CASE REPORT

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Desmoplastic Fibroma of the Skull

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Desmoplastic fibromas are rare intraosseous bone tumors. They are benign but locally aggressive and frequently found in the long bones and mandible. We report radiographic and histopathologic finding of a case desmoplastic fibroma involving right temporal skull bone. A 53-year-old woman presented at our hospital complaining of continuous right side headaches for a year. Simple skull X-ray film showed 3×2.5cm lytic lesion with mild sclerotic margin on right temporal area. A large craniectomy 1cm lateral to margin was fashioned. The resected mass showed encapsulated mass colored white gray. Histologic diagnosis was compatible with that of a the desmoplastic fibroma. There was no evidence of recurrence during the 15months of follow-up period.

KEY WORDS : Desmoplastic fibroma · Temporal bone · Skull.

Introduction

Jaffe described desmoplastic fibroma was distinct clinicopathologic entity in 1958[15]. Desmoplastic fibromas are rare intraosseous bone tumors. They are benign but locally aggressive and frequently found in the long bones and mandible[3,9]. About eleven cases of desmoplastic fibromas involving the skull have been reported[4-6,9,13,16-21,23]. We report radiographic and histopathologic finding of a case desmoplastic fibroma involving right temporal skull bone.

Case Report

This 53-year-old woman presented at our hospital complaining of continuous right side headache for a year. Physical examination revealed painful soft mass on right temporal area. Simple skull X-ray film showed 3×2.5cm lytic lesion with mild sclerotic margin on right temporal area(Fig. 1). Brain CT scan aslo demonstrtaed lytic lesion on right temporal area measuring 3×2.5cm(Fig. 2). Brain magnetic resonance imaging(MRI) showed high signal intensity lesion on T2 weighted image(Fig. 3).

At the time of surgery scalp was found intact. The tumor eroded the outer table of skull. A large craniectomy 1cm lateral to margin was fashioned. The exposured dura showed clear without any involvement. A cranioplasty was performed. The resected mass showed encapsulated mass colored white gray.

Microscopically, the tumors were composed large amounts of hyalinized collagen. rarely the fibroblastic cells were arranged in hyalinized collagen matrix. The cells had a monotonous appearance and lacked pleomorphism. No mitotic figures was founded in this case (Fig. 4).

After Surgical *en bloc* resection with wide margins, Headache disapperaed almost completely. There was no evidence of recurrence during the 15 months of follow up period.

Discussion

Desmoplastic fibroma is a rare tumor, accounting for 0.3% of benign bone tumors and 0.06% of all bone neoplasms[9]. Desmoplastic fibroma is relatively benign histological appearance, but it is thought as a category between benign and malignant bone tumors because of

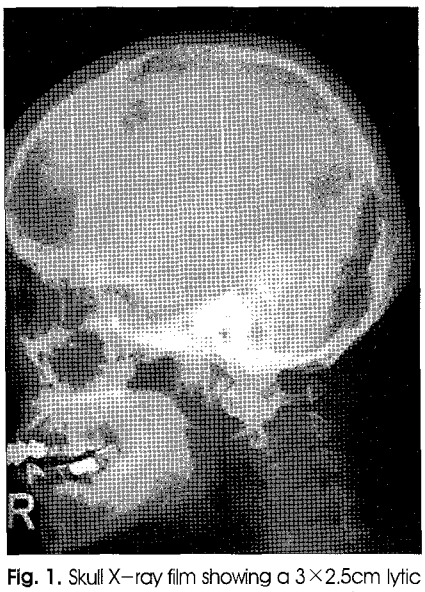

Fig. 1. Skull X-ray film showing a 3×2.5cm lytic lesion with mild sclerotic margin on the right temporal area.

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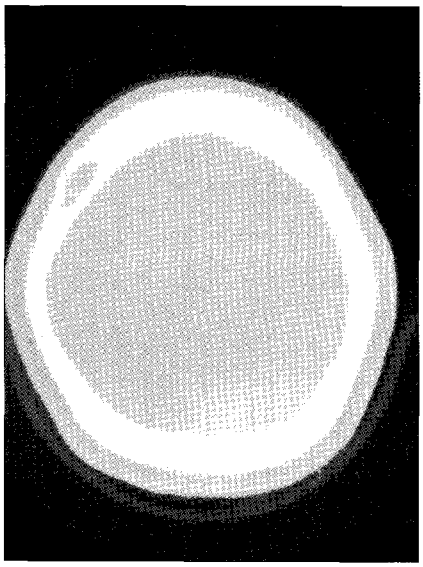

Fig. 2. Brain computed tomography scan aslo demonstrating a lytic lesion on the right temporal area measuring 3×2.5cm.

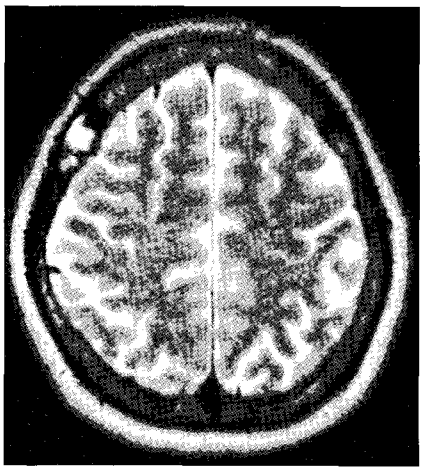

Fig. 3. Brain magnetic resonance imaging showing a high signal intensity lesion on T2 weighted image.

its locally aggressive nature[3,4].

Desmoplastic fibromas most commonly occur in the metaphyses of the long bones, the mandible, and the pelvis(approximately 70% of occurrences)[5], whereas the maxilla, calvaria, sternum, and vertebrae are less frequently affected. It occurs with equal frequency in both sexes and has a predilection for patients in the first 3 decades of life[14].

There are several clinical characteristics in desmoplastic fibromas of skull. Desmoplastic fibromas occurred with female predominantly (only two male in 12 cases including this case)[4-6,9,13,16-21,23]. This suggest a possible hormonal dependence of desmoplastic fibroma mentioned by Triantafyllou et al.[25]. The patient's age ranged from 7 years to 86 with a mean age 36. But over the half of reported cases were less than 30 years like desmoplastic fibroma occurring other parts. There are no predilection site within skull as shown Table 1.

Initial presenting symptom is headache in most of the patients. Most of the previously reported patients were treated with complete excision of the lesion, and no instances of recurrence were noted[4-6,9,13,16-21,23]. They are known as locally invasive and tendency to recur if they are not resected widely. The desmoplastic fibroma usually does not invade the dura if it was small but an advanced case was adherent to the underlying dura, requiring an extensive dural resection[21].

For patients with desmoplastic fibromas arising from the maxilla or mandible with extraosseous extensions, complete excision, including a margin of uninvolved soft tissue, is recommended[8,12,24]. Inwards et al.[14] reported no recurrence in seven patients with skeletal desmoplastic fibroma who underwent *en bloc* resection, whereas 9 of 11 patients treated with lesional curettage or marginal excision did develop a recurrence. Desmoplastic fibromas of skull could be resected more widely than desmoplastic fibromas of other area so there were no instances of recurrence[4-6,9,13,16-21,23].

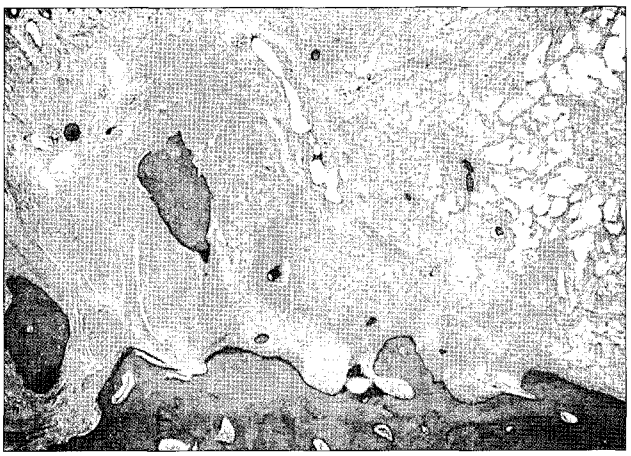

Fig. 4. Histologic finding of the desmoplastic fibroma. The tumors are composed a large amounts of hyalinized collagen(H&E×100).

Table 1. Summary of reported cases of desmoplastic fibroma of skull

Series(reference No)	Sex/Age(year)	Location
Garnini et al 1978	F/7	Frontal bone
Hufnagel, 1987	F/22	Parietal bone
Ovul, 1988	M/25	Parietal bone
Okuno 1990	F/86	Temporal bone
Goldberg 1995	F/42	Frontal bone
Selfa-Moreno 1995	F/28	Parietal bone
Pensak 1997	F/21	Temporal bone
Pensak 1997	F/28	Temporal bone
Celli 1997	F/64	Parietal bone
Kim 2001	M/22	Parietal bone
Rabin 2003	F/43	Temporoparietal bone
This study	F/53	Temporal bone

(F:female, M: male)

Radiotherapy may be an acceptable alternative therapy when *en bloc* resection is impossible. Sanfilippo et al.[22] reported a case of pelvic desmoplastic fibromas treated with radiotherapy alone. They noted no radiographic evidence of disease progression at 30 months of follow up[22]. Chemotherapy for desmoplastic fibroma has not been described. Desmoid, the soft tissue equivalent of desmoplastic fibroma[5,9,13], may respond to endocrine therapy[2,26]. Wilcken reviewed 35 cases of desmoid tumor that 51% of the cases treated with endocrine therapy were responsded partially[26].

Desmoplastic fibroma has several radiographic characteristics,

that is, local expansion, well-defined margins, and a soap bubble appearance[14].

Desmoplastic fibromas of the cranium are typically solitary, lytic lesions with a mild or absent sclerotic reaction at the margins[1,7]. A CT scan define the extent of local bone destruction. MRI can confirm displacement of local soft tissues in the absence of obvious local infiltration[11]. The dense connective tissue and hypocellularity of desmoplastic fibromas result in an intermediate signal intensity on T1-weighted images and a heterogeneous intensity on T2-weighted images[3]. Some author mentioned signal intensity on T2-weighted images may be of prognostic value in terms of interval growth of desmoid lesions[17]. But, the specific MRI characteristics are incompletely described because this lesion is relatively rare.

Desmoplastic fibroma has a gross appearance of a nodular, white mass that may be firmly attached to bone or periosteum[3,10,14]. Microscopically, desmoplastic fibroma is composed of slender or plump fibroblasts with ovoid nuclei sparsely dispersed within a collagenous and myxoid matrix[10]. A lack of mitotic figures and nuclear atypia may distinguish Desmoplastic fibroma from more malignant bone neoplasms[10]. The principal difference between desmoplastic fibroma and fibromatosis tumor is the site of origin. Desmoplastic fibromas arise within the bone, whereas desmoid tumors originate in musculoaponeurotic structures[5,13]. Desmoplastic fibroma act in an expansile fashion, eventually breaking through the cortex of the bone and extending into the surrounding soft tissues[3,10].

Conclusion

Although desmoplastic fibroma is uncommon, it should be considered in the differential diagnosis of a osteolytic skull tumor. Surgical *en bloc* resection with wide margins is the choice of treatment. Desmoplastic fibroma may be locally aggressive, total resection should be attempted.

• Acknowledgement
Desmoplastic fibromas are rare intraosseous bone tumors. We report radiographic and histopathologic finding of a case desmoplastic fibroma involving right temporal skull bone. There was no evidence of recurrence during the 15months of follow-up period.

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