OSTEOSARCOMA OF THE SPHENO-TEMPORO-ORBITAL BONE: IMAGING ASPECTS OF SUCH UNUSUAL LOCATION

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ABSTRACT

Osteosarcoma of the spheno-temporo-orbital bone is a sporadic tumour. Despite the fact that primary osteogenic sarcomas are the most common bone neoplasm, their location in the skull bone is uncommon representing less than 2% of all skull tumours. We report the case of a 41-year-old woman, who has experienced periorbital pain with exophthalmos and left eye vision loss. Neuroimaging analysis including both brain computed tomography (CT-scan) and Magnetic resonance imaging (MRI) was performed. Although radiographic features of skull bone osteosarcomas are not specific, the combination of several radiographic features could lead to this diagnosis in such rare location. A trans-temporal biopsy established the final diagnosis with immunohistochemical study. Neurosurgical resection of a primary tumour was not possible given the critical extent of a tumour and the involvement of adjacent structures, so the patient underwent conformational radiotherapy. Local and metastatic progression marked the evolution. The patient received palliative chemotherapy and died a few months later. The purpose of presenting this case is not only to report an uncommon malignancy of the skull bone, but also to provide imaging aspects of this unusual location and to raise awareness among radiologists to consider this radiological entity as a differential diagnosis when a skull bone process is identified.

KEYWORDS: Osteosarcoma, skull bone, spheno-temporo-orbital bone, imaging aspects.

Introduction

Osteosarcoma of the spheno-temporo-orbital bone is a sporadic tumour, accounting for less than 2% of all skull tumours and only 5 to 6% of craniofacial bone tumours [1, 2]. Clinical presentation is non-specific, most often symptoms depend on tumour's location [1, 3, 4]. Although no radiographic finding is pathognomonic, the combination of several radiographic features could help evoking this diagnosis in such rare location [1, 4, 5]. Through this report, we are trying to describe imaging aspects and radiological features of this unique skull bone tumour.

Case report

A 41-year-old Moroccan female plained three months before medical consultation (on August 2015) from a periorbital and left upper jaw pain. Then, she experienced a rapidly progressing exophthalmos associated with a gradual left eye vision loss. Neuroimaging analysis including both brain computed tomography (CT-scan) and Magnetic resonance imaging (MRI) was performed. Head CT scan showed a hyperdense lesion, measuring 43x35 mm, holding retro-orbital left triangle with the partial erosion of the posterior wall of the left orbit and the upper outer

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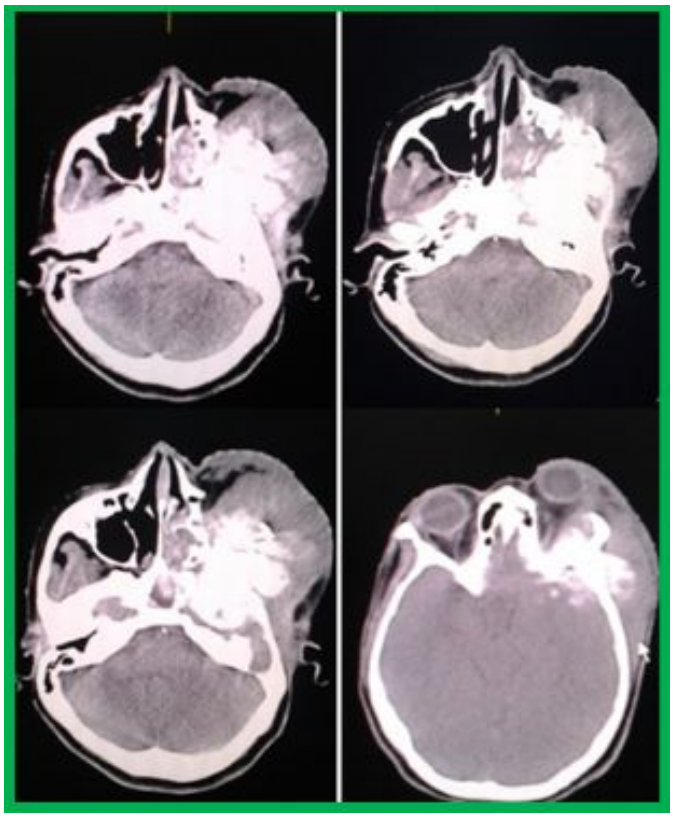

Fig.1. CT scan showing a hyperdense lesion, holding retro-orbital left triangle with partial erosion of the posterior wall of the left orbit and the upper outer wall of the left maxillary sinus with bone destruction of the left temporal bone.

wall of the left maxillary sinus with the bone destruction of the remaining temporal bone (figure 1). MRI demonstrated a het-

Table 1 A table summarizing care checklist organized into a timeline.

The event	Timeline
First symptoms	May 2015
First medical consultation	August, 23th 2015
Radiological investigations (CT - scan and MRI)	September 2015
Trans-temporal biopsie	October 2015
Radiotherapy	November 2015
Palliative chemotherapy (after progression)	February 2016

erogeneous signal mass containing hypointense areas of necrosis and hyperintense dotted calcifications. The process is focused on the more significant wing of the sphenoid with osteolysis of the left temporal bone and the inferior-lateral wall of the left orbit. An intracranial extension was noted. The mass involved the lateral pterygoid and the left temporalis muscles which were enlarged and heterogeneous (figure 2).

Despite being inconclusive, these radiological features and findings were suggestive for a locally advanced primary osteosarcoma of the skull bone. Taking into consideration that the

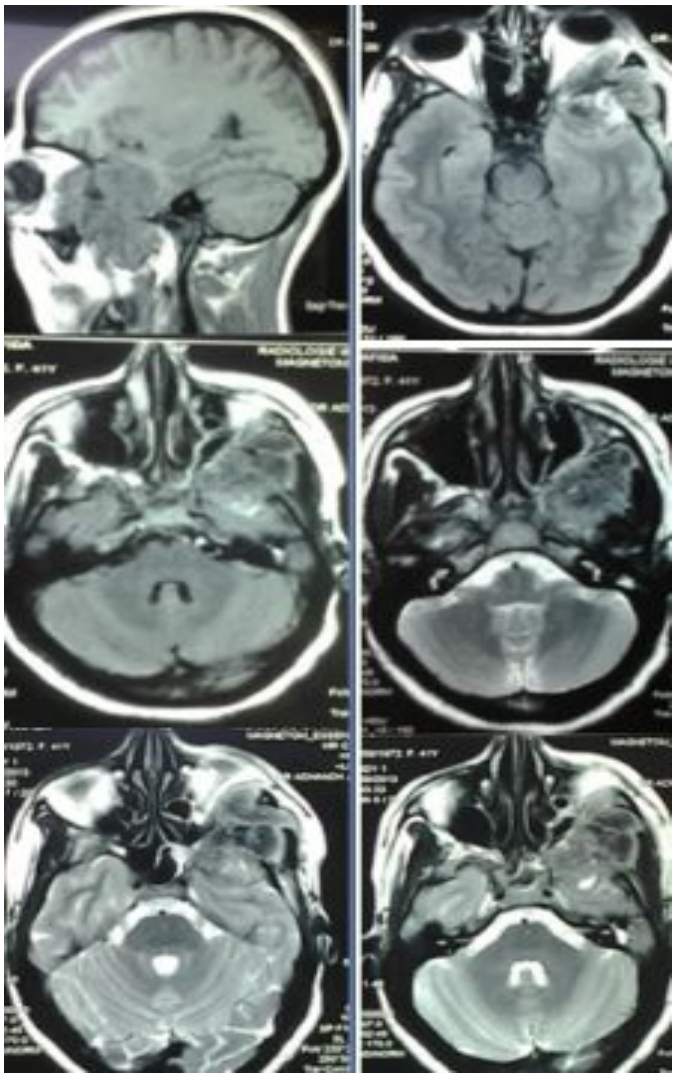

Fig.2. Magnetic resonance imaging pictures demonstrated an heterogeneous signal mass containing hypointense areas of necrosis and hyperintense dotted calcifications. The process is focused on the greater wing of the sphenoid with osteolysis of the left.

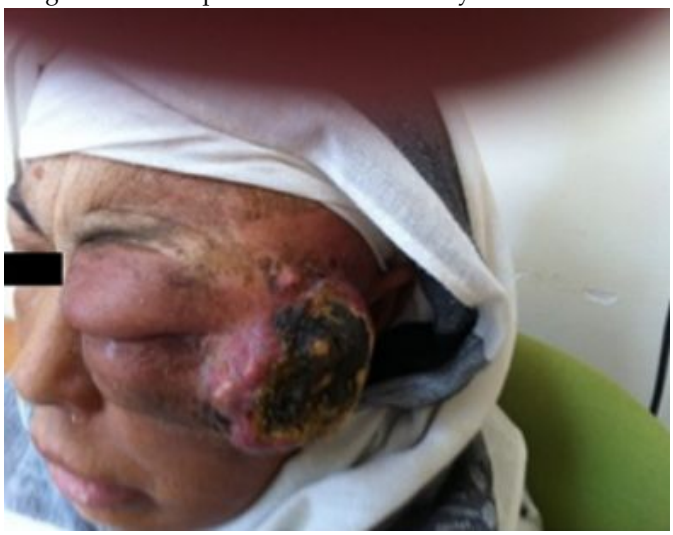

Fig.3. Photographic image showing clinical aspect of the tumor after local progression.

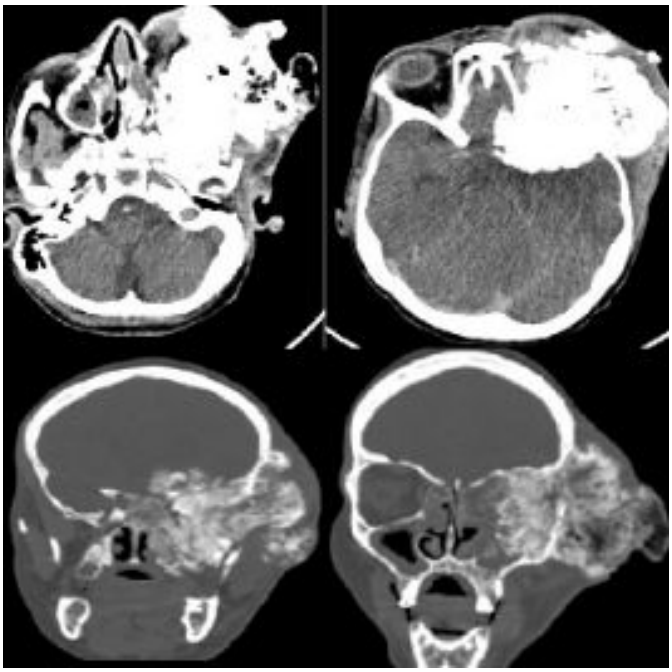

Fig.4. CT scan showing significant local tumor progression after failure of radiotherapy with intracranial involvement.

diagnosis of osteosarcoma is only histological, a trans-temporal biopsy with microscopic examination and immunohistochemical (IHC) study was performed (on October 2015), and the final diagnosis of osteogenic sarcoma was established.(Table 1) Neurosurgical resection of a primary tumour was not possible given the extent of a tumour and the involvement of adjacent structures including intracranial extension. The patient underwent an external conformational radiotherapy, targeting the lesion with a total dose of 66 Gy (2 Gy per fraction and five fractions per week started on November 2015). Evolution has been marked by local (figures 3 and 4) and metastatic progression three months later, in both pulmonary and bone level. The patient received palliative chemotherapy with ifosfamide and doxorubicin and died a few months later.

Discussion

We report a rare case of primary osteosarcoma, with a particular location in the spheno-temporo-orbital bone. Although osteosarcoma is the most common primary malignancy of bone, only 5 to 6% arise in the craniofacial extremity. Its location in the skull bone is even less common, accounting for less than 2% of all skull tumours [1, 2]. Moreover, the vast majority of craniofacial osteosarcomas are located in the zygomatic bone [6, 7].

Osteogenic sarcoma is more common in male than female [6, 7]. It is mostly a disease of adolescents and young adults, but more than 30% occurs in patients after 40 years of age with an average age of 34 years in one large series [4, 5, 8]. The presenting symptoms varied with the location of the tumours, common ones for locally advanced osteosarcoma are swelling, pain, or both. The most common finding in physical examination is a large and tender soft tissue mass [1, 3, 4].

Radiographically, the craniofacial osteosarcomas are infrequent and not extensively evaluated unlike those measuring long bones. Radiological investigations involve head CT-scan and MRI. For this atypical location, CT-scan provides an excellent detection of cortical erosion, tumour calcification and intramedullary as well as soft-tissue extension. Furthermore, CT-scan is more accurate than MRI in detecting the matrix calcifications and bone reaction or destruction [4]. However, MRI with T1 and T2 weighted images are more efficient than CT in demonstrating the intramedullary and extraosseous involvement. The T1 -weighted images are excellent for a clear and precise anatomic definition, whileT2-weighted images are better to explain the peritumoral edematous reaction [4]. Furthermore, in the paranasal sinuses the T2-weighted images allow to distinguish a retentional image from a true tumour extension, in fact, they will differentiate the relatively hyposignal of the tumour extent from the hypersignal of retained mucous debris in the obstructed sinuses [9]. General radiographic features of osteosarcomas are non- specific and maybe osteolytic, osteoblastic or mixed such as our case study [1]. In one extensive series of craniofacial osteosarcomas, excluding mandibular tumours, the radiographic presentation was osteolytic in the most cases (67%) [4].

The specular pattern of calcification is usually present, but the periosteal reaction is rarely observed in craniofacial osteosarcomas, and only in the mandibular lesions. Tumor matrix mineralisation is frequent, particularly in mandibular and maxillary lesions [4].

Although no radiographic finding is pathognomonic, the combination of several radiographic features could lead to this diagnosis in such rare location [1, 4, 5]. In fact, the presence of tumour matrix mineralisation, either osteoid or chondroid, with aggressive bone destruction and soft tissue involvement, leads directly toward a radiologic diagnosis of osteosarcoma without much difficulty [4]. The differential diagnosis of osteosarcoma and chondrosarcoma may be troublesome radiographically, and sometimes impossible even histologically. The chondrosarcomas are even rarer in the head and neck region. They appear less aggressive radiographically, with less bone destruction and more bone erosion [4]. Occasionally, osteosarcoma may be indistinguishable from the more common metastatic carcinoma, especially when it presents without matrix mineralisation [4].

In our case, CT scan and MRI findings were not conclusive but consistent with those of primary osteogenic sarcoma. However, a definite diagnosis is difficult to make by radiography alone as the final diagnosis of osteosarcoma is only histological [1, 3]. We performed a trans-temporal biopsy on our patient, which confirmed this diagnosis. The optimal treatment of primary craniofacial osteosarcoma has not yet been well established given the rarity of this tumour, so the available data are only from published isolated case reports and small case series. However, it seems that radical surgery with large margins is the most effective treatment as this condition represents the most significant prognostic factor for good outcome. Also, the use of pre or postoperative adjuvant therapy has not yet been well codified. In case of incomplete resection, radiotherapy could be considered, while the role of chemotherapy in locally advanced osteosarcoma is less clear as its efficiency remains uncertain in head and neck location given the lack of clinical trials including this entity [3].

Conclusion

Primary Osteosarcoma of skull bone is a very rare clinico-radiological entity. Although no radiographic finding is pathognomonic, the combination of several radiographic features including tumour matrix mineralisation, aggressive bone destruc-

tion and soft tissue involvement, leads directly toward a radiologic diagnosis of osteosarcoma without much difficulty.

The addition of our case to the literature offers new data of primary skull base osteosarcoma useful to raise awareness among radiologists adding this radiological entity as a differential diagnosis when a skull bone process is identified.

Authors' Statements

Competing Interests

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

There were no financial support or relationships between the authors and any organization or professional bodies that could pose any conflict of interests.

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