CASE REPORT

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Radiological difficulties in the diagnosis of fibrous dysplasia of the sphenoid sinus and the cranial base*

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SUMMARY

Fibrous dysplasia is a benign disorder of bone. We present the case of a 26-year-old man with fibrous dysplasia of the sphenoid sinus and the cranial base. The presentation and management of this rare condition are discussed highlighting the potential for misdiagnosis with MRI.

Key words: CT scan, MRI, monostotic fibrous dysplasia, paranasal sinuses, skull base

INTRODUCTION

Fibrous dysplasia (FD) is a term used to describe a group of benign bony lesions, previously designated by such varying titles as osteogenic or ossifying fibroma, fibrous osteoma or osteofibroma (Lichtenstein, 1938).

Histologically, FD is characterised by two elements: the first component being fibrous tissue with relatively uniform spindle-shaped fibroblastic cells; the second component usually consists of irregular trabeculae of woven bone without regular cement lines. Osteoblasts and osteoclasts are minimal or absent. The ratio of fibrous to osseous tissue varies from case to case and from area to area within the same lesion. Residual normal or reactive bone is occasionally present at the margins of the lesion (Fu and Perzin, 1974).

The craniofacial skeleton is involved in 10-27% of patients with the monostotic form and in 50% of those with the polyostotic form. Sites of predilection in the head and neck include the maxilla, mandible, frontal, ethmoid, sphenoid and temporal bones (Bibby et al., 1994). The onset of FD is usually in late childhood, with a female preponderance, although the clinical progression is unpredictable with many individuals remaining asymptomatic. Disease progression usually stops with the onset of puberty (Hirabayashi et al., 1998)

Fries (1957), described the plain film radiographic characteristics of craniofacial FD. Three types of X-ray appearance were reported, depending on the relative proportions of fibrous and osseous tissues:

1) Pagetoid: in which there is expansion of the involved bone, with alternating areas of relative density and lucency.
2) Sclerotic: with a homogenous density.
3) Cystic: with relative central lucency and dense margins.

CASE REPORT

A 26-year-old Caucasian male presented to the neurology department with a 4 month history of deep-seated headaches, photophobia in the right eye and a single episode of a generalized tonic clonic seizure. The neurological examination, lumbar puncture and initial ophthalmological examination (including visual acuity, fundus examination and visual field testing) were all normal. An E.E.G showed markedly abnormal irregular sharp and slow waves in the frontal region suggesting left frontal lobe pathology. A MRI scan revealed a large mass lesion replacing much of the normal marrow fat within the clivus and basisphenoid.

The lesion was of mixed signal intensity on both T1 and T2 sequences, with focal areas of hyperintensity. The sphenoid sinus was almost completely non-pneumatised, but the pneumatisation of the ethmoids was normal. On the right side the lesion extended inferiorly with apparent involvement of the pterygopalatine fossa, infratemporal fossa, and nasopharynx. The lesion appeared to be surrounding the intrapetrous-internal carotid artery bilaterally but with no vessel effacement. There was no evidence of retro-clival or brain stem involvement (Figures 1 A and 1 B). These findings suggested a diagnosis of a destructive skull base tumour, possibly a nasopharyngeal carcinoma, or even a chordoma.

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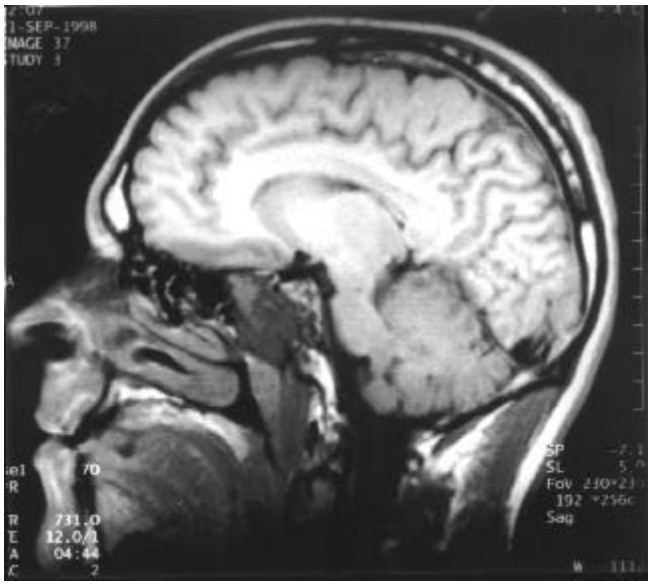

Figure 1A. Sagittal T1 weighted image showing a lesion of the sphenoid and clivus of mixed intensity with replacement of marrow fat.

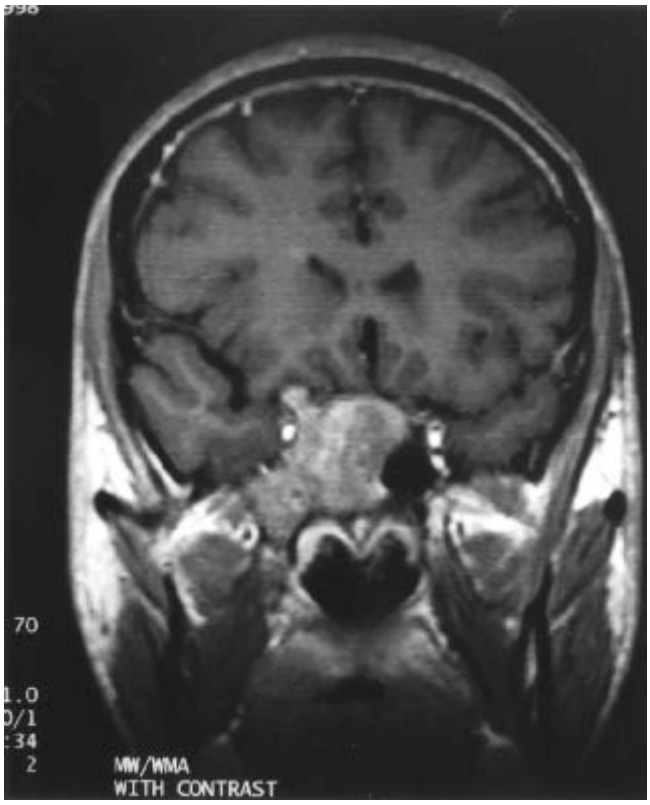

Figure 1B. Coronal T1 weighted image with contrast showing expansion of the sphenoid and planum sphenoidale with heterogenous enhancement.

The patient was subsequently referred to the ENT department for an endoscopic nasopharyngeal biopsy. A high resolution CT scan of the skull base with both coronal and axial images was then performed prior to an endoscopic biopsy. This showed an abnormality of the bone texture associated with expansion but with preservation of the cortical outline. The lesion was confined to bone and spread into the planum shenoidale, anterior and posterior clinoids and posterior ethmoids. The ground glass appearance was suggestive of a FD of the sphenoid and skull base (Figures 2A and 2 B). Nasal endoscopy was performed under general anaesthetics. The left sphenoid sinus ostium was negotiated revealing a considerably contracted left sphenoid sinus. The right sphenoid sinus ostium could not be identified. Biopsy of the face of the right sphenoid sinus yielded hard bone and uncapped a small mucocele, which was visible on the CT scan (Figure 2 B). The histology of the biopsied bone was consistent with fibrous dysplasia with no evidence of malignancy. The patient developed a progressive deterioration of vision in the right eye within a few months of presentation and had a right frontotemporal craniotomy and decompression of the right optic nerve with subsequent improvement in his visual field.

DISCUSSION

The MRI scan of our patient was initially misleading. There was a mixed signal intensity on both the T1 and T2 sequences. Inflammatory sinus lesions usually demonstrate an increased signal on T2 - weighted images. A loss of a T2 signal, within the sinuses, usually indicates an aggressive lesion such as a neoplasm or dried secretions (Shapiro and Som, 1989).

A MRI scan does however give useful information in cranial FD such as the evaluation of the impact of bony changes on neighboring soft-tissue structures, detection of sinus mucoceles (high signal intensity on T2-weighted image), and exclusion of an adjacent meningioma as a possible source of hyperostosis.

High-resolution CT is now the imaging method of choice for evaluation of FD. Widening of diploic spaces, osseous expansion, narrowing of neural and vascular foramina, and the characteristic ground-glass appearance are characteristic (Wayne Ham et al., 1998). Either CT internal attenuation coefficients, or Hounsfield units (HU), are used to differentiate FD from other benign bony dysplasias. FD lesions range from 70 to 130 HU compared with 20 to 40 HU for other benign lesions (Resnick, 1995).

The diagnosis of FD of the paranasal sinuses and cranial base is usually radiological. A number of authors, however, recommend biopsy to differentiate the lesion from other fibro-osseous disorders and to exclude malignant degeneration. (Ikeda et al., 1997) advocate an endoscopic approach over an external approach with biopsies of FD of the paranasal sinuses.

Coley and Stewart (1945) were the first to report malignant degeneration of FD. It is a rare but recognized complication of this disease. Diagnosis of malignant degeneration is made from a combination of biopsy and CT scanning which shows an osteolytic mass in a lesion that previously exhibited the classic features of FD. There appear to be no reports of malignant degeneration of FD of the sphenoid sinus.

It has long been considered that radiotherapy in patients with fibrous dysplasia plays an important role in malignant degeneration (Chetty et al., 1990; Campanacci et al., 1978; Schwartz and Alpert, 1964). There are, however, numerous reports of

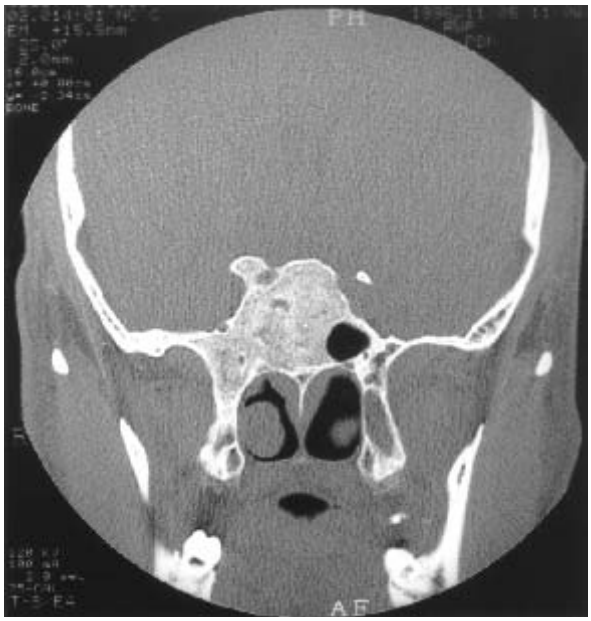

Figure 2A. High-resolution coronal CT scan showing the lesion with a ground glass appearance replacing most of sphenoid bone.

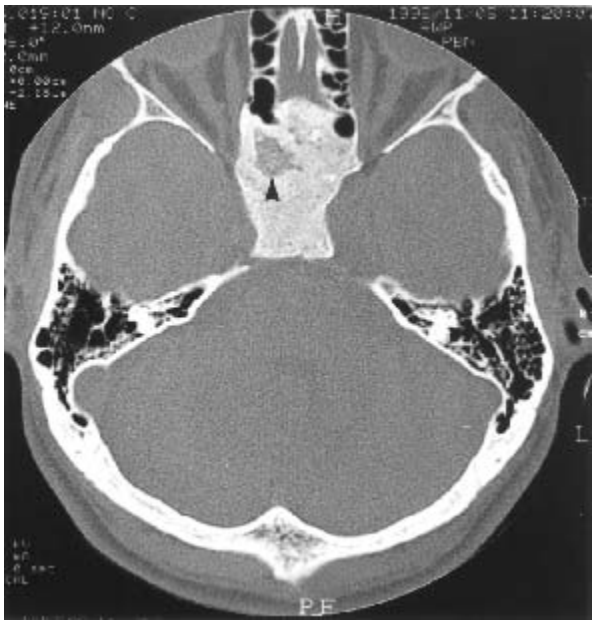

Figure 2B. High resolution axial CT through skull base showing non-pneumatisation of sphenoid with an area of soft tissue density (arrow) within anterior area of right sphenoid sinus confirmed as a mucocele at biopsy.

malignant transformation, with no preceding history of radiotherapy (Cheng and Chen, 1997; Schwartz and Alpert, 1964; Ishida et al., 1992).

Observation appears to be the best approach for patients with craniofacial FD who have minimal or no symptoms. Curettage of lesions may be followed by a high rate of recurrence, but the consensus for the treatment of symptomatic FD consists of surgical resection, particularly if there is optic nerve compression or any neurological deficit. Total resection of the craniofacial bones involving FD has been reported. The necessity for surgery and the extent of surgical resection must be based on the location of the disease, the severity of symptoms, and the biological behaviour of the lesion.

CONCLUSION

Fibrous dysplasia of the sphenoid sinus is a very rare disease. There are no classically associated symptoms, but deep-seated headaches and visual disturbance are often described. MRI on its own may give a misleading picture of a destructive malignant process. The diagnosis is best reached through a combination of CT, MRI and biopsy. Fibrous dysplasia of the sphenoid sinus is best observed with serial imaging if necessary and excision reserved for cases with severe symptoms.

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