Reconstructive Challenges in the Management of a Rare Case of Sphenoid Osteosarcoma – A Case Report

K B L Lee, E S W Ang, K C Tan

ABSTRACT

Primary osteosarcoma of the sphenoid bone is an extremely rare condition. This paper presents a case of a lady with recurrent oesteosarcoma of the sphenoid bone who had resection of the tumour via a combined neurosurgical and craniofacial procedure and reconstruction with a free flap and bone graft. It details the diagnosis, treatment and follow-up of this unusual condition.

Keywords: Skull base sarcoma, multi-modality therapy, combined approach, distant tissue transfer

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INTRODUCTION

Head and neck osteosarcomas are uncommon, accounting for less than 10% of all osteosarcomas. Most head and neck osteosarcomas occur in the mandible and maxilla[1]. Only a few cases of osteosarcoma of the skull base have been reported in the literature, most of which are radiation-induced[2,3]. Osteosarcomas arising *de novo* from the sphenoid bone are extremely rare[4-6].

Current mangement of head and neck sarcomas in general and osteosarcomas in particular stress multi-modality therapy, involving ablative surgery, reconstruction with or without radiation and chemo-therapy[7-10]. Multidisciplinary involvement by the neurosurgeon, head and neck surgeon and plastic surgeon is important in dealing with the complex issues involved in skull base surgery[11-16].

We report here a case of a lady with recurrent osteosarcoma of the sphenoid bone who had resection of the tumour via a combined neurosurgical and craniofacial procedure and reconstruction with a free flap and bone graft.

CASE REPORT

LBC is a 37-year-old lady who first presented in May 1997 with a bony lump over her left forehead for several months. A tumour was removed from the left sphenoid ridge via a coronal incision at another institution and the histology then showed fibrous dysplasia. She underwent six cycles of post-operative radiotherapy. She developed progressive diploplia on left outward gaze and left unilateral proptosis (Fig. 1) over the next few months prior to consultation at this institution. CT scan (Fig. 2) at this stage showed an osteoblastic tumour arising from the sphenoid bone extending to the temporal, fronto-orbital, zygomatic bones and the temporalis muscle. There was no involvement of the periorbita and orbital contents

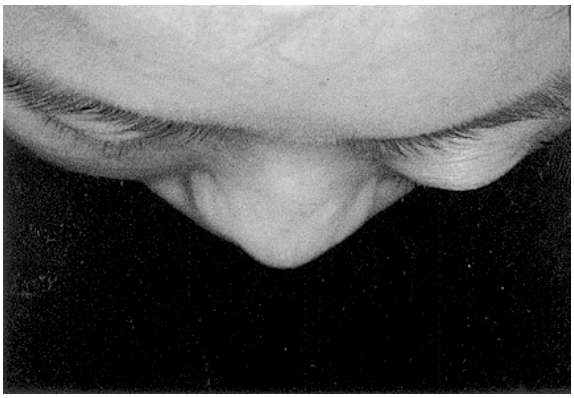

Fig. 1 Unilateral proptosis of the left eye.

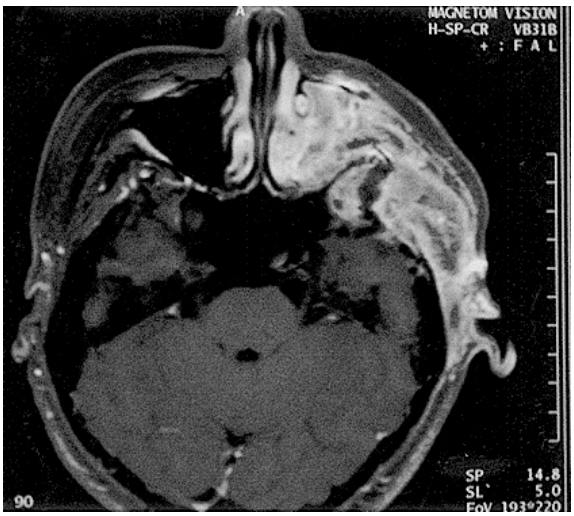

Fig. 2 Pre-operative CT scan showing an osteoblastic tumour rising from the sphenoid bone extending to the temporal, fronto-orbital and zygomatic bones and involving the temporalis muscle with sparing of the periorbita and orbital contents and dura covering the frontal and temporal lobes.

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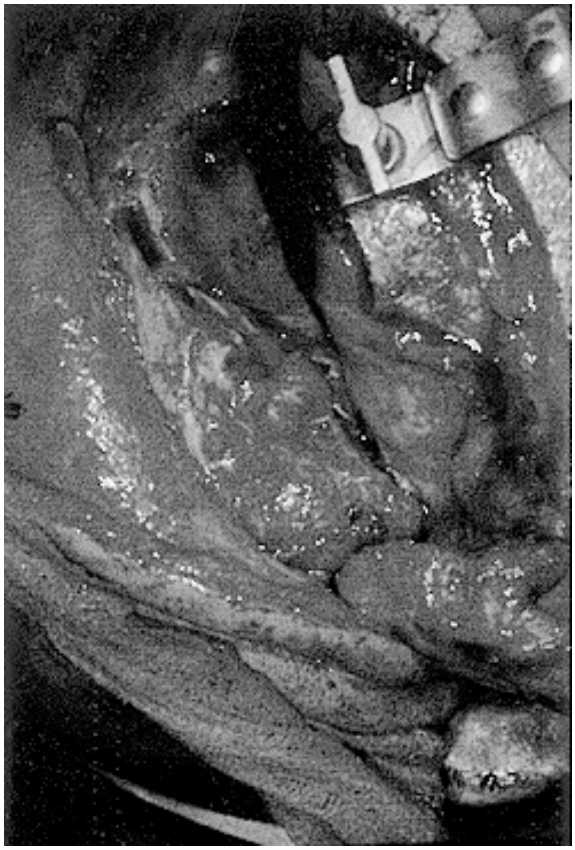

Fig. 3 The defect after the tumour was removed en-bloc and this included the greater wing of the sphenoid, lateral wall of the orbit and the squamo-temporal bone.

Fig. 4 The flap with the vascular pedicle located infero-medially.

and dura covering the frontal and temporal lobes. A biopsy through the previous craniotomy incision was done and this showed that the lesion was an osteogenic sarcoma.

She was seen in consultation with the ENT surgeon and the neurosurgeon and a wide-resection of tumour via a combined neurosurgical and craniofacial approach with immediate one-stage free rectus abdominis flap reconstruction of the resultant defect performed by a multi-disciplinary team was planned. The patient was advised on the potential complications of the operation with particular emphasis on the possibility of optic nerve damage and blindness due to proximity of the tumour to the orbital contents.

Resection of the tumour was achieved in a combined neurosurgical and craniofacial procedure. The tumour was approached via a standard craniofacial bicoronal incision with caudal reflection of the scalp flap to expose the fronto-orbital area where the supra-orbital ridge was osteotomised and laterally through a downward extension of the coronal scalp flap into the pre-auricular region with the removal of the left temporalis and zygomatic arch. The tumour was removed en bloc and this included the greater wing of the sphenoid, lateral wall of the orbit and the squamo-temporal bone (Fig. 3). The tumour was abutting but not involving the periorbita and temporal dura. The globe and its contents were thus preserved.

Reconstruction was necessary to seal off the frontal and maxillary sinuses from dura over the temporal lobe, obliterate dead-space and provide soft tissue bulk for the temporalis region,and to reconstruct the supra-orbital ridge. This was achieved with a calvarial split thickness bone graft reconstruction of the bony supra-orbital bar and a free rectus abdominis muscle flap which was orientated vertically with the vascular pedicle located inferio-medially (Fig. 4). The bone grafts were fixed rigidly with mini plates. Flap vessels were anastomosed end-to-end to the superficial temporal vessels at the level of the helix. The coronal scalp flap was then replaced over the muscle flap and the incisions closed in layers.

Histopathological examination confirmed that the tumour was a high-grade osteogenic sarcoma.

The patient had an uneventful recovery and on the fifth post-operative day, an MRI (Figs. 5, 6) was performed to document flap viability by demonstrating enhancement in the flap in the T2-weighted scan.

She received two cycles of adjuvant chemotherapy with doxorubicin and CDOP. At 12 months post-operatively, the patient was well, grossly free of tumour and continued to enjoy fair vision and an acceptable quality of life.

Fig. 5 MRI taken on the 5th post-operative day.

Fig. 6 MRI taken on the 5th post-operative day.

DISCUSSION

The goals of reconstruction are to achieve primary wound healing, obilterate dead space, seal off sterile cranial areas from contaminated oronasal cavities and to restore aesthetic contour[17,18].

This patient had a combination of a free flap and calvarial bone grafts for supra-orbital bar. The bone grafts were necessary to restore the supra-orbital ridge contour. Where there have been previous attempts at resection, there is a limitation of choice of surgical access and obliterated tissue planes[19]. In instances where a free flap reconstruction had been previously performed, there is a reduced choice of recipient vessels for microvascular anastomosis and limitation of donor site choice. Radiation results in impairment of wound healing and major wound healing complications[20-23] and small vessel changes[24,25], though these have not been shown clinically to have an adverse effect on microvascular anastomosis[26,27]. As this area had previously been irradiated, it was not ideal to use a synthetic implant like silicone or Gortex which would place the patient at high risk of infection.

The rectus abdominis muscle flap is a workhorse flap in reconstructive surgery and in particular, the skull base[28-30]. In this case the rectus abdominis muscle was ideal as it provided well-vascularised tissue with sufficient bulk to obliterate the dead space, it had a long and reliable vascular pedicle which facilitated microvascular anastomosis without the need for vein grafts, and it could be harvested at the same time as the resection, thereby reducing operative time.

We had to rely on hand-held ultrasonography and the use of the MRI on the fifth post-operative day (to demonstrate muscle viability) for the monitoring of this flap which was buried and had no skin paddle. MRI is able to distinguish viable from non-viable tissue, detect vascular occlusions[31] and to evaluate perfusion in muscle flaps[32].

Another use of the MRI following reconstruction for skull base tumour is to serve as a baseline for follow-up for local recurrences. As the original tumour is deep-seated and the resected site is covered by flap tissue and overlying skin flaps, tumour surveillance will have to depend on symptoms and regular imaging with CT and MRI scans.

CONCLUSION

Free tissue transfer enables the surgeon to reconstruct complex defects, especially those that result from surgical ablation of skull base tumours. It enables large portions of vascularised muscle to be imported to provide a water-tight seal between sinuses and brain.

This is a rare case of osteosarcoma of the sphenoid bone that was successfully resected and the resultant defect reconstructed with a combination of free bone graft reconstitution of the bony orbit and free tissue transfer of a rectus abdominis muscle.

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