Parosteal osteosarcoma of the skull

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Summary

Parosteal osteosarcoma of the skull is a distinct surface bone tumor, with a better prognosis than conventional osteosarcoma. The first series of this tumor was reported by Geschickter and Copeland4. POS is a primary neoplasm of bone that comprises 1.7% of total bone tumors, 1% of primary malignant bone tumors, and 4-6% of all osteosarcomas1,2,4,14. The most common location is on the surface of the distal femur which account for 46-66% of the cases6,14. The presentation in the skull is rare and only 11 cases have been described, one in the occipital bone7,13.

We describe the case of a patient with a POS developing from the occipital bone with extension to the parietal bone.

KEY WORDS: Parosteal osteosarcoma. Osteosarcoma. Skull neoplasm.

Osteosarcoma perióstico del cráneo

Resumen

El osteosarcoma perióstico del cráneo es un tumor de la superficie ósea de características bien definidas y con un mejor pronóstico que el osteosarcoma convencional. La localización más común es el fémur donde asientan aproximadamente entre el 46-66% de estos tumores. La presentación en el cráneo es muy poco frecuente habiéndose descrito pocos casos.

Describimos el caso de un hombre que desarrolló un osteosarcoma perióstico a partir del hueso occipital con extensión al hueso parietal. El paciente fue operado con resección tumoral completa. Se discute el caso y se revisa la literatura.


Introduction

Parosteal osteosarcoma of the skull (POS), also known as juxtacortical osteosarcoma, is a distinct surface bone tumor, with a better prognosis than conventional osteosarcoma. The first series of this tumor was reported by Geschickter and Copeland4. POS is a primary neoplasm of bone that comprises 1.7% of total bone tumors, 1% of primary malignant bone tumors, and 4-6% of all osteosarcomas1,2,4,14. The most common location is on the surface of the distal femur which account for 46-66% of the cases6,14. The presentation in the skull is rare and only 11 cases have been described, one in the occipital bone7,13.

We describe the case of a patient with a POS developing from the occipital bone with extension to the parietal bone.

Case report

A 49-year-old Caucasian man without personal or family history of Paget disease, presented 2 years before admission with a gradually enlarging, painless mass that occupied the occipital and parietal bones, with deformation of the calvaria. The patient had no other symptoms such as headache. Neurological examination and hematology and biochemistry parameters were normal. A mass, 6 cm in diameter, solid in consistency and not adherent to the overlying skin, was appreciated in parietal and occipital regions. Local pressure elicited pain.

Chest x-ray was normal and the CT of the skull revealed a cranial parietooccipital mass with its primary origin in the outer table of the skull and showing spicular reaction in its entire circumference (Fig. 1). It is noteworthy that medullar bone was not affected and there was no apparent intracranial extension.

Sagittal and axial MRI TI weighted images showed a diffuse circumferential involvement of the outer table and a radial spicular reaction (Fig.2). The signal intensity of the intramedullary space of the skull was normal.

Cerebral angiographic study showed that the lesion was avascular.

The patient was operated on and the lesion was removed in block, preserving the internal table of the skull (Fig. 3).

The pathological study showed that the lesion was a parosteal osteosarcoma.
Two months later a new lesion measuring approximately 1 cm was observed in the parietal bone. A new surgical resection was performed. After this second operation the patient received conventional radiotherapy and chemotherapy with good evolution (Fig. 4).

Two years later he developed a left hemiparesis and the chest x-ray showed lung metastases. A control MRI showed a local relapse with intracranial invasion compressing the brain.

The patient was reoperated on and again tumor removal was considered radical. However, the histological diagnosis at this time suggested an osteosarcoma and the patient died a few months later.

**Discussion**

Parosteal osteosarcoma is a rare primary neoplasm of the bone, affecting females more frequently than males. The most common location is in the metaphyseal end of long bones, but they are rare at the skull. Kumar et al. described eight cases located on the calvaria affecting six females and two males, with a mean age of 19 years; two of these tumors located in the mastoid region, two in the frontoparietal region, two in the parietal region and one in the occipital bone. Overall, the most common location of POS is in the temporal bone, whereas in the head the first location is in the jaw. POS of the long bone typically pre-
sent in the third or fourth decades in contrast to POS of the skull which tend to occur in the second decade of life. This is probably due to the comparatively smaller amount of tissue overlying the cranial bones.

Clinically POS are tumors producing external deformation of the head. They are firm to hard in consistency or minimally tender on deep palpation. Habitually they appear as painless masses fixed to the underlying skull and have a variable size. The average diameter in the reported cases is about 16 cm².

POS have characteristic radiological features which allow a preoperative diagnosis to be made. Skull x-ray shows a dense and lobular mass with a broad base of attachment to the external cortical bone. CT allows the possible presence of tumor in the intramedullary region to be excluded⁸,⁹.

Cerebral angiography, which in patients with low grade cranial POS usually shows an hypevascular lesion, turns hypervascular when the tumor is dedifferentiated and malignant⁸,⁹.

Differential diagnosis of POS include conventional osteosarcoma, chondrosarcoma, osteochondroma, giant osteoma and organizing scalp hematoma²,¹⁴.

Radical "on block" resection is the treatment of choice of POS. 20% of the large bone POS present malignant dedifferentiation and require more extensive surgery. POS of the skull showing dedifferentiation have also been reported. Usually no adjuvant therapy was necessary¹⁶.

Pulmonary metastases have been described in patients with POS of the long bones with medullary invasion. However, pulmonary metastasis have not been reported in cases of cranial POS³,¹².

The prognosis of POS is more benign than that of conventional osteosarcoma, and longer survival of patients has been reported¹⁴,¹⁵. Osteosarcoma of the skull is the second bone neoplasm in this location after mieloma. Previous history of Paget's disease or radiation is often found. It is more
aggressive than POS and therapy include surgery, radiotherapy and chemotherapy, with disease free survival rates of 80% at 3 years\textsuperscript{1}.

In our case we considered that the tumor resected at the second operation presented dedifferentiated areas and the diagnosis could be changed to osteosarcoma.

References


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