Giant Cell Tumor of the Sphenoid Bone

RASIKBALA DOSHI, ABDUL BASIT CHAUDHARI AND GORDON THOMSON

SUMMARY: *The clinical and histological features of two cases of giant cell tumor of the sphenoid bone are described. Both presented with similar symptoms and signs, comparable to previously described cases. The problems in histological differential diagnosis are discussed and radiotherapy as the treatment of choice is suggested.*

RÉSUMÉ: *Les données cliniques et histologiques d'une tumeur à cellules géantes de l'os sphénoïde sont décrites pour 2 patients. Les deux cas présentaient des symptômes et signes similaires, produisant un syndrome clinique comparable aux cas décrits auparavant. Les problèmes de diagnostic différentiel histologique sont discutés et la radiothérapie est fortement suggérée comme traitement de choix.*

From the Burden Neurological Laboratory Departments of Neurosurgery and Neuroradiology, Frenchay Hospital, Bristol.

Reprint requests to: Mr. A. B. Chaudhari, Department of Surgery, Lagos University Teaching Hospital, Surulere, PMB 12003, Lagos, Nigeria.

INTRODUCTION

The primary osseous tumors arising from the cranium have received little attention because of their rarity. Coley (1960) reported the incidence of primary cranial osseous tumors as 2 - 2.8% of all bone tumors. However, the spectrum of histological variety of osseous neoplasms is the same in the cranium as elsewhere in the skeleton. The relative rarity of the cranial tumors is the most conspicuous feature. These tumors are characterized by specific syndromes related to their customary locations. Among the primary cranial osseous neoplasms, giant cell tumor of the sphenoid bone is probably the rarest. Geissinger et al. (1970) found 11 cases in a review of the world literature. They added one new case.

This paper reports two cases of giant-cell tumor arising in the sphenoid bone and the purpose of the paper is to emphasize the importance of the clinical syndrome produced by the tumor, the value of histological diagnosis and the beneficial effect of radiotherapy.

CASE REPORTS

Case 1:

M. E. (F. B. 117/52) female, twenty-one was admitted to Frenchay Hospital, Bristol, U.K. in January 1952 with a six month's history of severe frontal headache and amenorrhea. She also had diplopia and ptosis for three months and five weeks respectively. Her general examination was unremarkable, but she had complete ophthalmoplegia externa on the left and the visual acuity in the left eye was limited to perception of hand movements in the periphery. She was completely blind in her right eye. There was hyperaesthesia over the left side of the chin, cheek and nose. An X-ray of the skull showed partial destruction of the dorsum sellae and the posterior part of the floor of the pituitary fossa.

A diagnosis of malignant primary tumor arising in the sellar region with rapid involvement of the optic nerve and chiasm was made. A right frontal craniotomy was performed in order to explore the chiasmal region, to decompress the optic nerves and to provide the material for histological examination.

Pathology:

The surgical specimen consisted of fragments of semi-solid tissue with blood clots. Paraffin blocks were prepared and sections were stained with hematoxylin and eosin, and by Gomori's method for reticulin. The chromatin pattern of nuclei of the stromal cells was surrounded by well developed pericellular reticulin. The chromatin pattern of the nuclei of the stromal cells was identical to that of the giant cell nuclei and mitotic figures were scant. A diagnosis of giant cell tumor of the sphenoid bone was made on the basis of these sections (Fig. 1).

Radiotherapy was started and a total dose of 3010 rads over a period of four weeks was given.

The patient remained in good health post-operatively and on reassessment six months later she had considerable improvement of the vision in her left eye. Fifteen years after the operation she had normal vision in the left eye and the right could perceive light in the periphery of the nasal half of the field. Her menstrual periods recommenced seven months after irradiation and she had a child four years after the course of radiotherapy.

https://doi.org/10.1017/S0317167100025257 Published online by Cambridge University Press

Case 2:

C. P. (N. P. 2697) female, aged eighteen years, was admitted to Frenchay Hospital, Bristol, U.K. in March 1972 with a year's history of severe throbbing occipital headaches with a frequency of three to four attacks per week. Six months previously she had had a rash which was thought to be rubella and since then she complained of a painful left eye, diplopia, photophobia and drooping of the left eyelid. Three months prior to her admission her headaches ceased and she stopped menstruating. Her general examination was unremarkable, but there was some proptosis of the left eye with almost complete ophthalmoplegia externa and interna with the exception of feeble and sluggish reaction of the left pupil to light while the right pupil was normal. She had hyperaesthesia over the distribution of the maxillary and ophthalmic divisions of the left trigeminal nerve. Her visual acuity was J20 on the left and J2 on the right.

Investigations:

The pre-operative investigations were as follows: Hb. — 13.8G% and plasma viscosity 1.64. Cerebrospinal fluid was clear and no cells were present. The protein was 49 mg%. Pregnancy test was negative. Plasma cortisol at 9. a.m. was 21.4 ug/100 cc and at midnight 10.0 ug/100 cc. Serum thyroxine was 4.8 ug/100 cc.

Skull X-rays revealed erosion of the posterior part of the floor of the pituitary fossa and poor definition of the posterior clinoids. The sphenoidal sinus was clouded, and in the tomograms a clear cut soft tissue mass of about 3 cm. in diameter with a rounded anterior margin could be seen. A left carotid angiogram showed straightening of the inferior limb of the siphon consistent with a mass spreading laterally from the pituitary fossa region, and also an area of pathological circulation across, above and behind the anterior limb of the siphon was seen.

A left fronto-temporal craniotomy was performed. An extradural tumor involving the sphenoid bone and cavernous sinus was found and was partially removed.

Pathology:

The smears received during surgery were stained with toluidine blue and the predominant feature was the presence of multiple giant cells and some mononuclear cells. The final diagnosis was deferred until the paraffin sections were available, but a non-malignant neoplasm arising in bone was suggested.

The biopsy specimen consisted of multiple portions of grey polypoidal tissue, together weighing 5 G. Cryostat sections were cut and stained by red sudan dye, and the remaining material was embedded in paraffin. Sections were stained with hematoxylin and eosin, Van Giesson hematoxylin, Gomori silver impregnation, and by the periodic-acid Schiff technique. Sections showed a tumor (Fig. 2) composed of stromal cells and multinucleated giant cells lying in a moderately vascular stroma. The prominent feature was the presence of numerous multinucleated giant cells with 10-60 nuclei situated at the center of these cells with abundant eosinophilic cytoplasm. The stromal cells were mononuclear, spindle or ovoid in shape with indistinct cytoplasm. The

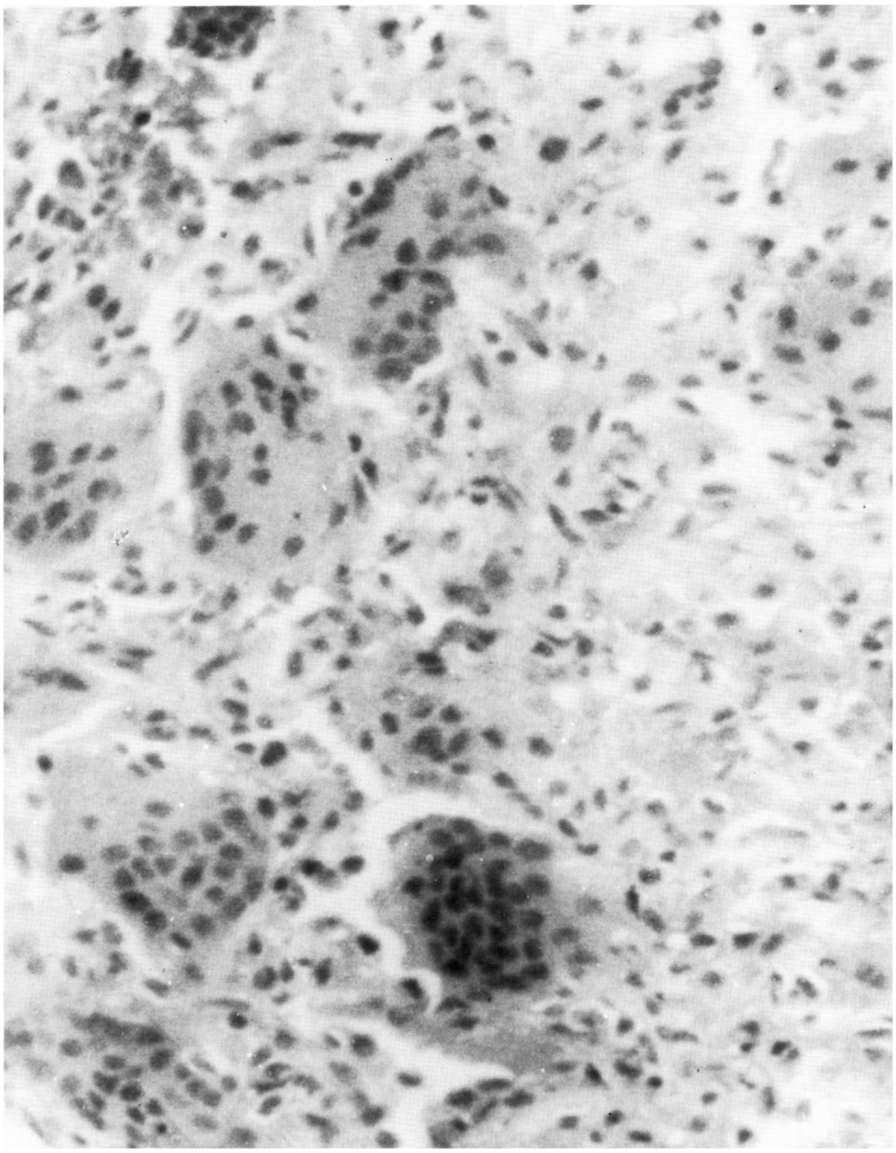

Figure 1—Case 1. Tumor from 1952 biopsy showing multinucleated giant cells and stromal cells. Hematoxylin and eosin X 300.

https://doi.org/10.1017/S0317167100025257 Published online by Cambridge University Press

chromatin pattern of the nuclei of the stromal cells was similar to that of giant cell nuclei and a few mitoses were present. A characteristic well developed pericellular reticulin pattern was demonstrated by the reticulin stain (Fig. 3). No intracytoplasmic fat was present.

A diagnosis of giant cell tumor of bone was made. Before post-operative treatment was started her parathyroid function was assessed. These revealed a serum calcium of 10.1 mg/100 cc, inorganic phosphate 3.2 mg/100 cc and alkaline phosphatase 75 i.u./litre.

Post-Operative Progress:

Radiotherapy was started and she received a total dose of 3400 rads in 27 days after discharge.

On reassessment, six months, eighteen months and four years later, she complained of occasional occipital headaches but she had gained one stone in weight, was feeling much better in herself and had started menstruating. There was improvement in her visual acuity and external ocular movements and the dimensions of the zone of hyperaesthesia had diminished.

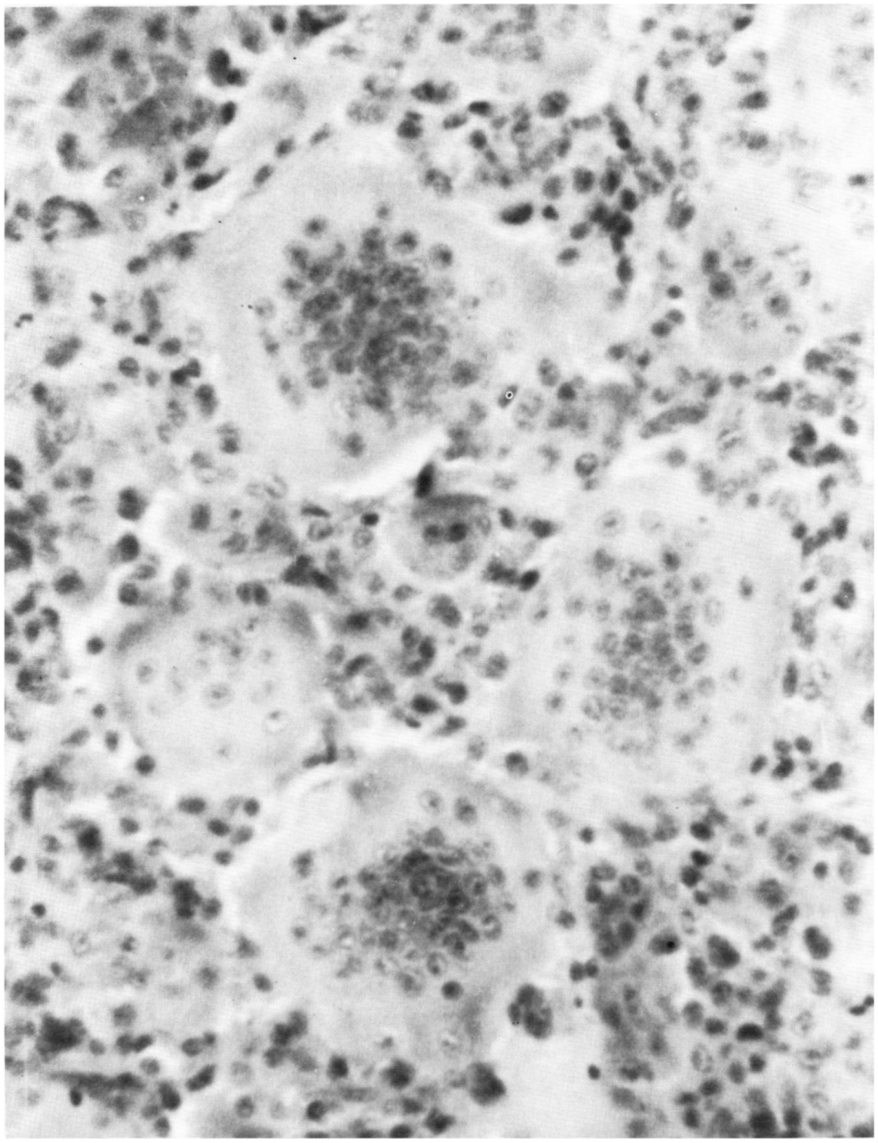

Figure 2—Case 2. Tumor from biopsy showing multinucleated giant cells with nuclei congregated at their center. Round to oval stromal cells are also present. Hematoxylin and eosin X 300.

DISCUSSION

The involvement of II, III, IV, V and VI cranial nerves is the most characteristic feature of these lesions because of their close association with the sphenoid bone. The clinical picture in the two cases is similar. This almost stereotyped pattern is apparent in all the reported cases reviewed by Geissinger et al. (1970). Endocrine symptoms were rare in some reported cases, but were present in the cases of McNerney (1949), and Pitkethley and Kemp (1969), although only post-operatively in the latter case. In the present cases, however, amenorrhea was an early feature and an important symptom in two young women.

Giant cell tumor of bone has been recognized as arising in the epiphysis of a long bone in young adults after the epiphysial-metaphysial union is complete. Although this tumor has been described in other skull bones, the sphenoid bone is a relatively uncommon site. The sphenoid bone develops from pre-sphenoid and post-sphenoid parts, which ossify from several ossification centers in preformed cartilage. These areas are comparable to the ends of long bones and thus provide a favorable site for the genesis of giant cell tumor. The average age of onset is 25 years, but there are examples of much younger and older cases. Echols (1945) reported a case of 12 years of age and Geschickter (1949) reported a case of 52 years. There is a definite female preponderance. Of the twelve cases reported in the world literature, eight were females and three were males. The sex and age of one case was not stated. The present cases bring the total to ten females in a total of thirteen.

The histological findings in both cases were identical. The tumor was composed of a variable number of giant cells interspersed between the stromal cells. The number of nuclei in the giant cells varied from 10-60 and the chromatin pattern of the nuclei of the stromal cells was similar to the nuclei in the giant cells. Prominent histological features were well developed pericellular reticulin

and few mitoses. No pleomorphism of the nuclei was present and both tumors were classified as Grade I (Lichtenstein, 1965). Hutter et al. (1962) suggested that if the growth rate was rapid there would be fewer giant cells and vice-versa. In both specimens the presence of many giant cells plus other benign characteristics gave a clear indication of a slow and benign course.

Before making a firm histological diagnosis of giant cell tumor of bone other conditions such as hyperparathyroidism, fibrous dysplasia, histiocytosis-x and non-ossifying fibroma must be excluded. Various histochemical tests such as the presence of acid phosphatase, acid mucopolysaccharides and absent lipid in cytoplasm have been described to differentiate giant cell tumor of bone from histologically similar conditions. None of these tests are absolutely specific. However, for the diagnosis of this tumor when it arises from the sphenoid bone, it is essential to cut cryostat sections before paraffin embedding in order to do lipid and P.A.S. stains to exclude histiocytosis-x. The presence of pericellular reticulin distinguishes it from the bony lesion of hyperparathyroidism, but estimations of serum calcium, phosphate and alkaline phosphatase are equally important to exclude this condition. On clinical and radiological grounds alone a precise diagnosis cannot be made, although certain features can be suggestive. The mainstay of diagnosis is histological examination without which appropriate treatment cannot be instituted. Success in the management depends on prompt histological identification and early institution of radiotherapy. One of the cases described (M. E.) showed an extremely good response to radiotherapy without recurrence or malignant change after nearly twenty years. The second case after a course of radiotherapy fared very well. There are numerous complications and problems associated with radical surgery because of the situation of the tumor and involvement of adjacent vital structures. In view of the results obtained in these two cases and the possible dangers inherent in other treatments, we think radiotherapy is the treatment of choice following diagnostic surgical biopsy.

ACKNOWLEDGMENTS

We wish to thank Mr. Huw Griffith of Frenchay Hospital, Bristol, U.K. for permission to quote details of the clinical histories and Dr. Betty Brownell from the same hospital, for her encouragement in writing the paper. We are also grateful to the photographic department of Frenchay Hospital who kindly carried out the photographic work.

REFERENCES

COLEY, B. L. (1960). Neoplasms of Bone and Related Conditions. Hoeber, 2nd ed., New York, p. 496.

ECHOLS, D. H. (1945). Giant Cell Tumors of the Sphenoid Bone: Report of a Case. J. Neurosurg. 2, 16.

GEISSINGER, J. D., SIQUEIRA, E. B. and ROSS, E. R. (1970). Giant Cell Tumors of the Sphenoid Bone. J. Neurosurg., 32, 665.

GESCHICKTER, C. F. and COPELAND, M. M. (1949). Tumors of Bone, Lippincott, 2nd ed., Philadelphia, p. 640.

HUTTER, R. V. P., WORCESTER, J. Jr. and FRANCIS, K. C. (1962). Benign and Malignant Giant Cell Tumors of Bone: A Clinicopathological analysis of the natural history of the disease. Cancer, 15, 653.

LICHTENSTEIN, L. (1965). Bone Tumors, Mosby, 3rd ed., London, p. 136.

McNERNEY, J. C. (1949). Giant Cell Tumors of Bones of the Skull. J. Neurosurg., 6, 169.

PITKETHLEY, D. T. and KEMPE, L. G. (1969). Giant Cell Tumors of the Sphenoid. J. Neurosurg., 30, 301.

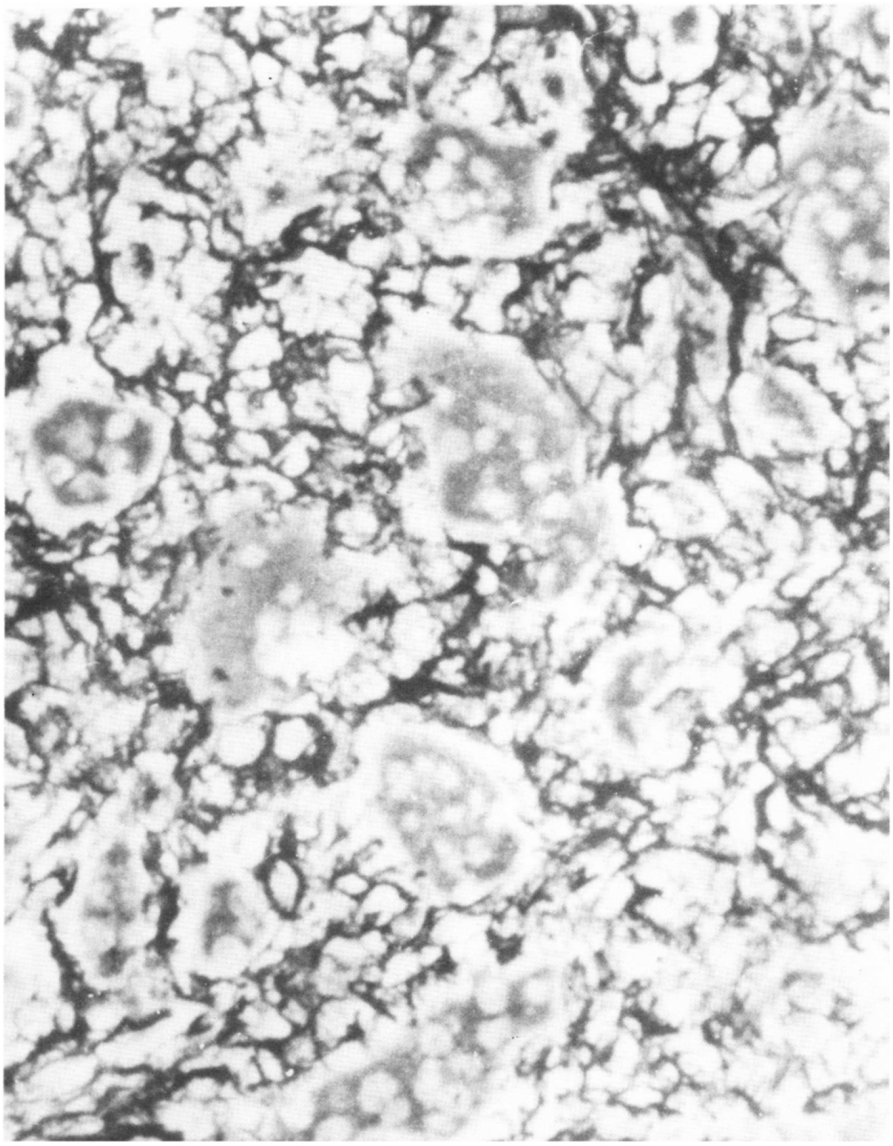

Figure 3—Case 2. A prominent reticulin network is present about blood vessels and individual tumor cells. Gomori's reticulin method X 300.