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CASE REPORT

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Cranial fasciitis with massive intracranial extension

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Abstract The case of a 10-month-old boy with cranial fasciitis is described. The patient had a rapidly growing subcutaneous mass in the left frontotemporal region. Computed tomography and magnetic resonance imaging clearly demonstrated a mass in the left temporoparietal bone extending both intra- and extracranially. The tumor seemed to originate from the calvarium, being located between the periosteum and the dura mater. Total resection of the tumor was performed, and the tumor was histologically identified as cranial fasciitis. A brief review of the literature is included that emphasizes the need for further investigation of this benign lesion that is frequently confused with a malignant neoplasm.

Key words Cranial fasciitis · Nodular fasciitis · Childhood · Intracranial involvement · Magnetic resonance imaging

Introduction

Cranial fasciitis of childhood, originally described by Lauer and Enzinger in 1980 [11], is a proliferative, fibroblastic, tumor-like lesion that resembles the nodular (or pseudosarcomatous) fasciitis described by Konwaler et al. in 1955 [9]. Both are benign, self-limited lesions, and they are histologically similar. Since the original description of nine cases of cranial fasciitis, ten additional cases have been reported [1, 2, 4, 5, 7, 10, 12–15]. Because none of these cases was associated with massive intracranial extension, all except two [7, 12] were published in non-neurosurgical journals. In this paper, we report on a child with cranial fasciitis with massive intracranial involvement, which was thought to have arisen from the cranial bone.

Case report

A 10-month-old boy was admitted to our department in November 1991 because of a rapidly enlarging tumor under the scalp in his left frontotemporal region. Three months prior to admission, his mother had noticed a slight nodule on his left temple. There was no episode of head trauma. Two months before admission to our department, plain skull X-rays and computed tomographic (CT) scan were performed by another physician. Skull X-rays showed an osteolytic lesion in the parietal bone near the pterion and immediately posterior to the coronal suture (Fig. 1A). CT scan demonstrated a subcutaneous soft tissue density mass 1.7×0.7 cm in diameter (Fig. 2A). There was slight destruction of the outer table but no intracranial involvement was seen.

Upon admission, neurological examination revealed nothing abnormal. There was a subcutaneous mass in the left temple, which was 5 cm in diameter, firm, not tender, and not mobile upon palpation. Skull X-rays demonstrated that the osteolytic lesion had increased in size, with a peripheral rim of osteosclerosis (Fig. 1B).

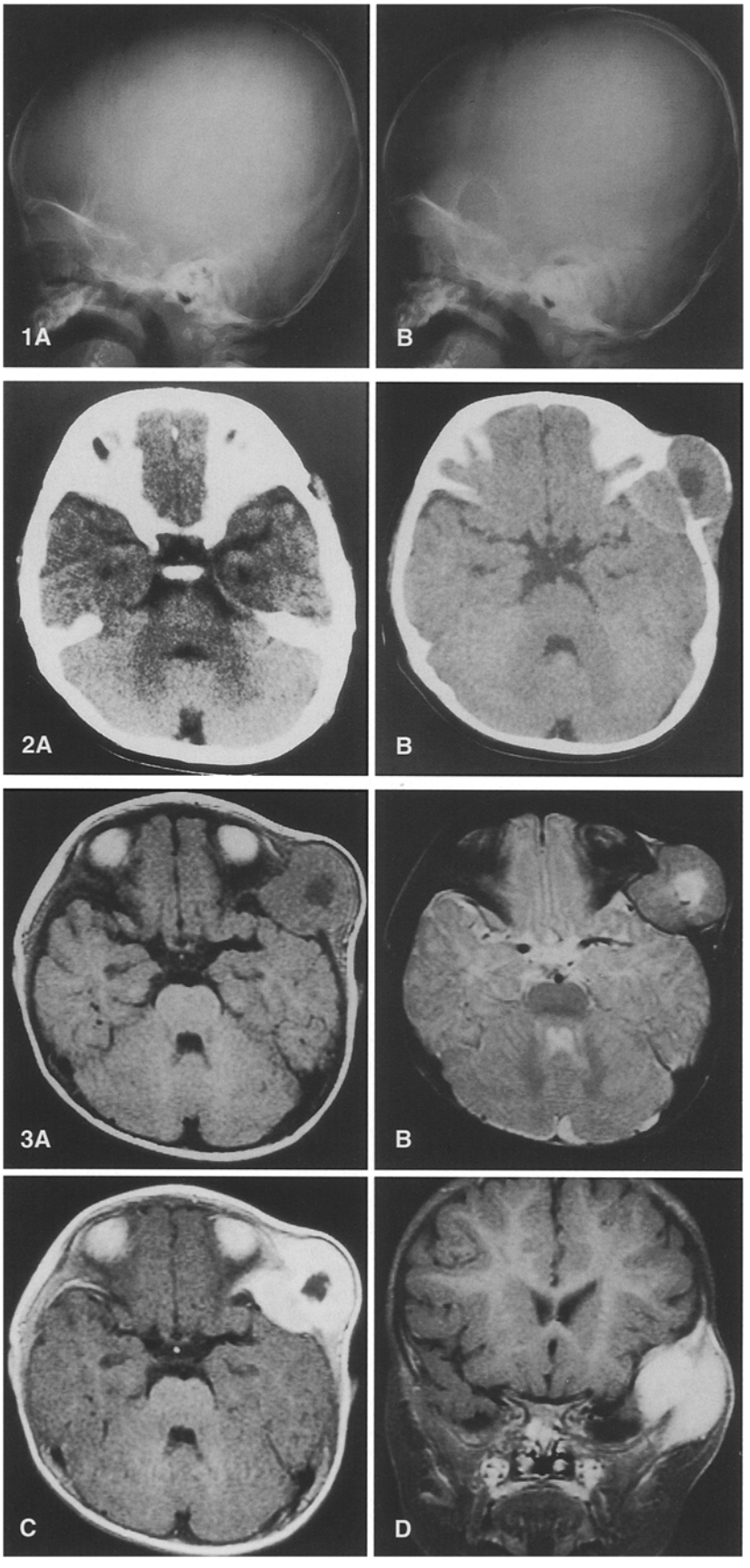

Fig. 1 **A** A plain radiogram shows osteolytic lesion of the left parietal bone near the pterion. **B** Two months later, an enlargement of the osteolytic lesion with a sclerotic rim is observed

Fig. 2 **A** Computed tomographic (CT) scan shows a subcutaneous soft tissue mass with slight expansion of the skull. **B** Two months later, an enlargement of the tumor with massive cranial involvement is demonstrated. There is a cystic lesion in the center of the tumor. The rim of the tumor is covered with a rim of bony density

Fig. 3 Axial views on magnetic resonance imaging (MRI) demonstrate an isointense mass both on T1- (**A**) and T2-weighted images (**B**) extending both intra- and extracranially. After intravenous administration of Gd-DTPA, the tumor is markedly enhanced except for at the center (**C** axial view; **D** coronal view)

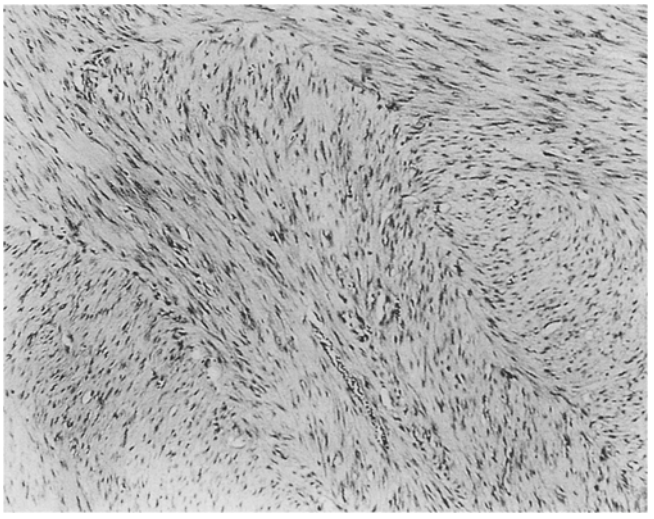

Fig. 4 Irregular whorls of spindle cells with scattered chronic inflammatory cells. Scattered mitoses are present. (Hematoxylin and eosin, ×75)

CT scan revealed a subcutaneous soft tissue density mass with massive intracranial involvement (Fig. 2B). A linear bone remnant skirted the outer rim of the tumor. The tumor caused scalloping of the edge of the cranium. These findings suggest that this mass arose from the cranial bone. After administration of contrast medium, the tumor was markedly enhanced except for a small cyst-like lesion in the center.

Magnetic resonance imaging (MRI) also clearly revealed a mass in the left temporoparietal bone extending both intra- and extracranially. This mass was isodense with gray matter on both T1- and T2-weighted images (WI) (Fig. 3A, B). The lesion showed marked enhancement with intravenous administration of gadolinium diethylene triamine pentaacetic acid (Gd-DTPA) (Fig. 3C, D). The central part of the tumor showed hypointense on T1WI and hyperintense on T2WI without contrast enhancement (Fig. 3).

The rapid growth of the lesion and the destruction of the bone with massive intracranial involvement were interpreted as signs of malignancy, and total removal of the tumor was performed. The extracranial part of the tumor was entirely covered with the intact periosteum. The outer table of the bone surrounding the tumor was thick and elevated. The extracranial tumor was whitish and elastically firm with little vascularity. Remnants of bone were identified on the surface of the tumor. The inner table of the bone surrounding the tumor was also slightly compressed inward. There was relatively tight adhesion of the tumor to the underlying dura, which was separated under a surgical microscope. In the center of the tumor a well-demarcated cyst was found containing yellowish-brown fluid. The tumor was totally removed including the thick surrounding bone, which did not show neoplastic involvement.

Histological examination revealed a mass composed of fibroblast-like spindle cells arranged in a fascicular or storiform pattern (Fig. 4). There were scattered microhemorrhages, inflammatory lymphoid cells, and foamy cells with occasional giant cells in the tumor. Cellular pleomorphism, necrosis, and mitotic activity were not found. The center of the mass was a cyst. The histological diagnosis was cranial fasciitis.

The postoperative course was uneventful. The boy was discharged without receiving radiation or chemotherapy. He is doing well and today, 8 months after the operation, is still without any sign of recurrence.

Discussion

Cranial fasciitis of childhood has been described as a nodular fasciitis-like lesion that arises in the scalp of young children [5, 11]. The exact nature of this entity is yet unknown. The term "fasciitis" does not mean inflammation of the cranial fascia, but is attributed on the basis of the histological similarity to nodular fasciitis.

The usual manifestation of this entity is a rapidly growing mass in the scalp of infants. Only 20 cases of cranial fasciitis of childhood have been reported including ours [1, 2, 4, 7, 10–15]. The age of these children ranged up to 7 years, and 12 cases occurred in children less than 2 years of age. There is a 2:1 male predominance.

No systematic description of the neuroradiological findings in cranial fasciitis exists for plain X-ray. In 11 of the reported patients the radiograph of the skull showed a lytic defect with a sclerotic rim [1, 2, 11–15]. This radiologic phenomenon was caused by the indentation/scalloping of the cranial bone by the fibroblastic proliferation [11, 13, 15]. This rather typical radiological feature is thought to occur in children due to the incomplete ossification of the membranous bones of the skull [15]. A rapidly growing skull mass with bone defect may occur in eosinophilic granuloma in children, which does not show osteosclerosis at the marginal bone of the defect. Other rare skull lesions manifesting with bone defects may occur with giant cell tumors, aneurysmal bone cysts, ossifying fibromas, but have not been shown to occur in children. Osteogenic sarcoma can be ruled out, since this tumor is exceedingly rare in the cranial bone.

CT scan findings were described in four cases [2, 7, 12, 14]. Most show a subcutaneous mass of soft tissue density, with involvement of the underlying cranium usually limited to the outer table of the skull. However, in some cases the mass extends to involve the inner table and is attached to the dura [2]. In our case, the first CT scan showed similar findings to those of the reported cases, but the second CT scan revealed the tumor to extend both intra- and extracranially.

To our knowledge, no report has been published of MRI findings in cranial fasciitis. The mass in our case demonstrated an isointensity on both T1- and T2WI. The mass exhibited enhancement following intravenous administration of Gd-DTPA. The cystic cavity in the center of the tumor showed a hypointense signal on T1WI and hyperintensity on T2WI. There are no MRI findings specific to cranial fasciitis. Although MRI clearly demonstrated the intracranial involvement, it showed the lesion as indistinguishable from a malignant tumor.

The case reported here is unusual for two reasons. First, this is the only documented example of cranial fasciitis with massive intracranial extension. Most of the reported cases show roentgenographic or operative evidence of involvement of the underlying cranium usually

limited to erosion of the outer table of the skull, but occasionally with extension through the inner table and attachment to the dura [11, 13, 15]. Seven out of 19 reported cases involved dural attachment, but none of them had massive intracranial extension.

Secondly, judging from the CT scan and operative findings, this cranial fasciitis arose from the cranial bone. In the original description, cranial fasciitis arose from the periosteum or the deep fascial layer [11]. Recent studies report that the microscopic features of the cranial fasciitis closely resemble those seen in infantile myofibromatosis (solitary congenital fibromatosis) of bone, making it likely that they all represent the same entity [8]. Infantile myofibromatosis is also a rare entity most commonly seen during infancy and childhood. Solitary myofibroblastic nodular lesions arise in bone as well as muscle, or subcutaneous tissue, and they follow a benign course [3, 6, 8]. Our case may provide evidence that supports this concept.

This lesion is essentially self-limited and benign in nature. A total excision of the lesion with curettage of the underlying bone, if involved, is curative [11, 14, 15]. No additional methods of treatment, including radiation and chemotherapy, are necessary. Therefore, when a scalp mass is seen in a child, even with bone destruction and/or intracranial involvement, cranial fasciitis of childhood should be considered.

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