one or more several translocations are possible. In all cases, an abnormal protein is produced, resulting in continued activation of the membrane receptor IgF-1, responsible for cell proliferation. The tumor can grow anywhere in the body, but it is mainly in the long bones (47 %) and pelvis (29%) that they appear. Some flat bones (ribs) and the spine are also commonly affected. There is a predilection for diaphyseal location, unlike osteosarcoma which is more frequently metaphyseal.

Orbital localization remains rare, with little specific clinical signs [5], only about thirty cases have been reported in the literature over the past fifty years.

The surgical difficulty and especially extensive surgery means that irradiation by conventional radiotherapy is often the only curative treatment as reported by numerous studies [6]

Our patient benefited from chemotherapy associated with radiotherapy with good progress. The most significant prognostic factor is the stage of the disease. Metastases and recurrences have a very poor prognosis [7].

Conclusion:

Ewing's sarcoma requires management depending on the different locations. Particular problems associated with orbital Ewing sarcomas are the difficulty of performing a very large surgery, yet a major prognostic element. Curative radiotherapy is widely used in this location but cannot compensate for limited surgery. The efficacy of chemotherapy in terms of survival has not been demonstrated in these locations where the major evolutionary risk remains local.

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Reference

Machado I, Noguera R, Pellin A, Lopez-Guerrero JA, Piqueras M, Navarro S, et al. Molecular diagnosis of Ewing sarcoma family of tumors: a comparative analysis of 560 cases with FISH and RT-PCR. Diagn MolPathol 2009; 18: 189-99.

Khoury JD. Ewing sarcoma family of tumors. AdvAnatPathol2005; 12: 212-20.

Turc-Carel C, Aurias A, Mugneret F, Lizard S, Sidaner I, Volk C, et al. Chromosomes in Ewing Sarcoma. I. An evaluation of 85 cases of remarkables consistency of t (11; 22) (q24; q12) / Cancer gent Cytognet 1988; 32: 229-38.

Kim HS, Kim S, Min YD, Kee KH, Hong R. Ewing´s Sarcoma of the stomach; rare case of Ewing´s sarcoma and suggestion of new treatment strategy. Journal of Gastric Cancer. 2012 0Dec; 12 (4): 258-261. PubMed | Google Scholar

Becroft DM, Pearson A, Shaw RL, Zwi LJ. Chromosome translocation in extraskeletal Ewing's tumor. Lancet. 1984 Aug 18; 2 (8399): 400. PubMed | Google Scholar

Ewing's sarcoma of the orbit. About a case and review of the literature M. Martani∗, A. Mansour, M. Bouaziz CHU d'Annaba, Annaba, Algeria. Scince direct, Pubmed / Google Scholar

Sabah N, Francis B, Valère D, Fatima S. Extraskeletal Ewing sarcoma at rare sites with review of literature. Annals of Pathology and Laboratory Medicine. 2017; 4 (1). Google Scholar