CASE REPORT

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Desmoplastic Fibroma of the Skull

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Desmoplastic fibroma is one of the uncommon osseous tumors that present in the skull. Although classified as benign tumor, desmoplastic fibroma exhibits local aggressiveness and has a high potential for recurrence. The most common sites include metaphysis of long bones and mandible. Only 15cases have been described in the skull. We report the 16th case of desmoplastic fibroma of the skull.

KEY WORDS : Desmoplastic fibroma · Intraosseous tumor · Complete excision.

Introduction

Desmoplastic fibroma is benign but locally aggressive intraosseous tumor that is frequently found in the long bones and mandible[3,14]. It was first described in the bone by Jaffe[10] in 1958 who differentiated it as an entity distinct from other types of intraosseous fibrous tumors. Histologically, the lesion is characterized by thick wavy collagen bundles and few fibroblasts with small oval nuclei and no atypia[5,15,16].

Nearly 200 cases of desmoplastic fibroma have been described in the literature. Desmoplastic fibromas of the skull are very rare. Only 15 cases have been reported and only two cases of these were in the infant. Here we present another case of desmoplastic fibroma of frontal bone in a 12-month-old girl.

Case Report

A 12-month-old girl visited our hospital with a three month history of progressive protuberance of the right frontal skull. The mass was about 2.5cm in diameter, nontender, fixed, and round over right frontal bone. The patient was neurologically and developmentally intact. Plain radiograph showed a round osteolytic lesion, 2cm in diameter with sclerotic margin. A computed tomography of the skull showed well defined osteolytic mass and diploic space enlarged with thinned cortices in right frontal bone (Fig. 1). MRI demonstrated an intradiploic soft-tissue lesion with prominent contrast enhancement. No dural enh-

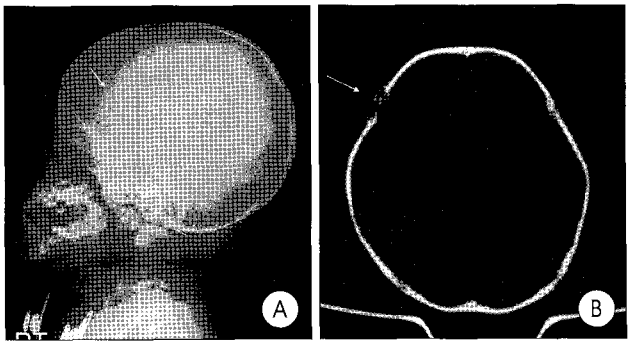

Fig. 1. Skull X-ray film of lateral view(A) shows well defined osteolytic mass with sclerotic rim, in right frontal bone just anterior to coronal suture. Axial computed tomography of the skull(B) demonstrates well defined osteolytic mass with bone remodeling and thinning in right frontal bone.

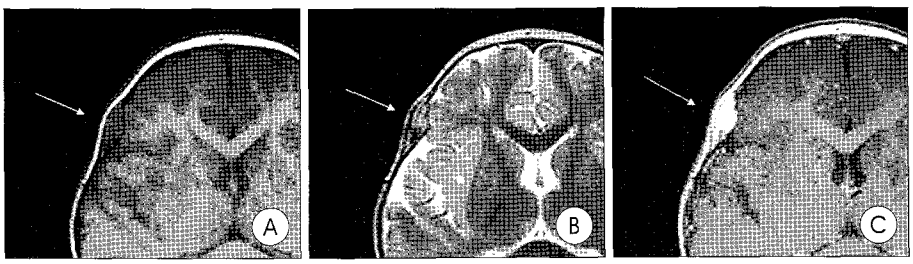

Fig. 2. In magnetic resonance imaging of brain, axial T1 weighted image (A), T2 weighted image (B), and contrast-enhanced T1 image (C) show well defined enhancing mass with sclerotic rim in right frontal bone.

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Fig. 3. A : The portion of the specimen consisted of three layers : the outer skull table, gray-white, firm, soft tissue, the inner skull table, and another portion of hard tissue with multiple bony islands. B : Section reveals irregular proliferating fusiform to spindle cells with bundles or whorling pattern in the abundant collagenous stroma(Masson's trichrome, ×100). C : There are occasional entrapped bony trabeculae and reactive bone formations(Masson's trichrome, ×200).

ancement or intraparenchymal extension was found (Fig. 2).

The patient underwent a craniectomy and complete mass resection. Grossly, the lesion appeared firm and yellow-white with erosion of outer table. There was no dural invasion. Pathological analysis revealed that desmoplastic fibroma was most probable. The color of the involved marrow had changed to a yellow-white, tumor-infiltrated, firm and rigid mass. Sections revealed irregular proliferating spindle cells with bundles or whorling pattern and abundant collagenous stroma. The tumor cells had oval to fusiform shaped nuclei. The evaluation of the exact cellular atypia or mitotic rate was difficult due to calcification. There were occasional entrapped bony trabeculae and reactive bone formation. Although there was limitation for histologic evaluation due to calcification, desmoplastic fibroma is most probable (Fig. 3).

Discussion

Desmoplastic fibroma(DF) is a rare tumor, accounting for 0.3% of benign bone tumors and 0.06% of all bone neoplasm[15]. DF can occur in all the bones of the skeleton. However, as a solitary lesion DF occurs predominantly in the metaphysis of the long bones (58%), mandible(14 to 29%) and pelvis. Firstly described by Jaffe[10] in 1958, DF of the bone is a distinct entity that is the only benign fibrous bone tumor accepted by the World Health Organization(WHO) with the following definition : 'A benign tumor characterized by the formation of abundant collagen fibers by the tumor cells. The tissue is poorly cellular and the nuclei are ovoid or elongated. The celluarity, pleomorphism and mitotic activity that are features of fibrosarcoma are lacking'[16]. Despite its slow growth and relatively benign histological appearance, DF falls within a intermediate or borderline category between benign and malignant bone tumor because of its locally aggressive nature[3,15].

Cranial DF is grossly a nodular, tan to white, firm but rubbery mass that may be attached to bone or the periosteum and may infiltrate the scalp muscles or underlying dura[3,15]. They expand the diploic space and commonly break through both the inner and outer cortex. Sequestrated bone fragments are found throughout the soft-tissue portion of the tumor. Because of the locally aggressive nature of this tumor, it is not encapsulated and usually fills the bone marrow spaces. Microscopically, DF is composed of slender or plump fibroblasts with wavy, ovoid nuclei sparsely dispersed within a collagenous and myxoid martrix[6]. There is no ossification and only rare mitotic figures. When examined with an electron microscope, DF have elongated spindle cells with minimally indented nuclei, rough endoplasmic reticulum with dilated cisterns, and bundles of intermediate filaments. Immunostaining is negative for S100, cytokeratin, estrogen, and progesterone[8,19].

Since the first case was reported by Gardini[5], fifteen cases of DF arising within the skull have been reported (Table 1). Most of the previously reported patients were treated with complete excision of the lesion, and no instances of recurrence were noted[6,12,14]. Extracranial DF have no sex predilection, but except two cases, all cranial DFs were found in female patients. Table 1 show that four lesions were found in frontal bone, five lesions in temporal bone, four lesions in parietal bone, one lesion in frontotemporal bone, and one lesion in temporoparietal bone and DF have no predilection site in cranial lesion. There was no recurrence.

Radiographically, DF in the long bones is characterized by local expansile osteolytic lesion in the metaphysis with thinning of the cortex, and a soap bubble appearance or diffuse trabeculation without periosteal reaction[10,11,20]. Infiltrative growth in the soft tissue was documented in 48% of patients[1]. DFs of the skull are typically solitary, osteolytic lesions of a honeycomb or trabecular pattern, with a mild or absent sclerotic reaction at the margins[6,10,15]. Typical CT findings are destruction and thinning of the bony cortex, and MRI can show the intermediate intensity on T1-weighted images and heterogenous intensity on T2-weighted images[6,18].

Lesions to be included in the differential diagnosis of des-

Table 1. Review of the literature on desmoplastic fibroma of the calvarium

researchers	Sex/age(yrs)	Site of lesion	Management	Follow-up
Gardini et al.[6]	F/7	Frontal bone	Complete resection and cranioplasty	NR(time not mentioned)
Hufnagel et al.[8]	F/22	Parietal bone	Complete resection and cranioplasty	NR(time not mentioned)
Ovul et al.[11]	M/3 months	Parietal bone		
Okuno et al.[10]	F/86	Temporal bone	Excision	NR(2 years)
Goldberg et al.[7]	F/42	Fronatal bone (supraorbital)	Complete resection and calvarial graft	NR(6 years)
Selfa-moreno et al.[15]	F/28	Parietal bone	Complete resection	NR(3 years)
Pensak et al.[12]	F/21	Temporal bone	Temporal craniotomy and petrosectomy	NR(4 years)
	F/28	Temporal bone	Temporal craniotomy and petrosectomy	NR(18 months)
Celli et al.[2]	F/64	Parietal bone	Excision and cranioplasty	NR(12 months)
Dutt et al.[3]	F/72	Temporal bone	Petrosectomy and cranioplasty	NR(6 months)
Rabin et al.[13]	F/43	Temporoparietal bone	Complete excision, petrosectmy and craniotplasty	Not mentioned
Yoon et al.[18]	F/1	Frontotemporal bone	Complete resection and cranioplasty	NR(12 months)
Wolfe et al.[17]	M/3	Frontal bone	Complete resection and cranioplasty	NR(12 months)
	F/7	Temporal bone	Excision and curettage	NR(3 months)
	F/22 months	Frontal bone	Complete resection and cranioplasty	Not mentioned
This study	F/1	Frontal bone	Complete resection	NR(6 months)

F = female, M = male

moplastic fibroma are hemangioma, eosinophilic granuloma, epidermoid, fibrous dysplasia, nonossifying fibroma, medullary fibrosarcoma, aneurysmal bone cyst, ameloblastoma, and metastatic tumors of the thyroid and kidney in children[8]. Hemangiomas have a honeycomb appearance on x-ray films with speculated trabeculations, similar to those of DF. Histologically, these two tumors can be easily differentiated. Eosinophilic granulomas are radiographically similar to DF as solitary, round, lytic lesions without sclerotic borders. In contrast to DF, eosinophilic granulomas are reddish purple, gritty, and soft due to their cellular histiocytic make up. Histologically, they are easily differentiated by their sheets of eosinophils and histiocytes. Epidermoid tumors are common skull lesions with a distinctly different radiographic appearance. They are usually found along the coronal suture and have sharp, sclerotic borders. Grossly, they are easily identifiable by their sebaceous and keratinized contents. Fibrous dysplasia has a classic soap-bubble appearance on CT scans that can be mimicked by DF. Their pathological hallmark is woven bone, a feature absent in DF. A nonossifying fibroma has classic, multinucleated giant and foam cells with osteoclastic activity at the edge of the lesion. These features are absent in DF. Medullary fibrosarcoma share the radiographic characteristics of DF, but as a malignancy they tend to be more aggressive. Histologically, fibrosarcomas are more cellular with a herringbone pattern. The cells are larger and plumper mitotic figures. An ameloblastoma is an odontogenic tumor arising from ectoderm. It is commonly found in the maxilla and mandible but rarely in the skull. Metastatic tumors of the skull have similar radiographic appearances but usually multiple and are rarely found in the pediatric age group. In our patient, the radiological differential diagnosis based on the preoperative films was fibrous dysplasia, desmoplastic fibroma, eosinophilic granuloma.

Complete excision of the tumor is the treatment of choice. Freedman et al.[4] advocated a thorough curettage to the lesion. However, curettage alone has shown high recurrence rates of up to 42%. No recurrence has been reported with wide and complete excision. In our case, the patient is being followed up and any evidence of recurrence was not found.

Conclusion

Desmoplastic fibroma of the skull is very rare, but it should be carefully differentiated from other cranial benign tumor due to its local aggressive nature. Complete resection with margins with possible cranioplasty and radiographic follow-up should be considered if DF is suspected by clinical and radiological findings.

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