Diagnosing cranial fasciitis based on distinguishing radiological features

Report of 4 cases

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Primary skull lesions, albeit rare in the pediatric population, have been well described and classified. These lesions are usually benign and commonly present as a painless mass. The most common lesions are epidermoid, dermoid, and Langerhans cell histiocytosis. Cranial fasciitis, encountered less frequently, is usually not considered in this differential diagnosis. Given such few cases reported, it is commonly misdiagnosed preoperatively.

The authors retrospectively reviewed data obtained in 4 patients with cranial fasciitis in whom the diagnosis was based on histopathological findings. In 2 patients the onset of the lesion was spontaneous. One patient had a lesion 4 months following a vacuum extraction and subsequent cephalohematoma formation. One patient developed a lesion following a previous craniectomy. Presentation, imaging studies, and histopathological findings were all reviewed and analyzed. All patients presented with a firm nontender mass. Radiological features included a lytic bone lesion with a mildly sclerotic margin, T1 isodensity, T2 heterogeneous hyperdensity, and heterogeneous enhancement. The enhancing portion was not bright on T2-weighted MR images, likely representing the fibrous component; the nonenhancing portion was bright on T2-weighted images, likely representing the myxoid matrix. Histopathological examination revealed proliferating fibroblasts in a myxoid matrix.

Cranial fasciitis is a benign, painless but rapidly growing lesion of the skull mainly limited to the pediatric population. It is histologically similar to nodular fasciitis, a fibroblastic proliferation of varying size. These lesions are often related to trauma but can also be insidious or can develop at a prior craniectomy site. The appropriate clinical picture and distinguishing radiographic features may help to differentiate cranial fasciitis from other lesions of the skull allowing for earlier intervention. (DOI: 10.3171/PED.2008.2.11.370)

KEY WORDS • cranial fasciitis • magnetic resonance imaging • nodular fasciitis • primary skull lesion
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Case Reports

Case 1

This 4-month-old boy presented with an increasing firm mass in the right frontoparietal region, which had been present since 6 weeks of age. The family denied that the child appeared to be in any discomfort. The patient was born at 39 weeks' gestation in a spontaneous vaginal delivery requiring no assistance; however, he was noted to have hypoxia at birth requiring a 6-day stay in the neonatal intensive care unit.

Physical examination revealed a nontender, well-circumscribed lesion over the right frontoparietal region measuring about 3 × 2 cm. The lesion was firm and there was no evidence of trauma around the mass.

An unenhanced CT scan of the brain revealed an expansive, partially lytic lesion arising from the interdiploic space with a mildly sclerotic margin involving both inner and outer tables.

Magnetic resonance imaging demonstrated a slightly heterogeneous mass lesion within the right side of the scalp, nearly isointense to brain on the T1-weighted images and somewhat hyperintense to gray matter on the T2-weighted images. We also noted evidence of relatively intense enhancement superficially, although centrally there was a nonenhancing region, which was noted to be bright on T2-weighted sequences. There was also no evidence of abnormal diffusion restriction.

At 4 months of age, the patient underwent gross-total resection because the lesion was enlarging. Intraoperatively, the mass was found to be firm and fleshy. The mass was dissected from the skull, exposing the dura mater, which was noted to be intact.

Histological examination showed a well-circumscribed tumor with dural attachment and prominent nodular pattern. The tumor was cytologically composed of bland spindle cells arranged in short, irregular fascicles separated by bundles of hyalinized collagen. The central areas had fewer cellular areas with a myxoid appearance. There were also scattered multinucleated giant cells and focal areas of ossification. The histological features were most consistent with those of cranial (nodular) fasciitis.

Case 2

This 11-year-old girl presented with a painless lesion in the right parietal scalp area that appeared to grow rapidly over a period of 4 months. Her medical history was significant for a right frontal endoscopic biopsy of a pineal primitive neuroectodermal tumor and placement of an extraventricular drain 3 years previously. Subsequent to her biopsy the patient underwent cranial radio- and chemotherapy.

Physical examination revealed a nontender, well-circumscribed lesion over the right parietal region measuring about 4 × 3 cm. The lesion had developed posterior to the cranietomy site but was within the irradiated field. The remainder of her examination was nonfocal.

A noncontrast CT scan of the brain revealed a moth-eaten appearance to the bone overlying a well-defined extraaxial mass lesion (Fig. 1). There was also a nodular-appearing rim with central low density.

Magnetic resonance imaging demonstrated a vividly enhancing extraaxial mass underlying the right parietal bone (Fig. 2A). There was decreased enhancement centrally and a component that appeared to extend through the diploic space. Again, the nonenhancing region was bright on T2-weighted MR imaging (Fig. 2B). There was also no evidence of abnormal diffusion restriction (Fig. 2C).

Given the patient’s history of malignancy and radiation therapy, we chose to undertake surgery. Gross-total resection was performed, during which the mass was found to extrude through the center of the exposed bone (Fig. 3). The lesion was very soft and was easily dissected off the dura using blunt dissection (Fig. 3A). We noted no dural defect (Fig. 3B). The hole in the center of the bone was repaired with titanium mesh.

Histopathological examination revealed a glistening, firm, pink lesion (Fig. 4). Histologically, there was a well-circumscribed mass in which we observed a distinct myxoid background (Fig. 4A). There were also scattered inflammatory cells consisting mainly of small lymphocyte-like cells (Fig. 4B). Focally, multinucleated cells were seen and had the appearance of osteoclast-like giant cells (Fig. 4C). The histological features were most consistent with cranial (nodular) fasciitis.

Case 3

This 4-month-old boy presented with a midline occipital scalp mass, the size of which, the family noticed, had increased over 3 weeks. His medical history was significant for a difficult vaginal delivery at 34 weeks' gestation when vacuum extraction led to the formation of a cephalohematoma. The hematoma resolved by 3 months of age, but a new nodule medial to the swelling was noted.

At that time a contrast-enhanced CT scan of the brain revealed a very mildly enhancing region of nodular soft-tissue swelling in the midline and left paramedian occipital scalp region measuring 2 × 3 cm.

Fig. 1. Case 2. Axial CT scan showing a lytic lesion involving both inner and outer tables.
On physical examination the head and sutural anatomy appeared normal. There was a 2-cm, firm, somewhat fixed midline occipital scalp mass that was nontender. The patient was seen again in 4 weeks at which time he had a new 2.5-cm right parietal scalp mass that was firm and fixed. The midline occipital mass was still present and stable in size.

Magnetic resonance imaging was conducted 1 month after this visit, and studies demonstrated T1-weighted hypointensity of both lesions (Figs. 5A and 6 left). The occipital lesion exhibited intense enhancement with a central noneenhancing area that had T2 signal hyperintensity and no diffusion restriction (Fig. 5B and C). The only radiological difference with the parietal lesion was the lack of a central noneenhancing area (Fig. 6 right).

One month later the patient returned to the clinic, and the parietal mass had resolved, but the occipital mass was unchanged. It was then decided to resect the lesion.

Grossly the lesion had a rim of glistening, firm, pink-white tissue. The central portion was softer with a boggy texture. Microscopic examination revealed a thick cluster of spindle cells. Frozen and permanent sections showed dense fibrocollagenous tissue with adherent adjacent skull bone. The central portion of the fibrous tissue showed a relatively well-circumscribed lesion with a distinctively basophilic and myxoid stroma. The histological features were consistent with a benign spindle-cell proliferation with the characteristic features of cranial fasciitis.

**Case 4**

This 4-month-old boy presented with an enlarging, nontender, mobile, firm occipital mass first noticed when the patient was 2 months of age. The parents denied any complications with the pregnancy or delivery.

Head CT scanning revealed an extracranial, nonlytic, nonsclerotic lesion. Magnetic resonance imaging demonstrated a lenticular, heterogeneous, intensely enhancing lesion centered in the subperiosteal region of the left occipital bone with a central noneenhancing area that was noted to be bright on T2-weighted images.

Gross-total resection was performed when the patient was 5 months of age. Histological examination revealed a well-circumscribed, subcutaneous lesion composed of nodular aggregates of reactive fibroblastic proliferation intermixed with inflammatory cells, consistent with cranial fasciitis.

**Discussion**

Cranial fasciitis represents a rare type of skull lesion, and since its first description in 1980 very few cases have been reported. Cranial fasciitis differs from nodular fasciitis in its age of onset (almost exclusively in young children) and its location in the scalp, but histologically the lesions are the same. They both consist of predominantly spindle-shaped and stellate fibroblasts with some mitotic figures. They also have similar immunohistochemical features with reactivity for vimentin and smooth muscle actin. The difference, however, between benign cranial fasciitis and a more malignant tumor with these same characteristics, such as fibrosarcoma or fibrous histiocytoma, is that with the former there is less cellularity and variation in the cellular arrangement within the myxoid stroma.
Radiological diagnosis of cranial fasciitis

Cranial fasciitis occurs chiefly in children < 6 years of age and involves the soft tissues of the scalp and underlying skull. It is more common in males, and although there are no definite predisposing factors, a history of local trauma is cited in many reports. There have also been reports of these lesions being related to birth trauma and prior craniotomy. The lesions typically grow rapidly and may attain an impressively large size. In most cases, they arise from the deep fascia of the scalp and erode the outer table of the cranium but infrequently penetrate the inner table, infiltrating the dura and sometimes the leptomeninges. The parietotemporal regions are the most commonly affected. Patients usually present with a history of a rapidly growing scalp mass that is firm and nontender. Symptoms if present are usually secondary to mass effect. This particular type of skull lesion is considered a benign, probably reactive process likely arising from the galea aponeurotica or the epicranial aponeurosis, and can infiltrate the incompletely formed cranial bone. Excision is the definitive therapy, and the lesions rarely recur thereafter.

All of the lesions in the aforementioned cases presented as nontender scalp masses, and all exhibited the same histological features. In Cases 1 and 4 there was no history of birth trauma, but the patient in Case 1 had perinatal hypoxia. The patient in Case 3, however, had undergone vacuum extraction, which correlated to the site of the lesion. Despite the fact that the patient in Case 2 had previously undergone a craniectomy, the lesion was actually distal to this site but was in the area of the irradiated field. There has been one other case report of this tumor occurring after radiotherapy.

Radiologically, these lesions have distinguishing features, specifically on MR imaging. In all 4 cases there was vivid enhancement with a central nonenhancing area that was hyperintense on T2-weighted images. We also observed hypointensity and no diffusion restriction on T1-weighted images. The only T2-weighted hyperintensity was the nonenhancing area of the lesion. In Case 3 we observed 2 lesions: 1 was histologically determined to be cranial fasciitis, continued to grow slowly, and did...
not resolve, although it shared the aforementioned MR imaging characteristics, whereas the second lesion, which did resolve, had slightly different MR imaging features. Both lesions appeared identical on CT. However, on MR imaging the lesion that resolved did not exhibit any area of central nonenhancement and therefore no T2-weighted central hyperintensity. Despite the fact that the diagnosis of cranial fasciitis is primarily a histological one, the unique MR imaging features have proven to be important in the differentiation between this benign condition and other lytic skull lesions.

**Conclusions**

Our cases clearly demonstrate that cranial fasciitis can occur in the absence of trauma as well as in the presence of prior radiotherapy. Each case demonstrated the previously reported presentation and histological pattern.\(^3\)-\(^6\),\(^13\),\(^14\) We, however, have demonstrated that all of these lesions have specific MR imaging characteristics, which is important in diagnosing and distinguishing cranial fasciitis from other skull lesions that may resolve and not grow, allowing for earlier intervention.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**


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