Sarcome d'Ewing du sinus sphénoïde avec extension orbitaire et intracrânienne : à propos d'un cas

Primary Ewing's sarcoma of the sphenoid sinus with orbital and intracranial extension: a case report

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RÉSUMÉ

Introduction: La tumeur d'Ewing est une tumeur osseuse primitive de haut grade de malignité appartenant au groupe des tumeurs neuro-ectodermiques. C'est la deuxième tumeur osseuse primitive de l'enfant après l'ostéosarcome représentant 4 à 10 %. Elle peut survenir sur tous les os du squelette. Cependant, la localisation au niveau du massif facial est rare (1 à 2%) et exceptionnelle au niveau du sinus sphénoïdal. Nous rapportons les résultats cliniques d'un cas rare de sarcome d'Ewing du sphénoïde avec extension intra orbitaire et intracrânienne.
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Mots-clés

Sarcome d'Ewing, sinus sphénoïdal, orbite, endocrâne, chimiothérapie

SUMMARY

Background : Ewing's sarcoma is a high-grade neuroectodermal primary bone tumor. This is the second primary bone tumor in children after osteosarcoma and represents 4 to10% of cases. It can occur in all skeletal bones. However, the location at the facial bones is uncommon (1 to2%) and extremely rare at the sphenoid sinus. We report the clinical results of a rare case of Ewing's sarcoma of the sphenoid with intraorbital and intracranial extension.

Key-words

Ewing's sarcoma, sphenoid sinus, orbit, intracranial, chemotherapy

Ewing's sarcoma is a high-grade neuroectodermal primary bone tumor (1). It represents 4 to 10% of all primary bone tumors (2). This is the second primary bone tumor of children after osteosarcoma. It can occur in all skeletal bones and can rarely arise from extra-skeletal sites. The location in facial bones is rare, representing 1 to 2% (3,4). In this region, it is the lower jaw that is most frequently involved (2,3). The location at the sphenoid sinus is rare.

CASE PRESENTATION

A previously healthy 4 years old girl presented with four months history of left proptosis and headache. Examination revealed an irreducible non-pulsating and painless left proptosis with an initially normal visual acuity (Fig.1). Nasal endoscopy showed a mass of the left middle turbinate area bleeding on touch.
Her routine hematological and biochemical investigations were within normal limits.

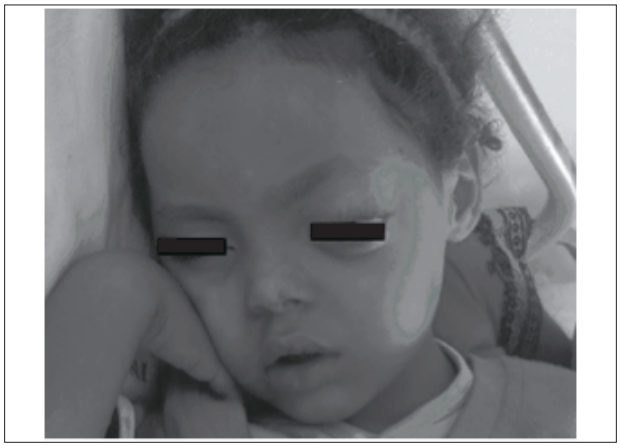

Figure 1 : left irreductible proptosis

Different MRI sequences with contrast revealed a soft tissue mass isointense on T1, heterogeneously intense on T2-weighted and heterogeneous enhancement following gadolinium administration. The mass was centered on the left sphenoid sinus with erosion of the basi-sphenoid (Fig.2) and sella and with eccentric extension into ethmoid cells, ipsilateral orbit, infratemporal fossa, the anterior cranial fossa, the maxillary sinus and the left nasal cavity (Fig.3, 4).
The diagnosis was established by biopsy, in which the tumor was seen as layers of small round cells, similar to lymphocytes, but larger. Mitotic cells were rare, intercellular stroma was scarce and a large portion of the tumor was necrotic. IHC study showed positivity CK and CD99.

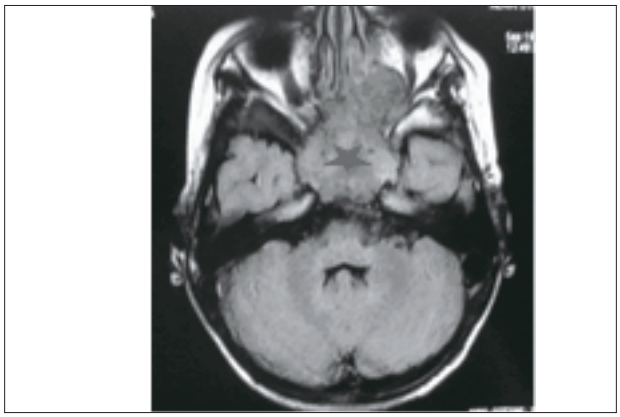

Figure 2 : MRI T1, axial sequence: isointense mass within sphenoid with involvment of ethmoid and left orbit

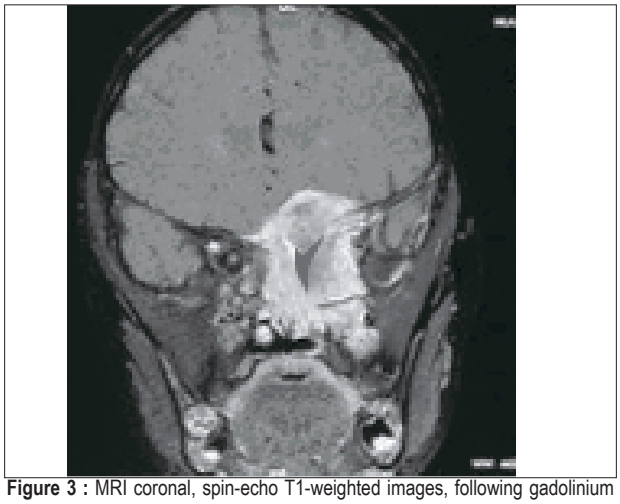

Figure 3 : MRI coronal, spin-echo T1-weighted images, following gadolinium administration: The sequences reveal an heterogeneous enhancement with intracranial and infra-temporal fossa extension of the tumor

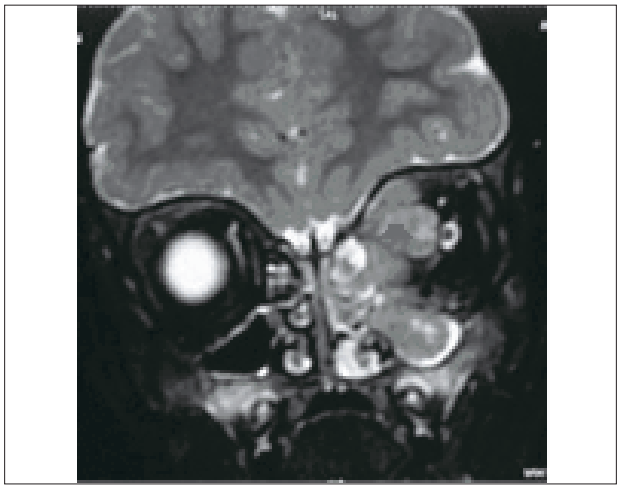

Figure 4 : MRI coronal spin echo T2 Fat Sat weighted images: The sequences reveal an orbital and left maxillary sinus extension of tumor

Given the extent of the tumor, surgery was not indicated and a chemotherapy was introduced using the protocol VIDE. (Vincristine, Isofosfamide, Doxorubicin, Etoposide) The tumor was extremely aggressive, with gradual loss of vision. The tumor did not respond to chemotherapy and the patient became comatose, due to massive intracranial extension (Fig 5, 6) and died after three cycles of chemotherapy.

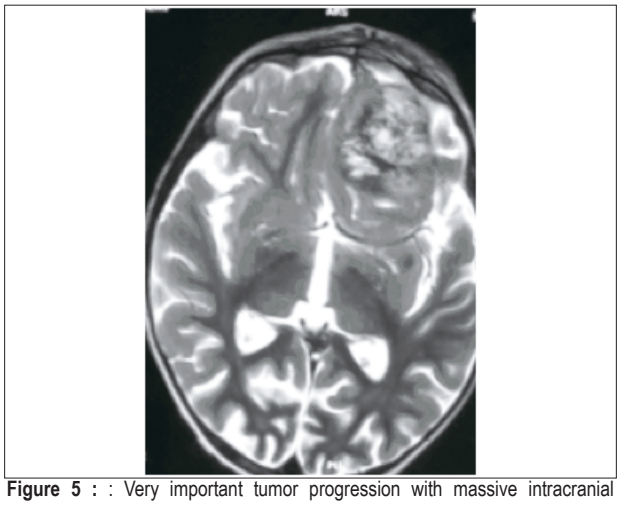

Figure 5 : : Very important tumor progression with massive intracranial extension under chemotherapy. MRI axial spin echo T2-weighted images

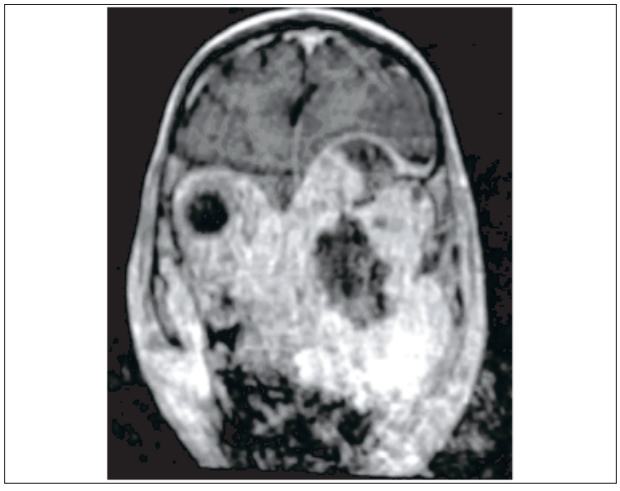

Figure 6 : Very important tumor progression with massive intracranial extension under chemotherapy. MRI axial, spin-echo T1-weighted images, following gadolinium administration

DISCUSSION

Ewing's sarcoma (ES) is a highly lethal round cell sarcoma, which was first described in 1921 by James Ewing as "diffuse bone endothelium." The cause is unknown, the origin cell is uncertain and even the potential of antigen expression is controversial (4,5). It is the second primary bone tumor of childhood after osteosarcoma (1, 4). It is a rare disease, representing approximately 10% of all primary bone tumors. It most frequently arises in the trunk and diaphysis of long bones with pelvis, ribs, femur and humerus being the predominant sites (1,2).

ES of the head and neck region is unusual and constitutes approximately 4% of cases, with primary ES originating from the paranasal sinuses being exceedingly rare (1,3,6). The skull and mandible are the most affected sites (1,2,4), less frequently the maxillary sinus (4,5), the ethmoid, (6,7), the orbit (7,8) and the petrous bone. Only a few cases of sphenoid Ewing's sarcoma have been reported in the literature (2,6,7).

The most common symptoms are headache, proptosis, oculomotor limitation, diplopia, fever and anemia (2,3). The optic nerve can be compressed and affect visual acuity (8,9).

Biologically we can observe an increased sedimentation rate and leukocytosis (3).

Most patients have a locally advanced tumor at diagnosis with a size greater than or equal to 10cm and/ or a metastatic extension (mainly lung and bone) in 10 to 15% of cases (9). The CT scan shows an osteolytic process with a soft tissue mass occupying the sinus cavity, destroying the bone wall and having a significant invasion of adjacent soft parts. This process is usually accompanied by a type of periosteal reaction spike "in sunshine", contrary to what is observed in long bones where it is rather multi lamellar type. The CT scan also makes it possible to appreciate the extension to other sinonasal cavities and the extra sinuses extension (1).

MRI is performed in addition to the CT scan to better assess tumor volume. It also allows better analysis of intracranial extension and neural involvement, as well as the assessment of response to chemotherapy (1).

ES is heterogeneous with hypo to iso intense in T1 weighted and iso to hyper intense in T2 weighted (1,2,10). Macroscopically, the tumor is often bulky, grayish containing areas of necrosis and hemorrhage (11). Pathological examination confirms the diagnosis (2,5,6). Microscopically there is a monomorphic proliferation of undifferentiated small round cells whose cytoplasm is slightly extended containing PAS positive glycogen granules (1,9). The nuclei are dense and hyper chromatic nucleoli. This poses the problem of differential diagnosis with other neuroectodermal tumors and other small round cell tumors such as neuroblastoma, lymphoma, osteosarcoma clear cell, the rhabdomyosarcoma and leukemia (1,3,6,11,12).

The immunohistochemical and cytogenetic study allow ask definitive diagnosis by showing a membrane marking MIC 2 (CD99) in 90% of cases, CK positivity in 25% of cases and translocation (11,22) d in 83% of cases

(1,4,9,11).
Appropriate treatment for Ewing's sarcoma is surgical excision of the tumor with radiotherapy and chemotherapy (1,3,5,8,10). The indications for surgery must be carefully examined for each patient: age, site, size and resectability of the tumor. By choosing chemotherapy, a combination of doxorubicin, cyclophosphamide, vincristine, actinomycin D, etoposide and ifosfamide has often been used.
The association of surgery, radiotherapy and chemotherapy has significantly improved the 5-year survival ratio, now reaching 40 to 75%. The most important prognostic indicators are the size and the primary site. Primary tumors of the head and neck, especially the mandible, have a significantly higher survival rate. Death is usually due to disseminated hematogenous spread (3,4, 11).

CONCLUSION

Ewing's sarcoma is a highly lethal round cell sarcoma. Location at the head and neck is rare representing 4%. Treatment must be prompt combining chemotherapy and radiotherapy or surgery in operable forms. The prognosis remains reserved, however, since these tumors are usually discovered late.

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