The Journal of Laryngology and Otology
October 1991, Vol. **105**, pp. 855–857

Giant cell tumour of the sphenoid sinus: An unusual skull base tumour

DAVID UTTLEY, F.R.C.S., DANIEL J. ARCHER, F.R.C.S. (London)

Abstract
An unusual skull base tumour is presented. Intraoperative smears made the diagnosis and dictated a change in surgical strategy. Giant-cell tumours of the sphenoid bone are discussed, together with current management.

Introduction

Skull base tumours are rare and account for less than one per cent of intracranial tumours. There is a rich diversity of pathology as they may arise from a variety of embryonal rests or specialized tissues contained within the basis cranii. Despite this diversity in origin, they tend to behave in a surprisingly similar manner. More often than not they are benign or locally malignant and rarely metastasise. These indolent characteristics contribute to their relative resistance to radiotherapy or chemotherapy. Thus surgical removal is the treatment of choice. Modern imaging techniques such as Computerized Tomography (CT) or Magnetic Resonance Imaging (MRI) readily reveal the presence of these lesions, but the lesions may be of such obscure origin that only an all embracing differential diagnosis is available to the surgeon. The following case report focuses on one of these rare tumours, and suggests a general scheme for dealing with them.

Clinical presentation

The patient is a 27-year-old Greek woman who noted visual loss in the right eye in the early stages of her first pregnancy some seven months earlier, associated with occasional headaches which were worse on stooping and bending. Some six weeks prior to her arrival in the UK for treatment she quite suddenly bcame blind in the right eye overnight. A caesarean section was performed a month later, the product of which was a healthy baby. Postoperative progress was uneventful apart from an impression of failing vision in the left eye, at which point the patient had an MRI investigation, was put on steroids and sent for treatment to the UK.

Examination: General examination was unremarkable apart from a healing caesarean scar. She was blind in the right eye where there was an afferent pupillary defect, and the optic discs were pale bilaterally. Visual acuity on the left side was 6/6 with a full field, but there was evidence of a left 6th cranial nerve palsy. There were no other abnormal neurological findings.

Investigations: Haematological and routine biochemical tests were within normal limits. An MRI scan revealed a striking abnormality in the skull base consisting of a large high signal relatively homogeneous mass arising in the region of the sphenoid sinus and clivus projecting anteriorly into the ethmoid sinus, occupying the pituitary fossa and elevating the gland, and extending into and expanding the clivus sufficiently to displace the pons posteriorly (Fig. 1). The AP scans revealed the crucial information that the carotid arteries and basilar artery were stretched around the periphery of the tumour and not enclosed within it (Fig. 2).

Operation: The optimal exposure of tumours in this situation is by the le Fort I maxillotomy and clivectomy. This approach has been described by the authors for the treatment of basilar aneurysms (Archer *et al.*, 1987) and excision of skull base tumours (Uttley *et al.*, 1989). The arc of surgical exposure is from the middle ethmoids in front down to the cranio-cervical junction posteriorly.

The tumour appeared relatively soft and contained areas which appeared haemorrhagic. These features suggested a degree of malignancy, but a perioperative smear showed that the lesion was a giant cell tumour of the sphenoid. A much more radical approach to the tumour than had been originally intended was adopted on receipt of this news, and a very satisfactory excision was accomplished with margins of what appeared to be normal bone on all four sides and normal dura in the depths of the decompression. There was no overt CSF leakage, the defect was filled with oxidized cellulose and human fibrin glue. The maxilla was repaired with compression plates, and the mucosa with silk sutures. A pharyngostomy was left in place for feeding purposes, and the patient was extubated at the end of the procedure. A lumbar drain was left *in situ* and allowed to freely drain for the next three days.

Postoperative progress was uneventful, with no CSF fistula or diabetes insipidus, there was no additional neurological deficit, indeed the left VIth nerve palsy began to resolve though there was no change in her vision. Twelve days after the operation she developed a temporary depressive state which was thought to be largely puerperal in aetiology, complicated by the stress of major surgery. This rapidly settled and she went on to receive radiotherapy.

Pathology: Microscopic examination revealed the tumour to be comprised of multinucleate giant cells with a number of smaller rounded cells. Areas of bone and osteoid formation were noted and considered to be reactive. No mitotic figures were noted in the prepared material. It was concluded that this was a giant cell tumour of bone (Fig. 3).

Discussion: The very rarity of these tumours and the fact that they are not necessarily aggressive in their behaviour highlights the need for a coherent approach to their management. new imaging techniques have enabled them to be visualized, and their boundaries can be plotted with considerable accuracy, particularly with MR images that delineate the vascular relationships of the tumour, thus reducing the necessity for invasive procedures, such as angiography, with a far from negligible morbidity.

The correlation between radiology and pathology is in its

Accepted for publication: 28 June 1991

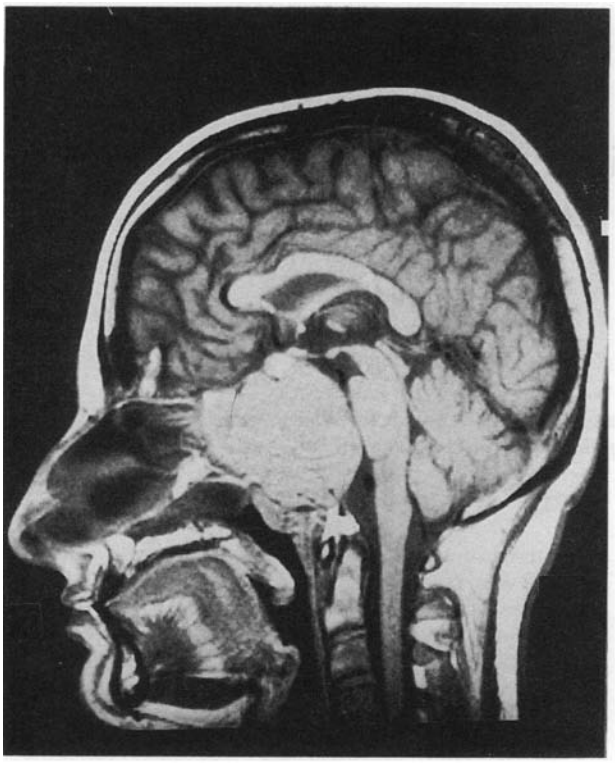

FIG. 1

A large homogenous tumour is seen occupying most of the central portion of the mid-line skull base. The pituitary gland is elevated and lies on top of the tumour, the brain stem is compressed anteriorly by the mass which extends almost down to the cranio-cervical junction.

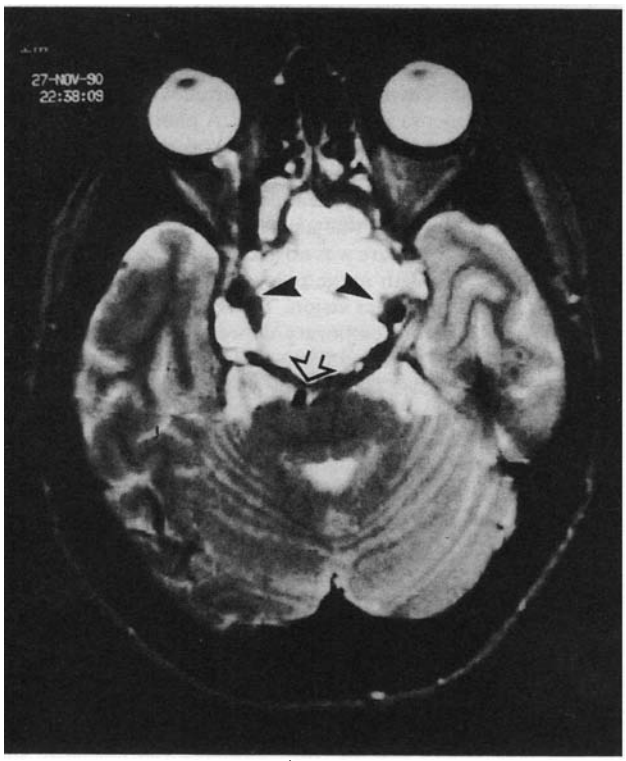

FIG. 2

Tranverse axial MR image of skull base. A large mass is centrally situated in the skull base. The closed arrows demonstrate the internal carotid arteries lying laterally, and the open arrow the basilar artery. The vessels are patent and disposed around the margins of the mass.

infancy in respect of these tumours, and a great deal more experience in this area is required. The acquisition of information on this type of tumour would be accelerated were they to be seen, investigated, treated and reviewed in centres where an active interest in them exists. The fact is that these patients are spread far too thinly between a variety of departments and disciplines which hinders the evolution of a common experience. Even with referral of all such patients to supraregional units, the low incidence with which these lesions emerge would lead to long delays in gathering such essentials as may influence practice, and controlled trials of treatment are inconceivable for the same reason.

The differential diagnosis of these tumours is exhaustive, and a plethora of possibilities interferes with the formulation of an accurate preoperative plan. For tumours of similar radiological appearances some patients may have a biopsy, others radical surgery, and many patients may be subjected to multiple procedures in the primary phase of treatment. With the best will in the world it is impossible to propose a single strategy for management when one of the most important parts of the equation, namely the pathology, remains unknown. In this case report the clinical operative findings suggested a malignant lesion, and without an intraoperative smear, which indicated a hitherto unsuspected and much more satisfactory diagnosis, the ultimate surgical resection would have been much curtailed. In other circumstances the opposite might happen: a palliative approach substituted for radical intervention. In any event we believe intraoperative pathological studies are of vital importance in acting as major arbiters of surgical tactics.

Giant cell tumours of the spheniod bone are exceptionally rare: as far as we can ascertain there are only 36 in the world lit-

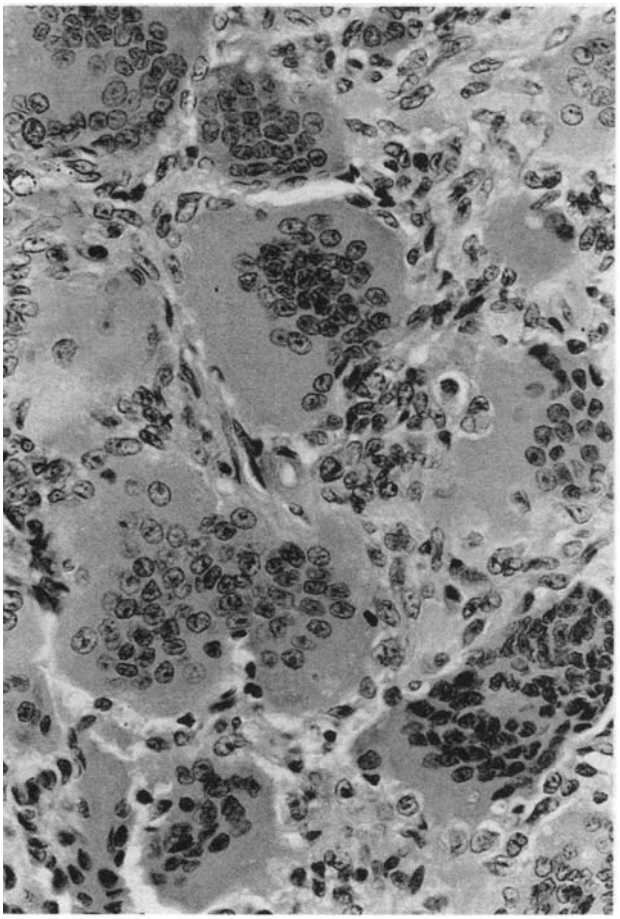

FIG. 3

High power photomicrograph of paraffin section, showing profusion of multinucleated giant cells in the specimen, original magnification ×40.

https://doi.org/10.1017/S0022215100117554 Published online by Cambridge University Press

erature. The largest series comes from the Mayo Clinic (Wolfe *et al.*, 1983), but the presentation is tyupical with headaches, ocular palsy and visual loss (Geissinger *et al.*, 1970; Wolfe *et al.*, 1983), and the duration of symptoms prior to treatment would have been much shorter had not the interval been artificially prolonged by pregnancy.

Many of the tumours are difficult of access, so the need for teamwork in achieving the best exposure is important (jackson *et al.*, 1986). In this case a Le Fort I osteotomy is tailor made for excision of the tumour and residual clivus. We have previously described this approach in connection with the treatment of basilar aneurysms (Archer *et al.*, 1987) and skull base tumours (Uttley *et al.*, 1989) and find it extremely efficacious in terms of exposure from the ethmoids to the cranio-cervical junction. In our hands it is associated with a low morbidity, and in those cases where recurrence appears the method can be repeated with great ease.

In many skull base tumours it is impossible to be certain that a complete excision has been obtained, so that, despite the low radiosensitivity of many of these tumours, radiotherapy has to be considered as a valuable adjunct for the destruction of inaccessible fragments. The role of this treatment modality in the treatment of giant cell tumours is undergoing a reappraisal. Reports in the 1970s recorded local failure rates of 40–60 per cent following radiotherapy (Dahlin *et al.*, 1970; Goldenberg *et al.*, 1970; McGrath, 1972), and this was associated with high rates of malignant transformation varying from 7 to 25 per cent (Goldenberg *et al.*, 1970; McGrath, 1972). this depressing scene has been challenged recently by another report, which fails to note either failures in tumour control or increased aggressiveness in behaviour in a series of 15 patients with a mean follow-up of 12 years (Bell *et al.*, 1983). This major improvement in response has been attributed to an increasing sophistication in radiotherapy techniques allied to precision in imaging (Schwartz *et al.*, 1989).

Although giant cell tumours are rare we believe that aggressive surgery followed by radiotherapy is the optimum current treatment, and that these measures may halt the progress of the disease for an indefinite period.

Key words: Paranasal sinus neoplasms; Giant cell tumour

Acknowledgements

We wish to thank Miss S. Rostron for her care and skill in the preparation of this manuscript.

References

Archer, D. J., Young, S., Uttley, D. (1987) Basilar aneurysms: a new transclival approach via maxillotomy. *Journal of Neurosurgery,* **67:** 54–58.

Bell, R. S., Harwood, A. R., Goodman, S. B. (1983) Supervoltage radiotherapy in the treatment of difficult giant-cell tumours of bone. *Clinical Orthopaedics and Related Research,* **174:** 208–216.

Dahlin, D. C., Cupps, R. E., Johnson, E. W. (1970) Giant-cell tumour. A study of 195 cases. *Cancer,* **25:** 1061–1070.

Geissinger, J. D., Sigueira, E. D., Ross, E. R. (1970) Giant-cell tumours of the sphenoid bone. *Journal of Neurosurgery,* **32:** 665–670.

Goldenberg, R. R., Campbell, C. J., Bonfiglio, M. (1970) Giant-cell tumour of bone. *Journal of Bone and Joint Surgery,* **52A:** 619–664.

Jackson, I. T., Marsh, W. R., Bite, U., Hide, T. A. H. (1986) Craniofacial osteotomies to facilitate skull base tumour resection. *British Journal of Plastic Surgery,* **39:** 153–160.

McGrath, P. J. (1972) Giant-cell tumour of bone. *Journal of Bone and Joint Surgery,* **54B:** 216–224.

Schwartz, L. H., Okunieff, P. G., Rosenberg, A. (1989) Radiation therapy in the treatment of difficult giant-cell tumours. *International Journal Radiation Oncology, Biology and Physics,* **17:** 1085–1088.

Uttley, D., Moore, A., Archer, D. J. (1989) Surgical management of mid-line skull base tumours: A new approach. *Journal of-Neurosurgery,* **71:** 705–710.

Wolfe, J. T., Scheithauer, B. W., Dahlin, D. L. (1983) Giant-cell tumour of the sphenoid bone: Review of 10 cases. *Journal of Neurosurgery,* **59;** 322–327.

Address for correspondence:
David Uttley, F.R.C.S.,
Department of Neurosurgery,
Atkinson Morley's Hospital,
Wimbledon,
London SW20 0NE.