Exceptional localization of Primary Ewing's sarcoma in the greater wing of the sphenoid bone

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Abstract

Ewing’s sarcoma is a primary bone tumor of high grade of malignancy. Locations in the head and neck are rare. We report the case of a patient with Ewing’s sarcoma of the greater wing of sphenoid with intra orbital and intracranial extension revealed by a diplopia. A stereotactic biopsy was done. The patient received radio-chemotherapy with good outcomes. Thanks to multidisciplinary approach the prognosis has been well improved.

Introduction

Ewing’s sarcoma is a rare bone tumor. Before the age of 20, it is the second malignant bone tumor in order of frequency (30%) behind osteosarcoma (60%) [1]. The sites of predilection are the long bones and the pelvis. Locations in the head and neck are exceptional (2 to 6% of cases) [1,2].

The diagnostic and therapeutic are difficult because of their low incidence and the difficulties of access to this anatomical region.

Case report

An 18-year-old patient, with an unremarkable medical history presented headaches associated with diplopia evolving for two months. The ophthalmological and central nervous system examination did not reveal any anomaly.

The patient underwent a cerebral CT-Scan which objectified a tumoral process centered on the right orbital apex measuring 26*17mm with an intra-orbital extension.

Cerebral magnetic resonance imaging (MRI) revealed a process centered on the greater wing of the right sphenoid and on the superior wall of the orbit. This process fills the optic hole anteriorly, invading the optic nerve and ophthalmic artery.

*Key Words:
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Figure 1: Cerebral CT scan showing an osteolytic tumoral process centered on the large wing of the right sphenoid invading the orbital apex.
A stereotactic biopsy of the tumor was performed objectifying a tumoral proliferation arranged in diffused sheet with areas of necrosis. It is made up of small rounded cells, with fine chromatin, and sparse basophilic cytoplasm. Figures of mitosis are noted. On immunohistochemical study, tumor cells express CD99 (intense and diffuse membrane labeling) and synaptophysin. They do not express the neurofilament, CD45 (lymphoid marker), desmin (muscle marker), or GFAP (glial marker). Based on these findings, the diagnosis of Ewing’s sarcoma was made.

The tumor was judged inoperable because its anatomical location and its relationship with the vasculo-nervous elements.

Chemotherapy and radiotherapy were started and gave excellent results. The patient underwent a CT-scan after three months and the results were very encouraging.

**Discussion**

Ewing sarcoma of the head and neck region is unusual and constitutes about 4% of cases. The skull and the mandible are the most affected sites [3,4,5], less frequently the maxillary sinus [4,5], the ethmoid [6,7], the orbit [7,8] and petrous bone. Only a few cases of sphenoidal Ewing sarcoma have been reported in the literature.

The most common symptoms are headache, oculomotor paralysis, ptosis, diplopia. The optic nerve can be compressed and affect visual acuity [8,9].

MRI is very important for the diagnosis and the management of the tumor [11,12].

The best treatment of Ewing’s sarcoma is radical surgery followed by radiotherapy and systemic chemotherapy [3,8,10], but the indications for surgery must be carefully considered for each patient: age, localization, size and the resectability.

In general, the prognosis of Ewing’s sarcoma is poor. This is due to the fact that metastases occur early in the lungs, the bones and neighbouring tissues.

**Conclusion**

Ewing’s sarcoma is a malignant bone tumor in young people. The primary location of the sphenoid sinus is not common. The treatment combines surgery, chemotherapy and radiotherapy. The prognosis is poor but it has been considerably improved thanks to a multidisciplinary therapeutic approach.
References


