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Osteogenic sarcoma of sphenoid bone: an extended lateral skull base approach

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Abstract
Osteogenic sarcoma involving the sphenoid bone is an extremely rare condition. The rarity of the disease and the close proximity of the sphenoid bone to the various important intracranial structures poses a real challenge in diagnosis and surgical management of these lesions. An extended lateral craniofacial resection by a multidisciplinary approach was carried out in one such case to attempt en bloc resection. This case is presented and also a review of the relevant literature.

Key words: Sphenoid bone; Osteosarcoma

Introduction

Osteogenic sarcoma of head and neck is a rare clinical entity. It may arise in the mandible or the maxilla (Bone *et al.*, 1973) but is rare in the ethmoid and sphenoid bones. Sataloff *et al.* (1988) reported 19 cases of osteogenic sarcoma in the temporal bone whereas Bradley *et al.* (1988) reported only two cases in the ethmoid. To our knowledge, no case of osteogenic sarcoma primarily arising from the sphenoid bone has been reported in the English literature. A case of osteogenic sarcoma of the sphenoid bone is reported here to emphasize the need for an extended lateral skull base approach for complete resection of the tumour. The approach adopted was the subtemporal transparotid transzygomatic approach. A review of the available literature failed to show any report of such an approach as a definitive surgical treatment for these tumours.

Case report

A 19-year-old female was referred to the ENT Department at Kasturba Hospital in March 1990 with the complaints of headache, diminished vision, proptosis and facial swelling on the left side of two years duration (Figure 1). On examination proptosis was axial, irreducible, nonpulsatile and nontender. There was chemosis and congestion of conjunctiva. The cornea was dry with macular opacity. Eye movements on this side were restricted. On palpation there was a hard irregular mass (5 × 4 cm in size) involving the superolateral orbital margin. Ophthalmological examination revealed finger counting at a distance of one metre. Fundoscopy showed optic atropy and the peripheral field was constricted. The right eye was normal. All cranial nerves except the IInd were normal. Nasal endoscopy revealed polypoidal mucosa of the left osteomeatal complex. CT scan showed a bony mass arising from the sphenoid wing (Figure 2) involving the lateral wall and roof of the orbit and left frontal bone. Other investigations were within normal limits. A limited lateral surgical approach was carried out

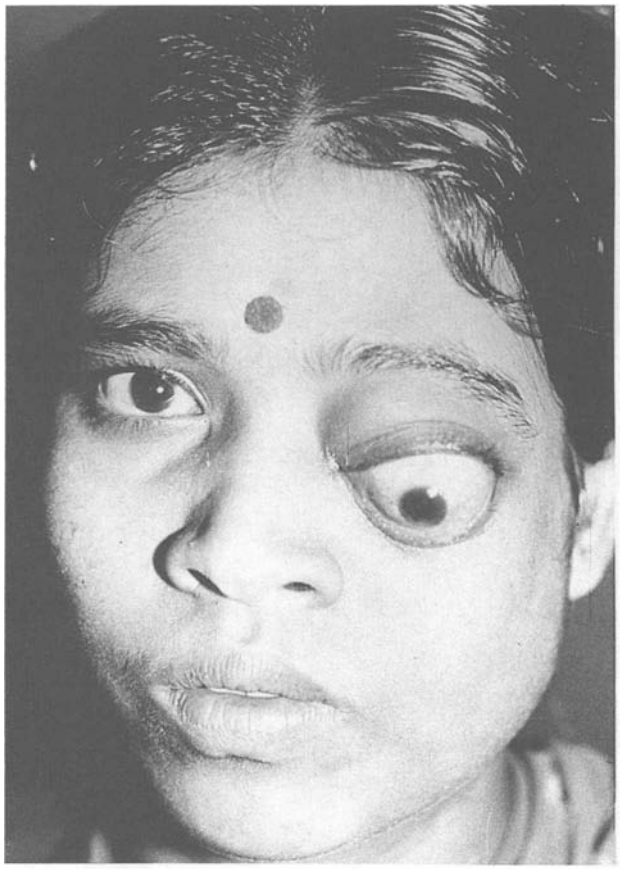

FIG. 1
Showing the proptosis and facial swelling.

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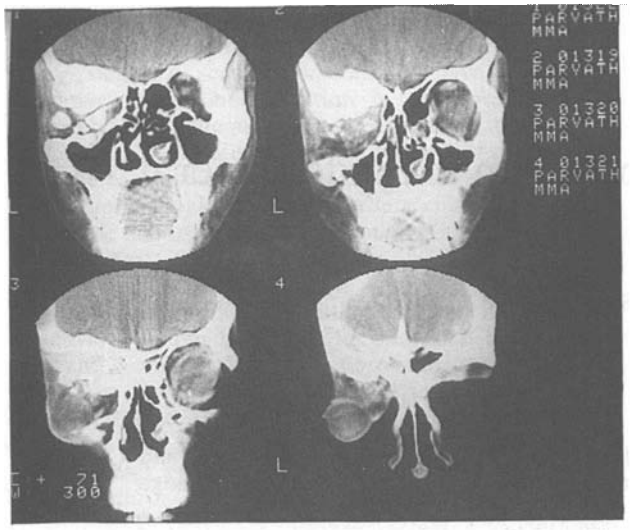

FIG. 2

CT scan showing bony mass arising from sphenoid wing involving left orbital roof and left frontal bone.

to remove the bony swelling for histopathological examination. This showed trabecullae of woven lamellar bone with a spindle cell stroma which contained an occasional mitosis. There was focal osteoblastic rimming with chondroid differentiation. A diagnosis of parosteal osteogenic sarcoma was made (Figure 3).

The patient was lost to follow-up for one year but represented in June 1991 with similar complaints. Ophthalmological examination revealed no vision in the left eye with loss of light and accommodation reflex. The pupil was dilated. A repeat CT scan showed the same bony mass with involvement of more cranial bones.

A left side extended lateral craniofacial resection was performed in July 1991 to attempt an en bloc resection. Under general anaesthesia, a subtemporal transzygomatic transparotid incision was made (Figure 4). The internal carotid artery was mobilized and controlled after mandibular condylectomy. The zygomatic arch was cut and mobilized downward. The temporalis muscle was incised close to its origin and was reflected downward. At this point the eyeball was exenterated and further resection of the facial flap was made to visualize the tumour. A burr hole was made in the region of pterion. The bone was nibbled out encircling all round the tumour margin including the anterior part of the squamous temporal bone, part of the parietal bone and the frontal one which were involved by the tumour. The tumour was mobilized from anterior and middle cranial fossa dura and removed. Extended lateral craniofacial resection included orbital extenteration, resection of left frontal bone, orbital roof, greater wing of sphenoid bone up to the cavernous sinus, portion of the left parietal bone and antero-aspect of left squamous part of temporal bone (Figure 5). The cranial defect was repaired with temporalis muscle. The postoperative period was uneventful except for CSF collection in the lower part of the neck which resolved completely

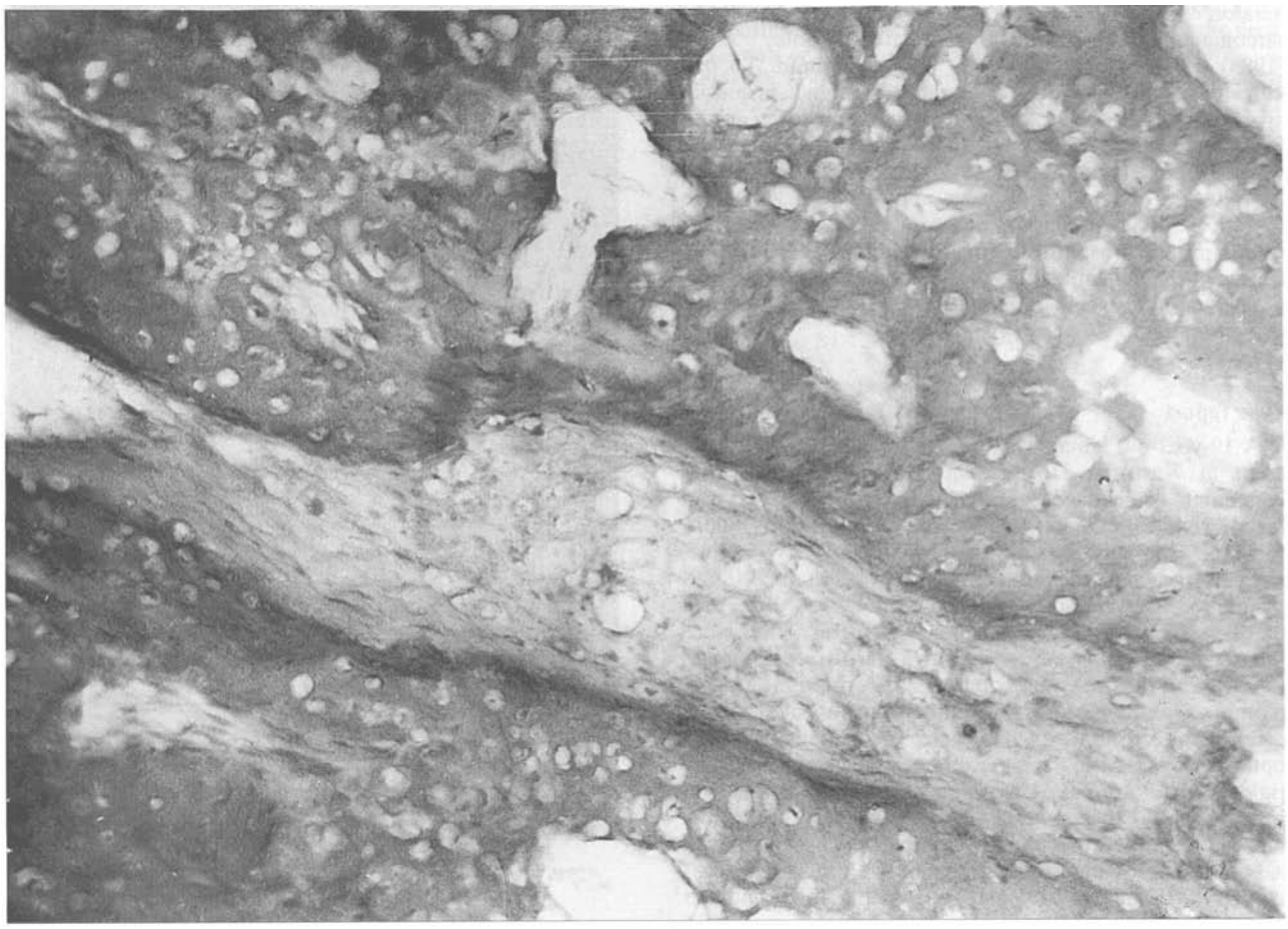

FIG. 3

Showing trabecullae of woven lamellar bone with spindle cell stroma, seen with an occasional mitosis. There is focal osteoblastic rimming with chondroid differentiation.

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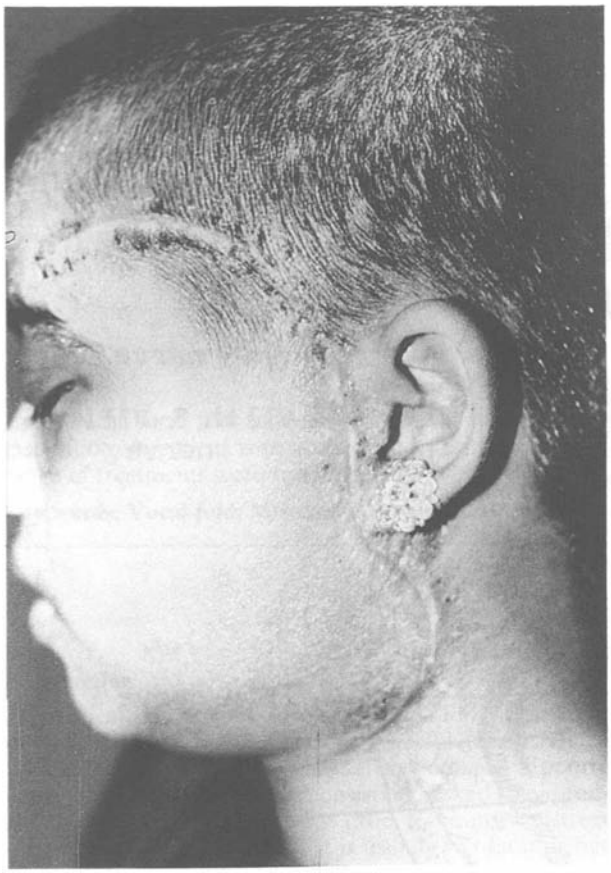

FIG. 4
Subtemporal transparotid transzygomatic incision.

after two weeks. The follow-up after three months revealed no evidence of disease. She refused post-operative chemotherapy and radiotherapy because of financial constraint.

Discussion

Osteogenic sarcoma is one of the most dreaded malignant tumours and has a very poor prognosis. It is commonly seen in the metaphyses of the long bones and is extremely rare in the head and neck region comprising only 0.5 per cent of all cancers (Defries *et al.*, 1979). The mandible and maxilla are commonly involved in the head and neck region. Involvement of other skull bones is infrequent (Bone *et al.*, 1973). Histologically osteosarcomas are of three types: (1) conventional, (2) parosteal and (3) extraoseous.

Our case was diagnosed as parosteal osteogenic sarcoma which was established as a different clinicopathological entity by Gechikter and Copeland (1952). It is defined as an osteosarcoma upon the external surface of the bone in close relation to the periosteum or immediately adjacent to periosteal soft tissue. Characteristic features of these tumours are the presence of woven lamellar bone with an inconspicuous focal osteoblastic riming. It accounts for three to five per cent of all osteosarcomas (Huvos, 1979). In contrast to conventional osteosarcomas it has a significantly better prognosis and is found predominantly (80 per cent of cases) in patients over the age of 20 years (Dahlin, 1978). Bras *et al.* (1980) identified seven cases occurring in the jaws, five in the mandible and two in the maxilla. It is extremely rare to see an osteogenic sarcoma arising from other parts of the skull (Bone *et al.*, 1973). We found only two cases of osteogenic sarcoma in the ethmoidal sinuses in the literature (Bradley *et al.*, 1988).

Early diagnosis of these tumours are difficult clinically because of their strategic position. Headache, ophthalmological symptoms and facial swelling are the first symptoms to appear as seen in our case. CT scan and MRI are helpful to observe the exact tumour dimensions. However three-dimensional high resolution volume rendering of computer tomography has also been developed as an important tool with which to determine the anatomical relationships of the tumour. This is often obscured on conventional CT scan (Davies *et al.*, 1991).

The treatment modalities of these tumours are difficult to determine because of the lack of literature on this subject. No single series has mentioned a definite line of management for a therapeutic procedure. Consensus of opinion is to perform surgical excision followed by chemotherapy or radiotherapy. En bloc resection of osteogenic sarcoma of the sphenoid is perhaps better dealt with by extended lateral craniofacial resection (as experienced by us). This approach gives a wider exposure and better control of the neurovascular bundle in the skull base.

Post-operative care is extremely important besides intra-operative monitoring. Reconstructive procedure for the post-resection large craniofacial defect should be planned in advance. In the present case temporalis muscle was used to reconstruct the defect. If the reconstruction is difficult using a local flap, a free microvascular flap can be used. CSF leak is a common problem in the post-operative period but usually resolves within a month. If it does not repeated lumbar puncture may be carried out until the leak is completely healed.

The prognosis for osteogenic sarcoma in general is grave, in spite of complete surgical excision. Chemotherapy has been claimed to improve the five-year survival (Link, 1981). Its efficacy in tumours involving head and neck has yet to be proved. Bradley *et al.* (1988) found no improvement in their case of ethmoidal osteogenic sarcoma and the patient died five months after surgery despite further chemotherapy. Defries *et al.* (1979) reported a good result with the combined approach of pre-operative radiotherapy, surgery and pre- and post-operative chemotherapy in osteogenic sarcoma of the mandible. In the past the prognosis for osteogenic sarcoma arising in the head and neck has been comparable to that of other sites with five-year survival rates of around 25 per cent (Garington *et al.*, 1967). It remains to be seen whether recent improvement in staging, investigation facilities and treatment will increase the survival rate in parallel with that at other sites.

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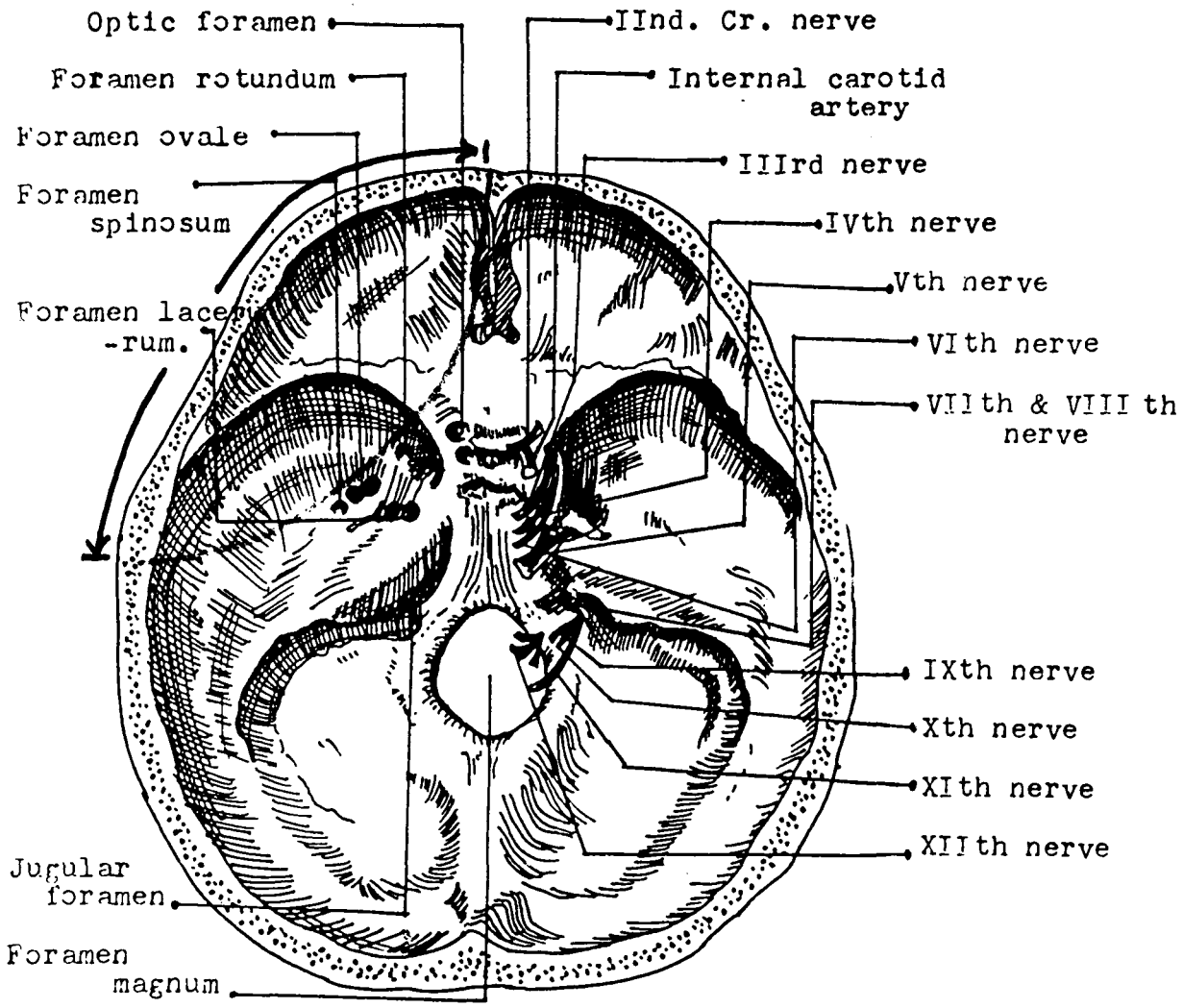

FIG. 5
Line diagram showing the excised area of the cranial bones.

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