

Spheno-orbital mesenchymal chondrosarcoma: A rare cause of exophthalmia

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Abstract

Intracranial primitive mesenchymal chondrosarcoma is a highly malignant and extremely rare tumour. Orbital chondrosarcomas was reported in only 18 cases until 2004.

Case description: A 36-year-old woman presented with left painless exophthalmia with temporozygomatic tumefaction. This exophthalmia was progressive during 4 months, with decrease in visual acuity in left eye. Oculomotricity examination showed near ophthalmoplegia. Palpebral occlusion was impossible. Visual acuity was 10/10 on right eye and 6/10 in left eye. A temporal tumefaction was noted. This swelling was firm, non mobilisable and adhered to deep zygomatic area. Cranial and skull base CT scan showed a process occupying space lesion in the orbit and sphenotemporal area with extension in intracranial temporal fossa. Cranial and orbital MRI showed a hypo intense signal on T1- and hyper signal on T2- images. Cerebral angiography showed intense tumoural blush fed by external carotid and internal maxillary artery. Twenty-four hours before the surgery the internal maxillary artery was embolised. Surgery was performed using a left fronto temporal approach with lateral orbitotomy to access intraorbital tumoural extension and complete removal was performed. Histopathological specimens showed a mesenchymal chondrosarcoma with immunoreactivity to antivimentine body and no reactivity to leucocyte commun antigen, epithelial membrane antigen, cytokeratine, protein S 100 or desmine.

Postoperative period was unremarkable with regression of exophthalmia. The patient received chemotherapy and radiotherapy. After 12 months follow-up the patient is asymptomatic.

Conclusion: Spheno-orbital chondrosarcoma is a rare cause of tumoural exophthalmia. (p86-89)

Keys words: *Exophthalmia, mesenchymal chondrosarcoma, spheno-orbital, surgery, radiotherapy and chemotherapy.*

Introduction

Chondrosarcoma represents 6% of all skull base tumours and 0.15% of all cranial space occupying lesions. Only 192 intracranial chondrosarcoma were reported in the literature since 1998.⁷

Chondrosarcoma of orbit is extremely rare; only 18 cases have been reported until 2004.⁹ A case of spheno-orbital mesenchymal chondrosarcoma revealed by exophthalmia is reported.

Case Report

A 36-year-old Moroccan woman presented to the Department of Mohamed V Military Hospital in Rabat, for left exophthalmia with temporozygomatic area tumefaction.

The medical history was unremarkable, no medical antecedent was noted. This exophthalmia was progressive during 4 months, with decrease in visual acuity in left eye and this exophthalmia was painless.

On examination, the patient was in good health and normotensive but left non axial and not reducible exophthalmus was noted. This exophthalmus was not pulsatile. Oculomotricity examination showed near ophthalmoplegia, with limitations in gaze movement. Palpebral occlusion was impossible. Eye annex examination showed no normality. Anterior segment was unremarkable. Visual acuity was 10/10 on right eye and 6/10 in left eye. Fundoscopy was normal in both eyes. A temporozygomatic tumefaction was noted. This tumefaction was firm, non mobilisable and adhered to deep zygomatic area. The somatic examination

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showed no abnormalities.

Cranial and skull base computer tomography (CT) scan showed a process occupying space lesion in the orbit and sphenotemporal area with extension in intracranial temporal fossa. Bone window CT scan showed central specula calcification. After intravenous infusion of contrast this lesion showed an intense enhancement (Fig. 1). Cranial and orbital magnetic resonance imaging (MRI) showed a hypo intense signal on T1- and hyper signal on T2- images (Fig. 2).

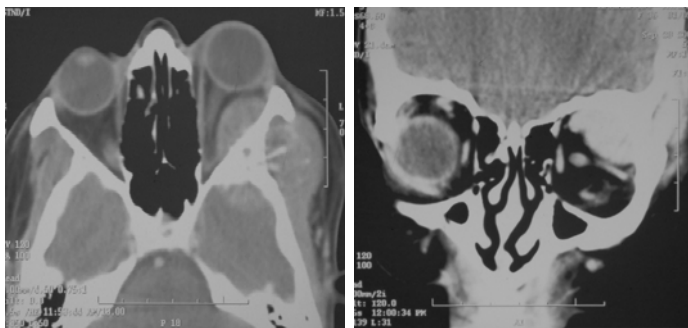


Figure 1 - Cranial CT scans on axial and coronal section showing an iso-intense lesion in left orbit and temporal fossa responsive of exophthalmia with optic nerve compression. Note the intense enhancement after intravenous infusion of contrast.

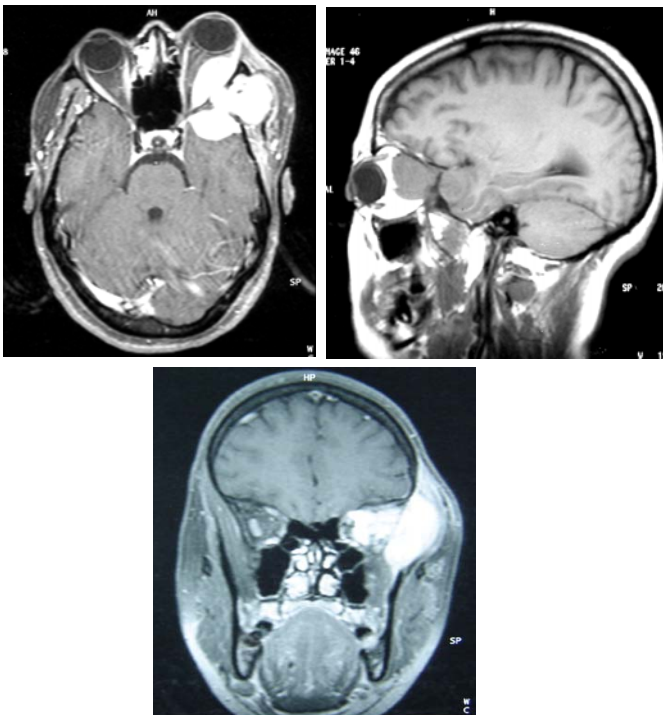


Figure 2 - Cranial MRI on axial, sagittal and coronal section showing three components in orbit, temporal fossa and temporozygomatic area responsive of left exophthalmus.

Cerebral angiography showed intense tumour blush fed by external carotid and internal maxillary artery. Twenty-four hours before the surgery the internal maxillary artery was embolised (Fig. 3).

Surgery was performed using a left fronto temporal approach with lateral orbitotomy to access to intraorbital tumoural extension. A grayish-white infiltrant processus was removed. Intraorbital component of lesion showed more bleed than temporal one during excision. Complete removal was performed.

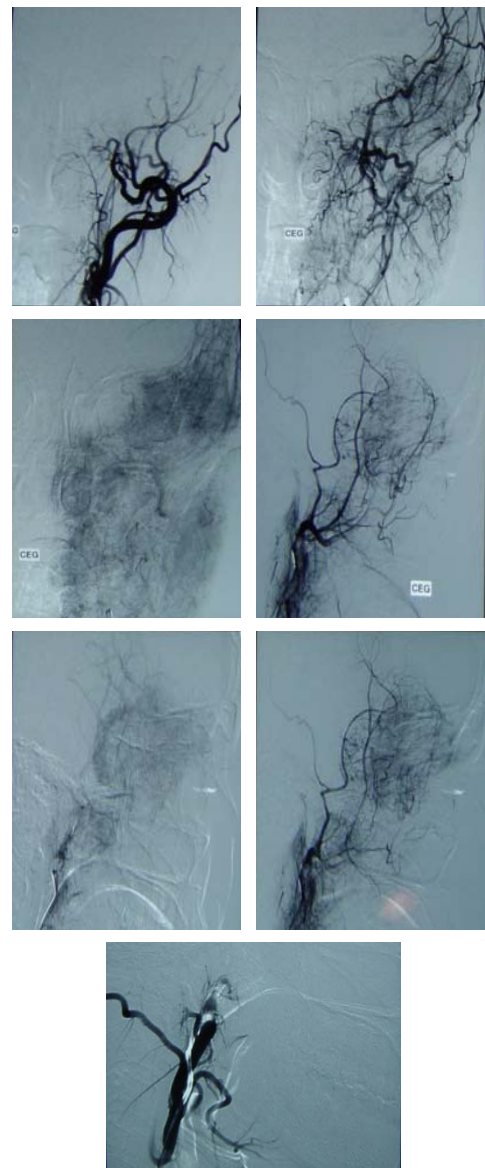


Figure 3 - Left carotid cranial angiography showing intense tumour blush fed by external carotid artery before (a) and after embolization (b).

Histopathological specimens showed a mesenchymal chondrosarcoma with immunoreactivity to antivimentine body and no reactivity to leucocyte common antigen (LCA), epithelial membrane antigen (EMA), cytokeratine, protein S-100 (PAS) or desmine (Fig. 4).

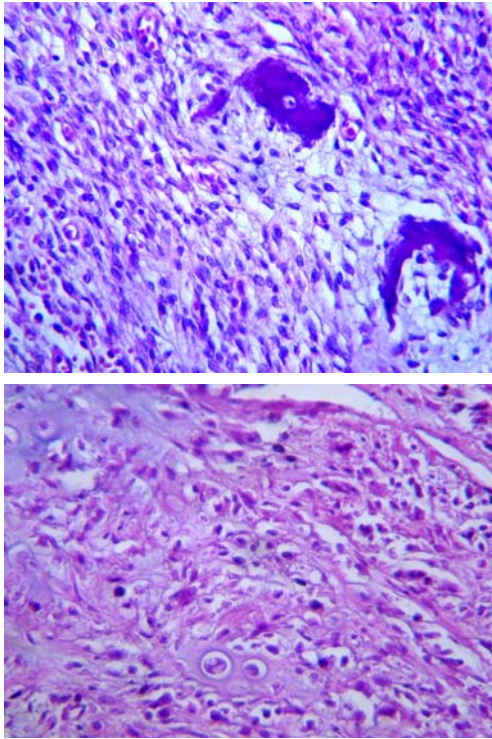


Figure 4 - Histological study with HE $\times 400$ showing (a) tumoural proliferation of fusiform cells and undifferentiated avoids cells and (b) area of cartilaginous differentiation cells.

Postoperative period was unremarkable with regression of exophthalmia. The patient was oriented to Oncology Department for adjuvant therapy of chemotherapy and radiotherapy. She received three cycles of cyclophosphamide, vincristine, actinomycyne D, spaced by 3 weeks. A 65 Gray fractionated radiotherapy was given. Two weeks postoperative CT and MRI showed total removal and good and aesthetic orbital reconstruction (Fig. 5). After 12 months follow-up the patient is asymptomatic.

Discussion

Our patient presented a tumoural exophthalmia revealing a mesenchymal chondrosarcoma of sphenoidal area. These findings are very rare.⁷

Chondrosarcoma represents 6% of all skull base tumours. It is assumed that chondrosarcoma originate from remnants of embryonic cartilage or from metaplasia of meningeal fibroblasts.¹

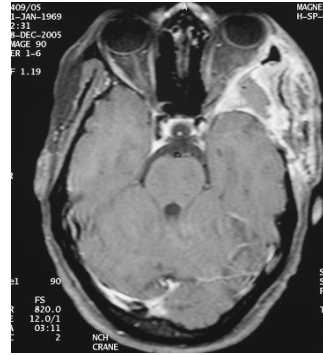


Figure 5 - Two weeks postoperative MRI showing total resection and regression of exophthalmia.

Arthur et al, in his review of literature found only 192 cases intracranial chondrosarcoma.⁷ The chondrosarcoma were located in the petrosal bone in 37%, in the occipital bone and clivus in 23%, in sphenoid bone in 20% and to a lesser extent in frontal, ethmoidal and parietal bones 14%.⁷ Only 18 orbital chondrosarcoma have been reported until 2004 by Tuncer, et al.⁹

No sex dominance existed. The mean age was 37 years. Our patient was 36-years-old. Chondrosarcoma occurred in both very young and old age groups, from 3 months to 76 years of age. The mesenchymal subtype showed a tendency to occur at younger age.⁷

The first clinical manifestation of the chondrosarcomas was mainly caused by oculomotor dysfunction, related to the preferable location of chondrosarcoma in the petrosal part of the skull base. Central lesions caused by intradural and intracerebral expansion were reported in 30 cases. Most cases were described as originating from skull base, possibly from remnant of embryonic cartilage.

Four types of skull base chondrosarcoma were described in the literature: grade I, II, mesenchymal, and myxoid type.⁷ The pathological classification of the subtype's grade I-III is based on differences in characteristics such as nuclear size, cellularity, mitotic rate and frequency of lacunae with multiple nuclei.⁷ In the mesenchymal subtype, primitive spindle cells are present. The myxoid type as composed of strings of rounded cells in a more or less myxoid matrix.

Mesenchymal chondrosarcoma occurred in the younger age group (10 - 30 years). The grade I chondrosarcoma had no clear age preference.

On histological examination, 51% of tumours were classified as grade I, 11% grade II, 30% mesenchymal and 8% myxoid. The mesenchymal type was the most malignant as illustrated by a strong tendency to intradural

and cerebral growth and possible occurrence in younger age groups.

Immunohistology study can differentiate chondrosarcoma from chordoma. Chordomas lack vimentin immuno-reactivity and chondrosarcoma fail to express cytokeratin, S-100 protein expression is present in both. In our case it was immunoreactivity to vimentine, and no immunoreactivity to LCA, EMA, cytokeratine PAS 100 or desmine.

Plain skull radiography showed only bone destruction and calcifications. Computed tomography scan showed bone destruction and calcifications. On T1-weighted MRI, chondrosarcoma had a low to intermediate signal intensity and were isointense or hyperintense to gray matter. On proton density and T2- weighted images they had high signal intensity and were hyper intense to gray matter.⁸

The use of fat-saturation sequence is helpful in analysing intraorbital extension of chondrosarcoma. The MRI gives better tumour demarcation and visualisation of dural extension of neurosurgical extirpation.⁸

Angiography had shown a tumoural blush in our case. The preoperative embolization was very helpful to prevent excessive bleeding during surgery.

The usual methods of treatment were neurosurgical and conventional or proton radiotherapy.^{7,9} Adjuvant chemotherapy was scarcely mentioned. Macroscopically, total resection was accomplished in 56 - 67% of cases and subtotal in 20% cases.⁶ In 53% of neurosurgical treated patients recurrence of the tumour was found (mean interval

32 months). The high recurrence rate is caused by partial resection due to the proximity of critical neuronal and vascular structures.⁷

Recent promising results imply that charged particle radiotherapy, in combination with surgery, may be the therapeutic choice of the future.

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