Case Report

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An Unsual Site about Small Cells Osteosarcoma Parietal Region and Classique Osteosarcoma Occipital Region Mimicking Meningioma with Literature Review Management and Outcome

Bankole NDA[1,3]*, Sayore M[1,3], Denou M[1,3], Cherradi N[2,3], Melhaoui A[1,3], Boutarbouch M[1,3] and El Ouahabi A[1,3]

[1]*Service de Neurochirurgie de l'Hôpital Des Spécialités Rabat, Marocco*

[2]*Service de Neuro-Anatomopathologie de l'Hôpital Des Spécialités Rabat, Marocco*

[3]*Mohammed V University of RABAT*

***Corresponding author**
Bankole NDA, Service de Neurochirurgie de l'Hôpital Des Spécialités Rabat, Marocco, E-mail: bankolenouroudine@yahoo.fr

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Abstract

Introduction: *Primary osteosarcoma (OS) is the second most common primary bone malignancy, the first being multiple myeloma. OS occurs in the second decade, with a predilection for ends of long bones. Head and neck involvement is seen in 2-9% with extragnathic craniofacial bones in 1–2% of cases. Small Cell OS (SCO) constitutes 1.3-4% of all OS, skeletal distribution and age range being similar.*

Materials and Methods: *We report two rare osteosarcoma and we done the review of the literature about the management and the outcome about intracranial osteosarcoma in our department of neurosurgery.*

Results: *It is two osteosarcoma cases about a 72-year-old man and one 49-year-old man who both mimiking first meningioma. The first case is an unusual site parietal and the second case is occipital. The both benefited surgery with excision and exam of histology confirm diagnosis. But the first case died 15 days after surgery in intensive unit care and the second cases died after one year, he benefited surgery and chemotherapy.*

Conclusion: *Small cell osteosarcoma (SCO) is an extremely uncommon entity that mainly involves the metaphysics of long bones and, rarely, the skull. Histopathology is the key to establishing the correct diagnosis, including sub typing for appropriate management and prognostication, as radiological features are not specific.*

Keywords: Osteosarcoma, Intracranial, Small Osteoid Cell, Osteosarcoma Outcome

Introduction

Skull-based OS clinically presents as scalp swelling, pain or both, with a duration of a few weeks to months. Non-specific radiological features may pose a diagnosis difficulty [1,2]. Histologically, small cells *osteosarcoma* shows a predominant population of malignant small round cells, instead of the more common spindle cells, with foci of bone formation. However, extensive tissue sampling may be required for *osteoid* demonstration. The problem in diagnosis is further compounded as its immune histochemical profile overlaps with other more common malignant small round cell tumours (MSRCTs), including Ewing sarcoma (ES), mesenchymal chondrosarcoma and lymphoma. Rare locations, such as the skull, and lack of awareness of this rare histological subtype, may lead to erroneous diagnosis. The aim of this work is to highlight such an entity at an unusual site like the parietal region and Occipital region and how sometimes they mimicking meningioma like diagnosis and the different sub-types of hystogy about intracranial *osteosarcoma*, treatment and outcome considerations in a 72-year-old man and 49-year-old man and the review of the literature about the management and the outcome about intracranial *osteosarcoma*.

Cases Presentation

Case 1

A 72-year-old patient with a history that dates back to 12 months by the appearance of a mass of the parietal scalp with a brutal installation, 3 months later left hemiplegia following a trauma at the point of cranial impact. This symptomatology complicated by deep vein thrombosis of the left lower limb 2 months later and put on anti-vitamin K treatment, Sintrom. Objective clinical examination revealed a left pyramidal syndrome with a soft mass at the level of the immobile vertex compared to the deep plane with bone lysis and without inflammatory sign.

Investigation

MRI of the brain showed a large heterogeneous lesion in the right parietal region. Overlying parietal bone was not visualized. The mass was intracranial with subgaleal extension and had an enhancing soft tissue component with areas of calcification and High density of bleeding. On radiology, possibility of metastasis, meningioma was

suggested (Figure1 and 2).

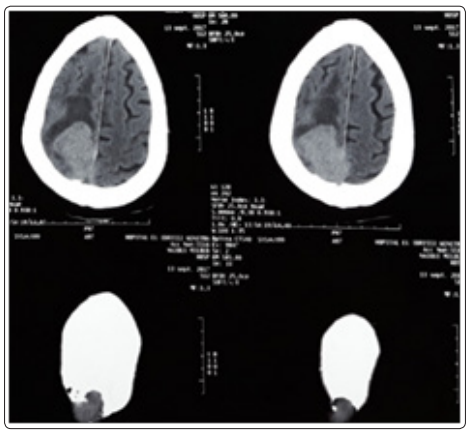

Figure 1: CT scan show the mass and lyticBone and attached of sinus longitudinal

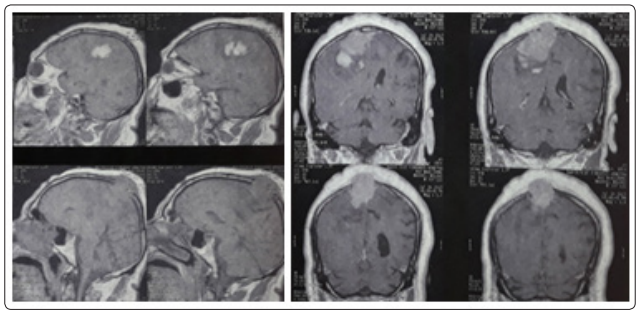

Figure 2 : Sagital and Coronal T1 MRI showinga mass lesioninvolving the right parietalregion with overlyingparietalbone.The mass isintracranial with subgaleal extension and has an enhancing soft tissue component with an area ofcalcification (arrow). The mass isassociated with oedema and falcosinusal invasion.

Management

Patient underwent complete resection with bone resection and cranioplasty, in pre procedure of exicion, possibility of métastasis or ostesarcoma (Figure 3a).

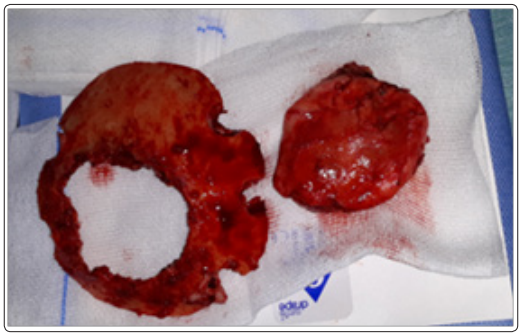

Figure 3a: Macroscopic Aspect of the lesion per-procedure

Histopathological examinations revealed a Focal presence of lace-like material (osteoid) in between the cells and confirm that is an *osteosarcoma* (Figure 3b and 3c).

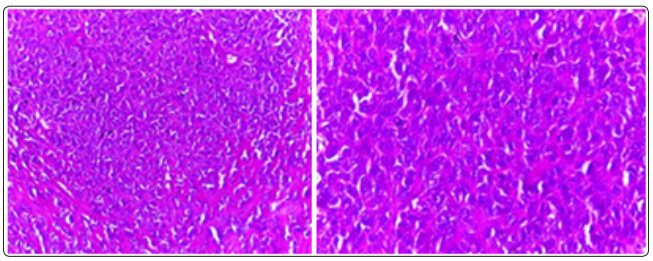

Figure3b: Small Rounds Cells Proliferation with lace-likeosteoid

Figure 3c: Higher Magnification ofsmall round cellsproliferation

Outcome and Follow-up

On follow-up, the patient developed frequent seizure episodes in post-op. The patient then degraded 3 days after the tumor excision of his Glasgow Coma Scale, presented respiratory disorders, and succumbed in intensive care 15 days later. So he could not benefit from chemotherapy.

Case 2

This is a 49-year-old smoking and alcoholic patient with no significant pathological history. The onset of the disease traced back to 5 months by the appearance of an occipital swelling of progressive onset associated with frontal headaches without vomiting with a decrease of visual acuity. He has benefited first time surgery by a neurosurgeon with a cytopathology anatomy examination who concludes a meningioma, 2 months later he found a significant recurrence of the occipital mass.

Investigations

MRI of brain showed a large hypo-intense occipital process with an extra-axial intracranial component and an extra-cranial component with extensive occipital bone lysis with an extension of the upper longitudinal sinus and some intratumoral calcifications. The process strongly takes the contrast in a heterogeneous way (Figure 4 and 5).

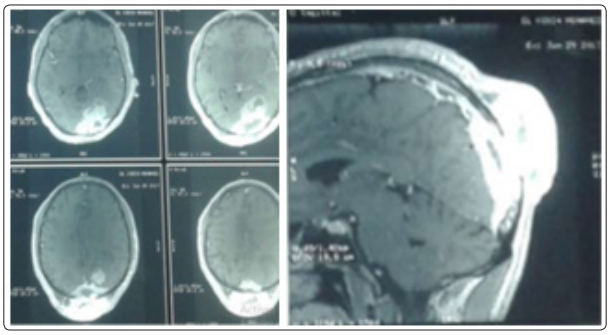

Figure 4 **Figure 5**

Management

The patient benefited from a partial excision of the extra cranial mass at first then an excision of the intracranial mass in a second time following the recurrence. The 2nd histology examination revealed tumor proliferation composed of moderately atypical ovoid-sized cells with richly vascularized anisocaryototic hyperchromatic rounded nuclei and articulate around small spicules of an osteoid substance revealing the morphological appearance of *osteosarcoma* contrary to the 1st histology examination (Figure 6a and 6b), which showed WHO grade I meningioma.

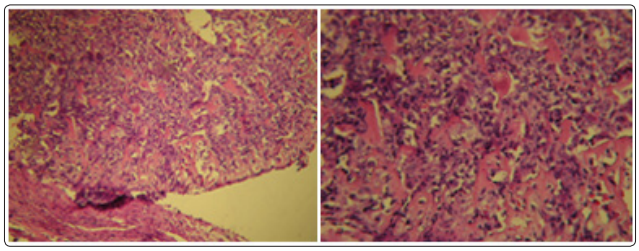

Figure 6a **Figure 6b**

Outcome and Followup

The patient subsequently referred to oncology for radiotherapy with a recurrence at 1 year and died after this recurrence.

Discussion

Primary *osteosarcoma* is the second most common primary bone malignancy, the first being multiple myeloma. *Osteosarcoma* is the most common bone tumor of the child and the adolescent represents 42% of the malignant tumors of the bone. Its frequency peak is between 10 and 25 years old. Occurred before 5 years and after 30 years is rare. Men are more affected with a sex ratio of 1.5 [3,4]. Ours patients are 49 and 72 years old man significantly, in their series; pain was the most common clinical manifestation, being present in all, with duration of pain ranging from 1 to 20 months [5]. However, in the series by Ayala and al, pain and swelling of the affected area were the most common symptoms [6]. In our 1st case, the symptoms is this appearance of a mass of the parietal scalp with a brutal installation, 3 months later left hemiplegia following a trauma at the cranial impact and in the 2nd case the onset of the disease traced back to 5 months by the appearance of an occipital swelling of progressive onset associated with frontal headaches without vomiting with a decrease of visual acuity.

Small cell *osteosarcoma* of bone was first described by Sim et al., in 1979, as a small round monomorphic tumour with cells separated by a fibrous component, occurring mainly in the second decade of life [7]. The WHO defines it as a high-grade malignant neoplasm composed of small cells, with a variable degree of osteoid formation. Metaphysis of long bones (90%) is the predominant site of involvement. Awareness of this morphological subtype of OS is important. Exact diagnosis is mandatory owing to poorer outcome than conventional *osteosarcoma* and Ewing sarcoma. Being non-radiosensitive, aggressive radical surgery with multi agent adjuvant chemotherapy constitutes the main treatment modality.

A local recurrence rate of 26-69% and 5-year survival rate of 25-37% have been observed for craniofacial OS, the former being the major cause of death [8-10]. In our parietal case, he is died few days after surgery, the patient didn't have time to benefit chimiotherapy, and we confirm that OS is tumors with high bad pronostic. Our occipital case benefited chemotherapy and died after one year. We confirm that OS is tumors with high bad pronostic.

Conclusion

Small cell *osteosarcoma* (SCO) is an extremely uncommon entity that mainly involves the metaphysis of long bones and, rarely, the skull. Histopathology is the key to establishing the correct diagnosis, including subtyping for appropriate management and prognostication, as radiological features are not specific. Being non-radiosensitive, aggressive radical surgery with multi agent adjuvant chemotherapy constitutes the main treatment modality. Anyway the pronostic is very bad [11].

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