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Case Report

Desmoplastic fibroma of the skull

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Summary

Desmoplastic fibroma is a primary bone tumour which occurs exceedingly rarely in the cranial bones. We report a histopathologically confirmed example in a 21 year old man with a painless swelling over the sagittal suture. İn the light of our experience and reports of previous examples, we review the features of the condition and its management.

Keywords: Desmoplastic fibroma; skull; sagittal suture; marijuana.

Introduction

Desmoplastic fibroma (DF) is an unusual intra-osseous, locally aggressive tumour. The sites of predilection for DF are the long bones and mandible [13]. A review of previous reports yielded only sixteen examples in the cranial bones, of which four had involved the parietal bone. In this paper we report a further patient with a desmoplastic fibroma involving the sagittal suture and review the nature of the condition in the light of previous literature and our experience.

Review of literature

The results of our literature search via Medline using the keywords ''desmoplastic fibroma AND skull'' yielded twenty-nine articles between 1966 and 2006. Those reporting cranial DF examples were identified. Fourteen articles reporting DF in locations other than cranial such as mandible, maxilla, zygoma, parapharyngeal space were excluded. One article describing a patient with a low grade osteosarcoma of the skull and another entitled ''Limitations to mobilizing the intrapetrous carotid artery'' were also excluded. Finally thirteen articles describing DF in the calvarium were included in this review. The first cranial example of DF was reported by Gardini *et al.* in 1978 [6]. Up to date, sixteen patients with DF of the cranium have been published in 13 articles.

This review included 17 reports of cranial DF including ours and Table 1 summarises the clinicopathological findings [2, 4, 6, 8, 12, 16–19, 21–24]. Their ages ranged from 3 months to 86 years. Eleven of the seventeen were in adults and the other six in the paediatric age group. Twelve of the 17 cranial DFs occurred in the first three decades of life. The sex ratio was 1:2.4 (male: female 5:12).

The sites of occurrences were, 5 in the temporal bones, 5 frontal, 4 parietal, 1 frontal and temporal, one temporal with parietal bone involvement and ours involving the sagittal suture. There is no site predilection between paediatric and adult sufferers.

DFs usually present as slow growing mass lesions. The clinical presentation includes headache, hearing changes, ear drainage and head asymmetry. The duration of symptoms ranged from several weeks to 12 years. One of the examples is said to be congenital [16].

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Table 1. *Clinical data of the published reports of cranial desmoplastic fibroma*

Ref.	Tumor location	Age/sex	Presentation	Tumour size and gross appearance	Radiographic appearance	Treatment	Follow-up	Recurrence
Gardini *et al.* [6]	frontal	7/F	tender mass for 1 month	NA	NA	complete excision, cranioplasty	NA	NA
Hufnagel *et al.* [12]	parietal	22/F	6 month history of headaches	2 × 1 cm – firm mass	lytic lesion with irregular borders	surgical resection, cranioplasty	NA	NA
Ovul *et al.* [18]	parietal	3 month/M	3 month history of growing mass (present at birth)	5 × 4 × 0.8 cm – grey, white, solid, elastic	hypodense mass	complete excision	2 years	–
Okuno *et al.* [17]	temporal	86/F	ear drainage for 1 month	2–3 cm – smooth, firm	patchy mass	surgical resection	2 years	–
Goldberg *et al.* [8]	frontal	42/F	headaches for 3 months	NA – white, firm, rubbery	abnormal trabecular pattern with expansion	en bloc resection	6 years	–
Selfa-Moreno *et al.* [22]	parietal	28/F	headaches for several weeks	2 cm – grey-white, firm, rubbery	lytic lesion	complete excision	3 years	–
Pensak *et al.* [19]	temporal	21/F	over 6 months history of aural fullness	3.5 cm – NA	bony lesion	petrosectomy	4 years	–
	temporal	28/F	1 year history of aural fullness and intermittant suppuration	NA – grey-white, firm, rubbery, nonencapsulated	destructive mass	petrosectomy with tympanoplasty	18 months	–
Celli *et al.* [2]	parietal	64/F	cerebral MRI control (incidental)	2 × 3 cm – white, firm, fibrous, shinny	lytic lesion	surgical excision, cranioplasty	12 months	–
Dutt *et al.* [4]	temporal	72/F	painless head swelling for 10 years	2.3 cm and 2.5 × 1.5 × 1 cm – firm, rubbery	trabecular bony defect with intracranial (dural) extension	complete excision, cranioplasty	6 months	– (patient died from another cause)
Rabin *et al.* [21]	temporo-parietal	43/F	head asymmetry over 12 years	10 × 8 × 5 cm – nodular, white, firm, whorled cut surface	isointense mass, scalp and temporo-mandibular joint and minimal dural extension	complete excision, cranioplasty	2 years	–
Wolfe *et al.* [23]	frontal	3/M	mildly tender mass enlarged over 2 years	3 cm – tannish-grey mass	expansion of diploic space	complete resection	1 year	–
	temporal	7/F	nontender mass for 14 months	NA – rubbery, grey mass	lytic lesion associated with soft tissue component	excision and curettage	3 months	–
	frontal	22 month/M	nontender frontal mass	NA – soft mass	enlarged diploic space	complete excision	NA	NA
Yoon *et al.* [24]	fronto-temporal	1/F	10 month history of progressive swelling	8 × 7 × 1 cm – grey-white, firm	round lytic lesion with trabeculation	surgical excision, curettage	NA	NA
Lath *et al.* [16]	frontal	18/M	headaches, scalp swelling of 1 month duration	NA	frontal mass with intracranial extension and dural involvement	radical excision, duroplasty	1 year	–
Our case	sagittal suture (parietal)	21/M	nontender mass for 3 years of duration	3 cm – firm, white mass	lytic lesion	complete excision	15 months	–

NA Not available.

Skull radiographs revealed lytic lesions with irregular borders without sclerosis. CT scans showed bony defects and diploic space enlargement. Most of the cranial DFs are confined to the bone, although two showed soft tissue invasion [21, 23] and three others had intracranial extension [4, 16, 21]. None of them have been found to invade the brain parenchyma. The reported cranial DF lesions ranged in size from 2 to 10 cm.

The treatment of choice was complete surgical resection in nearly all patients. The follow-up duration ranged from 3 months to 6 years. All of the reports with follow-up data did not show any evidence of recurrence.

Illustrative clinical history

A 21 year old male was admitted to the neurosurgery clinic with a swelling over the parietal region. The lesion had been present for 3 years and progressively increased in size. There were no additional symptoms. He had a history of habitual marijuana use for 6 years. Physical examination revealed a firm, 2–3 cm swelling over the parietal region. The overlying scalp appeared intact. There was no discernable discrete margin with the underlying bone. Neurological examination was unremarkable. A computed tomography (CT) scan demonstrated a 2.5×3 cm osteolytic intra-osseous lesion located mainly in the sagittal suture with extension into the parietal bones (Fig. 1). There was no soft tissue and intracranial invasion. Routine laboratory test results were within normal limits.

The lesion was approached surgically through a scalp incision to provide full exposure of the lesion. Excision of the outer skull table and of the intraosseous tumor was performed.

On gross examination the whole specimen was $8 \times 4 \times 1$ cm in size. The outer skull table was in continuity with the firm irregular underlying lesion with a greyish–white cut surface. Light microscopic examina-

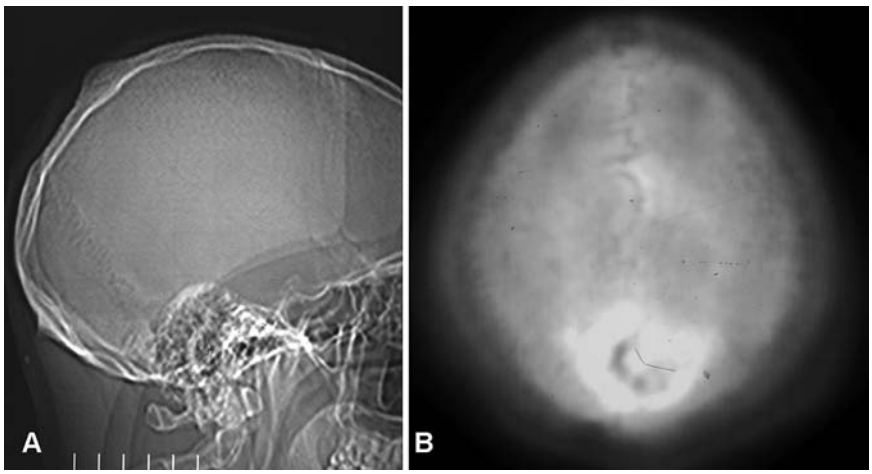

Fig. 1. (A) Cranial radiograph demonstrating a lytic lesion. (B) Axial computed tomography scan demonstrating an intradiploic lytic lesion

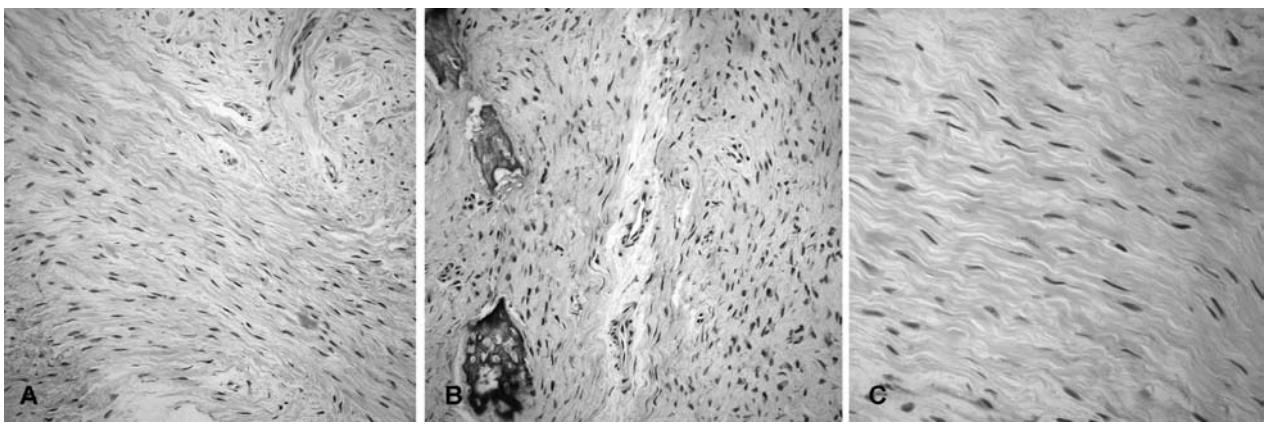

Fig. 2. (A) Lesion composed of spindle shaped cells with abundant collagen (haematoxylin and eosin, $\times 200$). (B) Entrapment of the destroyed bone is seen at the peripheral parts of the lesion (haematoxylin and eosin, $\times 200$). (C) Spindle shaped cells are in uniform appearence with elongated nuclei. No mitotic figures are present (haematoxylin and eosin, $\times 400$)

tion of the lesion demonstrated fascicles of monomorphic, spindle shaped cells. The lesion had a relatively low cellularity and tumour cells embedded in a highly collagenized background. The lesion was not encapsulated and was invading the bone. Fragments of destroyed bone entrapped by the tumour were observed at the periphery (Fig. 2). Immunostains for cytokeratins, epithelial membrane antigen (EMA), smooth muscle actin, desmin, S-100 protein, and CD34 were negative. The spindle cells were typically positive for vimentin. There was no immunoreactivity for oestrogen and progesterone receptors.

The post-operative course was uneventful. There was no local recurrence during 15 months of follow-up.

Discussion

Desmoplastic fibroma has been first described by Jaffe as a distinct entity in 1958 [14] and is accepted as a bone tumour by the World Health Organisation under the following definition: ''A rare, benign bone tumour composed of spindle cells with minimal cytological atypia and abundant collagen production''. DF is also known as desmoid tumour of bone and is an intra-osseous counterpart of soft tissue fibromatosis. This relatively uncommon tumour tends to occur in young adults and and may involve any bone [5]. Up to date more than 200 examples of DF have been reported in different sites [24]. Most of the DFs arise in the metaphysis of the long bones and they show predilection for the humerus and femur, while mandible, maxilla, sternum, vertebrae are less frequently affected [7]. Cranial DFs are exceedingly rare, as only 16 cases within the skull have been reported. To our knowledge no other example with cranial suture involvement has been reported. This is the first report of a DF involving the sagittal suture.

DF occurs with equal frequency in both sexes, but the cranial lesions show female predilection. Interestingly, all male patients except one with skull involvement reported to date have been in children [16]. We report the second adult male patient with cranial DF. As the number of reports of DF increase an equal gender distribution in the skull may be seen. Although DF can affect any age group, most patients tend to be in the first 3 decades of life [13]. Radiographically, DFs are well defined, lytic and expansile lesions with thinning of the cortex. CT and MRI may be useful in assessing the degree of bone destruction and soft tissue extension [9]. In our patient the plain radiographs demonstrated a lytic lesion and cranial CT confirmed that the lesion was without any soft tissue extension both of which are in line with previous reports.

Histologically, DF is characterised by a proliferation of elongated, spindle-shaped cells of uniform appearence with abundant collagen. The cells lack hyperchoromasia and atypia. Mitotic figures are usually infrequent. The edges of the DF were irregular with the tumour cells infiltrating into the bone. Therefore, the detection of tumour free margins is an important feature of radical excision [7, 13].

The histopathological differential diagnosis may include benign and malignant spindle cell tumours of the bone such as fibrous dysplasia, low grade intra-osseous osteosarcoma and non-ossifying fibroma. These lesions have sufficiently distinct histopathological features to distinguish from DF. Another important differential diagnosis is low-grade fibrosarcoma. The presence of cellular pleomorphism, hyperchromatism and mitotic figures are helpful in differentiating these two entities. Low grade fibrosarcoma can be distinguished from DF by the presence of mitoses and high cellularity in the former. Immunohistochemical studies are not useful in the discrimination of these lesions [13]. Intra-osseous meningioma sometimes causes a diagnostic problem, however the usual positivity of EMA allows discrimination of DF from meningioma [1, 20]. Malignant fibrous histiocytoma of the bone rarely causes difficulty in differential diagnosis of DF, because of its clear cut malignant histologic and radiologic features [5]. A detailed differential diagnosis of DF is summarised in Table 2. The correct diagnosis of DF is necessary to prevent inadequate or over treatment.

The pathogenesis of desmoid tumours and DFs are poorly understood. Development of desmoid tumours can be the result of genetic, endocrine and physical factors. Trauma is also a likely contributory factor [10]. Marijuana smoking has been implicated as an aetiologic factor in head and neck, lung and prostate cancer [3, 11]. Paternal marijuana use during gestation was associated with childhood leukaemia, rhabdomyosarcoma, and astrocytoma [11]. However, sufficent studies are not available to adequately evaluate the impact of marijuana on soft tissue and bone tumours. It is speculative that marijuana use may lead to development of DF by metabolic alterations. However, there is no report in the literature published in English indicating a cause-effect relationship between marijuana and fibroblastic tumors. On the other hand, the association between marijuana smoking and DF may be incidental. Additional studies are necessary to assess the association of fibroblastic tumours and marijuana use.

Table 2. *Differential diagnosis of desmoplastic fibroma [1, 5, 20]*

Tumour	Radiologic features	Histologic features	Immunohistochemical features
Fibrous dysplasia	geographic lesion with ground glass matrix; no soft tissue extension and periosteal reaction present	composed of fibrous spindle cells and osseous component with irregular trabecula of woven bone	vimentin (+) no specific marker
Benign fibrous histiocytoma	well defined radiolucent lesion, 2/3 of the lesions have sclerotic margins	spindle cell proliferation in storiform fashion with no atypia. The lesion is sharply demarcated from uninvolved bone	vimentin (+) no specific marker
Non-ossifying fibroma	sharply demarcated radiolucent lesions with sclerotic margins	cellular fibrous tissue with frequent collections of haemosiderin laden-macrophages and osteoclasts	vimentin (+) no specific marker
Cranial fasciitis	usually involves the outer skull table; lytic defect with sclerotic margins	regular spindle fibroblastic proliferation lacking nuclear atypia, the border is typically infiltrative and mitoses may be abundant	vimentin (+) smooth muscle actin (+) desmin and CD68 rarely (+)
Intraosseal meningioma	mostly hyperostotic or mixed osteoblastic-osteolytic; purely lytic lesions are rare	spindle cell proliferation with whorls and psammoma bodies	vimentin (+) EMA (+) S-100 and cytokeratin (+) in some cases
Low grade fibrosarcoma	destructive geographic lesion with ill-defined appearence and cortical destruction; periosteal reaction present	uniformly cellular spindle cell proliferation with readily identifable mitoses	vimentin (+) no specific marker
Low grade osteosarcoma	poorly marginated lesion with trabeculation and sclerosis; cortical destruction is common with or without soft tissue extension	moderately cellular spindle cell proliferation with some degree of atypia; osteoid production is generally evident	vimentin (+) no specific marker
Malignant fibrous histiocytoma	osteolytic lesion with ill-defined borders, cortical cortex is commonly involved and soft tissue extension present	mixed population of spindle, histiocytoid and pleomorphic cells; most of these tumours are high grade with abundant mitoses and necrosis	vimentin (+) smooth muscle actin and CD68 may be focal (+)

The microscopic features and the local aggressiveness of the desmoid-type fibromatosis (desmoid tumour) and DF are similar. DF is also associated with a high rate of local recurrence. The presence of oestrogen receptors was detected in approximately one third of desmoid tumours and endocrine therapy has become an alternative treatment [15]. Despite the morphological overlap with desmoid tumour cranial DFs did not show oestrogen receptor positivity [4, 19, 21]. Immunohistochemical studies showed negativity for oestrogen and progesterone in our patient. However, oestrogen receptor analysis should be done not to overlook the hormonal responsive DFs, since the published data on this issue mainly depends on single case reports [4, 19, 21].

En bloc resection of the tumour is the therapy of choice and most of the previously reported examples were treated with complete excision. It is essential to provide tumour-free margins, because of the infiltrative nature of DF [21]. We therefore removed the lesion with a margin of grossly normal appearing bone. Microscopic examination revealed tumour-free margins supporting the intraoperative evaluation. Sometimes it is difficult to estimate the surgical margins because of the abscense of a capsule. If the tumour is suspected to invade the margin, we recommend to extend the surgical margins. The recurrence rate reported is as high as 20–30% in DFs of the bone other than cranium. The median time to recurrence is approximately 2.7 years [13]. None of the reported skull lesions were found to have recurred. The follow-up period is sufficient only for 3 patients with cranial DF [8, 13, 22], the others having only short term clinical follow-up. Thus studies of series with longer follow-up are needed to show the exact behavior.

In summary, we describe a patient with cranial DF involving the sagittal suture associated with marijuana use in a young man in whom there are features different from other previous reports. Although our observation is based on a single patient and a small number of previous reports, the triad of findings that characterise this entitiy are: 1) Lytic lesion 2) microscopic appearence of benign spindle cell proliferation with entrapment of peripheral bone; 3) an indolent clinical course. A constellation of radiologic, microscopic and immunohistochemical features allows correct diagnosis and leads to appropriate management.

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