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Pathology in Focus

Desmoplastic fibroma of the temporal bone

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Abstract
An unusual case of a desmoplastic fibroma of the temporal bone is presented. Although classified benign, this intraosseous lesion exhibits local aggressiveness and has a high potential for recurrence. This rare condition occurs predominantly in the mandible and in the long bones and is seldom seen in the calvarium. Here we add another case to the previously described eight in the skull and this is the fourth such case reported in the temporal bone. The clinical features, radiology, histopathology and the therapeutic considerations of this lesion in a 72-year-old female are discussed. In addition, a literature review of all the cases affecting the skull bones is presented.

Key words: Fibroma bone neoplasms; Temporal bone

Introduction

Desmoplastic fibroma of the bone is a locally aggressive benign tumour of fibrous tissue origin.[1] This distinctive and rare neoplasm biologically and histologically mimics the more common soft tissue desmoid tumour. It was first described in the bone by Jaffe[2] in 1958 who differentiated it as an entity distinct from other types of intraosseous fibrous tumours. Histologically, the lesion is characterized by thick wavy collagen bundles and few fibroblasts with small oval nuclei and no atypia.[3,4]

Nearly 200 cases of desmoplastic fibroma have been described in the literature. The most common sites include metaphyses of long bones and mandible (70 per cent).[5] Rare sites include maxilla, skull bones, sternum and the vertebral column. Only eight cases have been described in the calvarium and of these only three have been in the temporal bone.[6,7] Here we present another case of desmoplastic fibroma of the temporal bone in a 72-year-old woman. The clinical presentation, radiological features and histopathology of the condition are discussed in detail. Also presented is a brief review of the literature pertaining to the disease affecting the skull bones.

Case report

A 72-year-old woman presented to the Neurotology Clinic at the Queen Elizabeth Hospital, Birmingham with a history of swelling in the right temporal region above the ear for nearly 10 years. The swelling was painless and constant in size until six months prior to ENT consultation, when she noticed a gradual increase in size with it becoming increasingly difficult to lie on her right side. The patient denied any previous history suggestive of ENT disease. Her past medical history included carcinoma of the cervix treated by surgery and radiotherapy 40 years ago, carcinoma of the bronchus treated by pneumonectomy 36 years ago and carcinoma of the breast treated by lumpectomy and tamoxifen nine years ago. Examination revealed a large irregular and painful protrusion of the right squamous temporal bone extending into the right mastoid region. Otoscopy, neck examination and a thorough neurological examination were all unremarkable.

A high definition computerized tomographic scan of the right temporal region showed a large triangular bony defect involving the right lateral petrous temporal and adjacent squamous and mastoid parts. Soft tissue seen within the bony defect extended anteriorly to result in some narrowing of the external auditory meatus. Middle and inner ear structures were spared. The lesion involved predominantly the outer table of the skull, expanding the diploic space and partly destroying the inner table of skull (Figure 1). There was a small area on the inner table of the postero-lateral aspect of the temporal bone where an intracranial extension could not be excluded. A magnetic resonance image (MRI) scan performed with gadolinium enhancement confirmed the intracranial extension. This showed an enhancing soft tissue mass, which appeared to have breached the dura mater in a small area with an intracranial extension to the temporal lobe (Figure 2). An open biopsy of the lesion was performed which was reported as a desmoplastic fibroma of the bone.

Surgical procedure

A complete excision of the tumour by a lateral temporal bone resection with repair of the dural defect and a split calvarial graft cranioplasty was performed with the assistance of the maxillofacial surgeons. The procedure involved a subgaleal exposure of the tumour (Figure 3) and the entire lateral temporal bone followed by closure of the external auditory canal. An extended cortical

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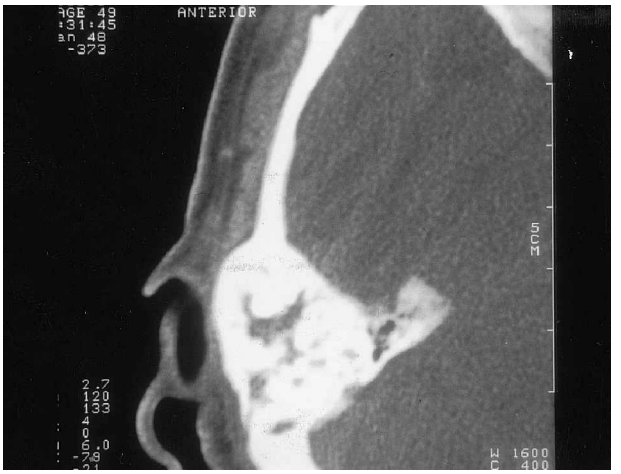

FIG. 1

Axial, high definition CT scan showing the tumour in the right temporal bone, involving predominantly the outer table of the skull, expanding the diploic space and partly destroying the inner table of skull.

mastoidectomy was performed with removal of the middle-ear contents apart from the stapes. Additional craniotomies were performed to excise tumour from both the middle and posterior fossa extradural spaces. The tumour had breached the middle fossa dura over a small area and had extended on to the surface of the temporal lobe of the brain. The tumour was excised completely with the craniotomies and lateral temporal bone resection. The dural defect was repaired with a pericranial patch and fibrin glue and the skull defect was closed with a split calvarial graft.

During the post-operative period she had one episode of seizures and was commenced on anti-epileptic medication. She also developed a wound CSF leak, which was treated by re-suturing and pressure dressings. This resolved subsequently. She was discharged home on the 10th post-operative day. The patient had been well and showed no sign of recurrence of this tumour for six months. However, she was subsequently admitted to the medical ward with an acute onset of ascites and general malaise. A computed tomography (CT) scan revealed a hitherto undiagnosed ovarian tumour with liver metastases. The patient died from carcinomatosis shortly after admission.

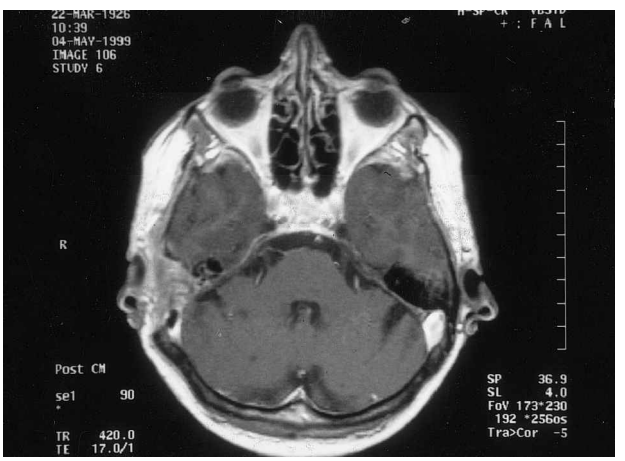

FIG. 2

An axial T_1-weighted MRI scan with gadolinium enhancement showing an enhancing soft tissue mass in the right temporal bone, which appears to have breached the dura mater in a small area with an intracranial extension to the temporal lobe.

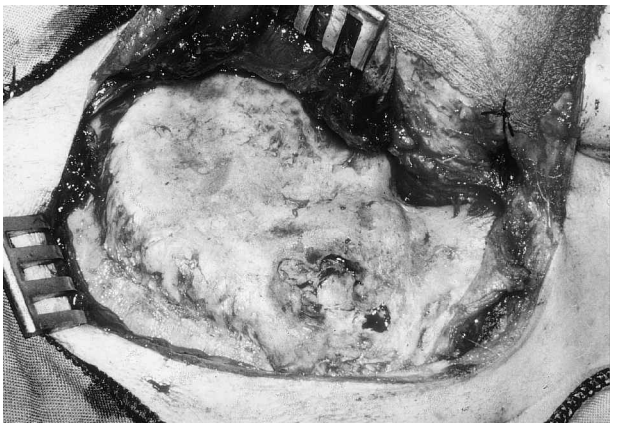

FIG. 3

An operative photograph of the lesion showing a large, irregular, greyish tumour in the squamous and petromastoid parts of the right temporal bone.

Histopathology

The specimen was received in five parts consisting of two portions of muscle, a portion of greyish-white tissue 2.3 cm in diameter attached to bone, right temporal bone measuring 7.5 × 4.0 × 0.8 cm in thickness and a portion of white tissue 2.5 × 1.5 × 1.0 cm.

The lesional tissue consisted of elongated spindle cells with fascicular architecture and paucicellular areas (Figure 4). The proliferating spindle cells had bland ovoid nuclei and the collagen fibres had a wavy pattern with entrapped fragments of lamellar bone (Figure 5). Mitoses were not seen. Abundant collagen fibres were present that were focally hyalinized. The fibroblastic tissue showed a deeply

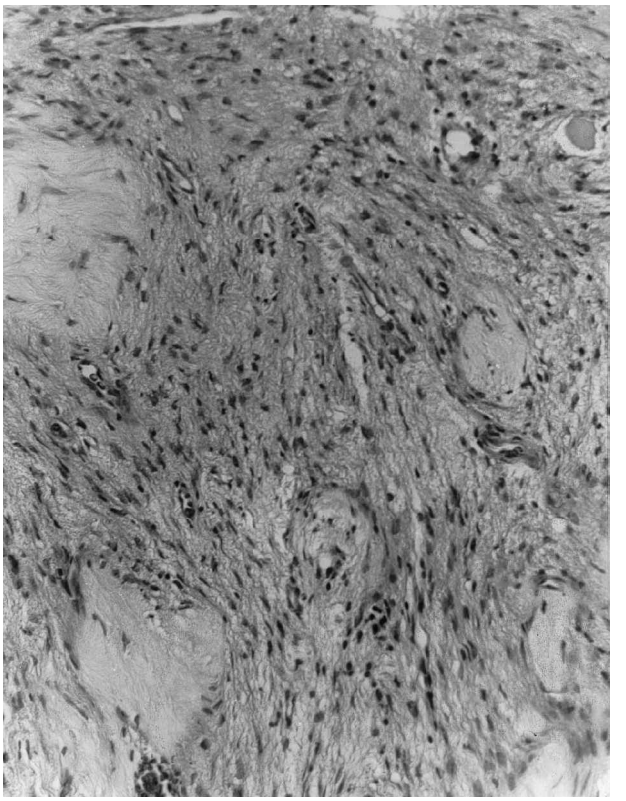

FIG. 4

Spindle cells arranged in compact fascicles alternating with loosely cellular areas (H & E; ×171).

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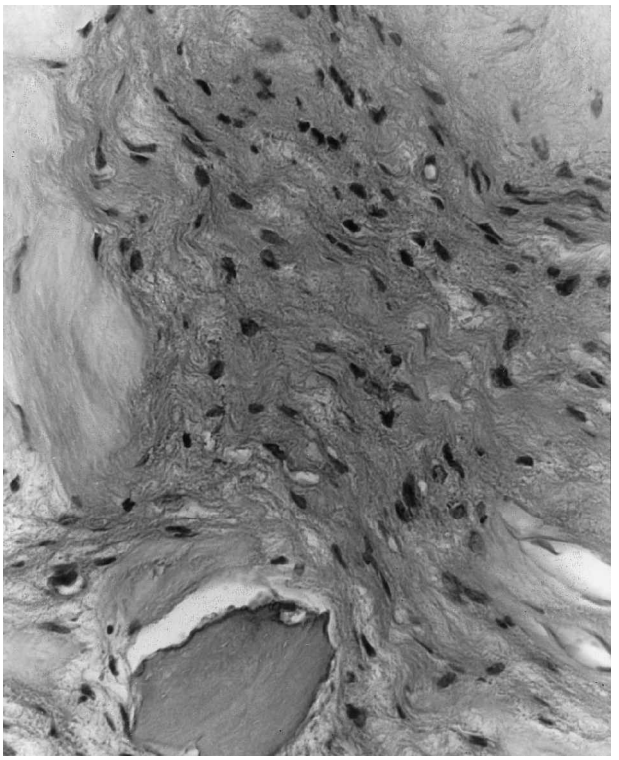

FIG. 5

Spindle cells with bland ovoid nuclei interspersed between wavy collagen fibres. A remnant of bone is present (H & E; ×342).

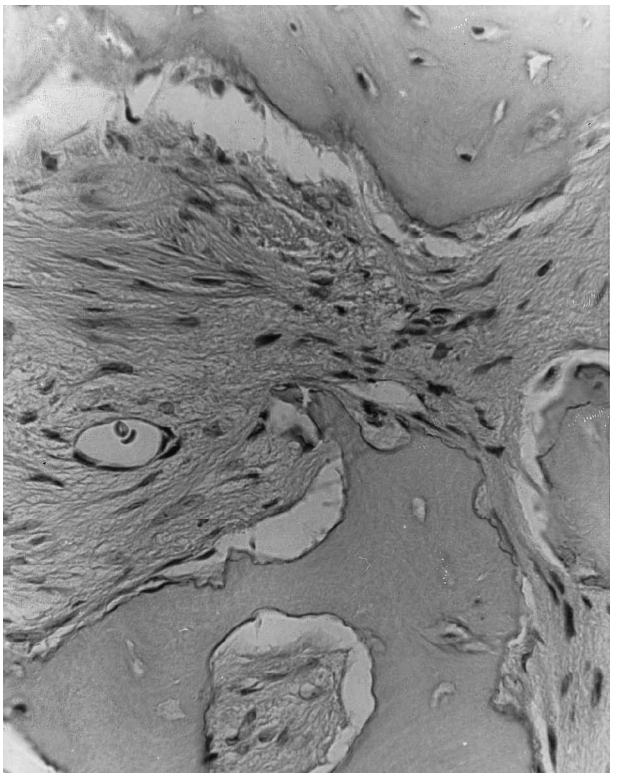

FIG. 6

Fibromatosis showing permeative growth pattern through bone (H & E; ×342).

permeative growth pattern infiltrating between the bony trabeculae (Figure 6) and extended through the full thickness of the bone involving dura and extending to the level of the cerebral cortex.

Immunostains for cytokeratins, S 100 protein, oestrogen and progesterone receptors were negative.

The appearances were those of a benign fibro-osseous lesion with a permeative growth pattern as seen in a fibromatosis involving bone (desmoplastic fibroma). Whilst lacking the ability to metastasize, these lesions tend to be locally infiltrative with a tendency to recurrence. The completeness of excision in this case could not be definitely confirmed.

Discussion

First described by Jaffe[2] in 1958, desmoplastic fibroma (DF) of the bone is a distinct entity that is the only benign fibrous bone tumour accepted by the World Health Organisation (WHO) with the following definition: 'A benign tumour characterized by the formation of abundant collagen fibres by the tumour cells. The tissue is poorly cellular and the nuclei are ovoid or elongated. The cellularity, pleomorphism and mitotic activity that are features of fibrosarcoma are lacking'.[5] DF is placed in the intermediate or borderline category between benign and malignant tumours because of its locally aggressive character.[8]

The tumour affects almost all the bones of the skeleton. However, as a solitary lesion DF occurs predominantly in the metaphysis of the long bones (58 per cent of cases), mandible (14 to 29 per cent) and pelvis.[9] Less common and rare sites include maxilla, sternum, thoracic spine and the parapharyngeal space.[10–13] DF is an uncommon neoplasm comprising less than 0.1 per cent of bone tumours.[4]

In the calvarial part of the skull, only about eight cases of DF have been described in the literature. We have reported here the ninth case and fourth such in the temporal bone. Table I details the reported cases of DF in the calvarial bones including their clinical presentations, modality of treatment and the follow-up. It is interesting to note that DF has a clear predilection for the first three decades of life and is more common in females.[8] However, our patient was in her early seventies.

Radiologically, the tumour is generally lytic with a honeycomb or trabeculated pattern and the differential diagnosis includes a unicameral bone cyst, fibrous dysplasia, chondromyxoid fibroma, nonossifying fibroma, giant cell tumour of the bone, fibrosarcoma of the bone, eosinophilic granuloma and metastatic lesions.[14]

Gross examination of the tumour reveals a firm but rubbery irregular lesion with a greyish white cut surface. Areas of bone and old haemorrhagic nodules can be identified. Tissue invasion is quite characteristic and hence a definite capsule is seldom found. DF is a non-metastasizing, but often locally aggressive neoplasm composed of fibroblasts and myofibroblasts in an abundantly collagenized stroma. It is regarded as the bone equivalent of soft tissue aggressive fibromatosis. It is capable of exhibiting locally aggressive, infiltrative growth and in our case extension had occurred through the dura to the cerebral cortex. Some authorities regard them as 'borderline' rather than purely benign even though metastases do not occur. The main consideration in the differential diagnosis is a low grade fibrosarcoma that has minimal nuclear atypia and abundant collagen production. Usually an occasional enlarged nucleus is present often possessing a nucleolus and a few mitoses are present. Sometimes the histological appearances of the two lesions overlap. Other lesions which have to be differentiated are a fibro-osseous lesion such as fibrous dysplasia and

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TABLE I
REVIEW OF THE LITERATURE ON DESMOPLASTIC FIBROMA OF THE CALVARIUM

Researchers	Age of patient (yrs)	Sex	Site of lesion	Presentation	Management	Follow-up
Gardini *et al.*[17]	7	F	Frontal bone	Tender nodule – 1 month	Complete excision, cranioplasty	NSR (time not mentioned)
Hufnagel *et al.*[18]	22	F	Parietal bone	Headache – 6 months	Complete excision, cranioplasty	NSR (time not mentioned)
Okuno *et al.*[6]	86	F	Temporal bone	Otorrhoea, deafness, swelling – 8 months	Excision	NSR (2 years)
Goldberg *et al.*[14]	42	F	Frontal bone	Headache – 3 months	Complete excision, calvarial graft	NSR (6 years)
Selfa-Moreno *et al.*[19]	28	F	Parietal bone	Headache – weeks	Complete excision	NSR (3 years)
Pensak *et al.*[7]	28	F	Temporal bone	Aural fullness, Otorrhoea – 1 year	Temporal craniotomy and petrosectomy	NSR (18 months)
Pensak *et al.*[7]	21	F	Temporal bone	Headache, aural fullness – 6 months	Temporal craniotomy and petrosectomy	NSR (4 years)
Celli *et al.*[9]	64	F	Frontal bone	Headache, swelling, – few months	Excision and cranioplasty	NSR (12 months)
Dutt *et al.* (present paper)	72	F	Temporal bone	Headache, swelling, aural fullness – 1 year	Petrosectomy, craniotomy and cranioplasty	NSR (6 months)

NSR = no sign of recurrence; F = female.

ossifying fibroma, a nonossifying fibroma, a myxoma, an eosinophilic granuloma and an odontogenic tumour. All of these lesions have different histological appearances which should enable distinction to be made.

Some authors have mentioned the need to demonstrate the presence or absence of oestrogen and progesterone receptors by immunohistochemistry in these tumours,[7] with a view to considering hormonal treatment with tamoxifen. Tamoxifen has been shown to produce regression of a recurrent desmoid tumour of the left upper arm and scapular region in a 30-year-old female patient by Thomas *et al.*[15] The tumour in our patient did not express hormonal receptors (ER or PR). There are no reports in the literature of a hormonal receptor positive DF of the bone.

As with the majority of authors, we believe the key to avoid recurrence is complete excision of the tumour. This is followed by a reconstructive procedure as deemed necessary. Freedman *et al.*[16] advocate a thorough curettage of the lesion. However, curettage alone has shown high recurrence rates of upto 42 per cent.[4] No recurrence has been reported with wide and complete excisions.[10] Awareness of this condition and its correct management are important to the otolaryngologist.

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