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Case Report

Extremely rare occurrence of multiple cranial fasciitis: A case report

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Cranial fasciitis (CF) is a rare, rapidly growing, benign fibroproliferative lesion of the skull in the pediatric population. It is characterized by benign mesenchymal proliferation of spindle cells arranged as short, intersecting loose fascicles within a fibromyxoid stroma, and mostly appears as a single mass. A surgical excision with clear surgical margins is definitively curative for CF. Up to date only two cases with multiple CF have been reported in the literature. In this report, we describe a 1-year-old girl with multiple locations of CF, as the first case to be reported in the Turkish population. The radiological and morphological findings of our case were comparable with the observations of the two previous reports in the literature. Histopathological examination remains to be the gold-standard for differential diagnosis of CF, as the treatment of this lesion differs from other malignancies of the skull in the pediatric population.

Key words: clinicopathological correlation, cranial fasciitis, fibroblastic proliferation, multiple lesions, pediatric population.

INTRODUCTION

Cranial fasciitis (CF) is a rare, rapidly growing, non-neoplastic, myofibroblastic lesion of the scalp and underlying bones, which occurs between the ages of 47 and 61 years, but mainly occurs in the pediatric population at 3 weeks to 6 years of age.[1] This benign lesion is histologically similar to the spindle cell mesenchymal tumors such as meningioma, schwannoma, or sarcomatosis and histiocytosis due to its rapid, nodular growth pattern. CF is characterized by loose fascicles of stellate cells in a fibromyxoid background, findings similar to those described in the closely related variant nodular fasciitis.[2] Although the concept of malignant nodular fasciitis has recently been reported histomorphologically,[3] CF is a self-limiting myofibroblastic neoplasm with a low risk of metastasis or malignant recurrence. Generally, complete resection is considered curative for CF.[4] Up to date, there are only two reports of multiple CF cases, presenting as distinct small scalp masses in the pediatric population.[5,6] In this paper, we present the findings of clinical, radiological and histopathological examinations in a 1-year-old girl with two distinct CF lesions, located in the temporal and frontal regions. This is the first case of multiple CF in a pediatric patient in Turkish population.

CASE PRESENTATION

Clinical summary

A 1-year-old girl presented with a painless mass on the pterional region of the left temporal bone, which had been first noticed by the family when she was four months old and had been kept under observation only. The patient had been referred to our pediatric neurosurgery unit with a history of accelerated growth of the mass in the last two months. There was no history of seizures, evident trauma, infection or previous systemic disease. She was a healthy young girl with an obstetric history of a nontraumatic vaginal delivery after an uncomplicated pregnancy. Head examination revealed a soft, nontender and immobile left temporal lesion of about 2 cm. The overlying skin and scalp hair seemed normal and were not adherent to the mass.

For further clinical examination, a magnetic resonance imaging (MRI) of the cranium was performed. MRI examination revealed gadolinium-contrast-enhancing mass lesions in the left temporal bone and in the right frontal bone. Both lesions were broad-based masses, measuring 20 × 18 × 12 mm in the temporal site and 12 × 10 × 5 mm in the frontal site. The lesions were seemed to have destroyed the inner and middle tabulae of the skull

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regions, and showed possible invasion of the dura mater (Fig. 1). The differential diagnosis based on the imaging findings included dermoid/epidermoid cyst, eosinophilic granuloma, Langerhans cell histiocytosis, leptomeningeal lipoma cyst, meningioma, neurofibroma, lymphoma and osteosarcoma.

Surgical resection

The temporal site lesion, which was larger in dimensions and had a history of rapid growth, was resected surgically. During the surgery, the periosteum overlaying the lesion was observed as normal and not to be adherent to the lesion. After the lesion borders were explored, an ultrasonic bone cutter was used to perform a small craniectomy around the tumor, maintaining a grossly clean margin of 5 mm. The gray-brown colored tumor mass was easily dissected from the neighboring dura mater and excised totally. No dural adherence or invasion was observed by the surgeons. Regarding the lesion located in the frontal bone, which did not cause any neurological or cosmetic problems, the treatment was decided to be planned according to the results of the histopathological examination.

The patient had an uneventful post-operative course and was discharged home after two days. A repeat radiological examination was planned four weeks after surgery.

Pathological findings

Morphologically, the tissue sections stained with hematoxylin and eosin showed no evidence of dermoid/epidermoid cyst or Langerhans cell histiocytosis. The extravasated red blood cells, proliferation of spindle and stellate fibroblastic cells forming short fascicles with variable and loose myxoid stromal background were observed in histological sections. A number of cellular lesions were detected to erosion the cortical bone (Fig. 2). Despite this erosion, there was no evidence of malignancy and no necrosis/mitosis was detected. Considering the location of the mass, the meningioma and schwannoma were also included in the differential diagnosis for this proliferation of spindle cells (Fig. 3).

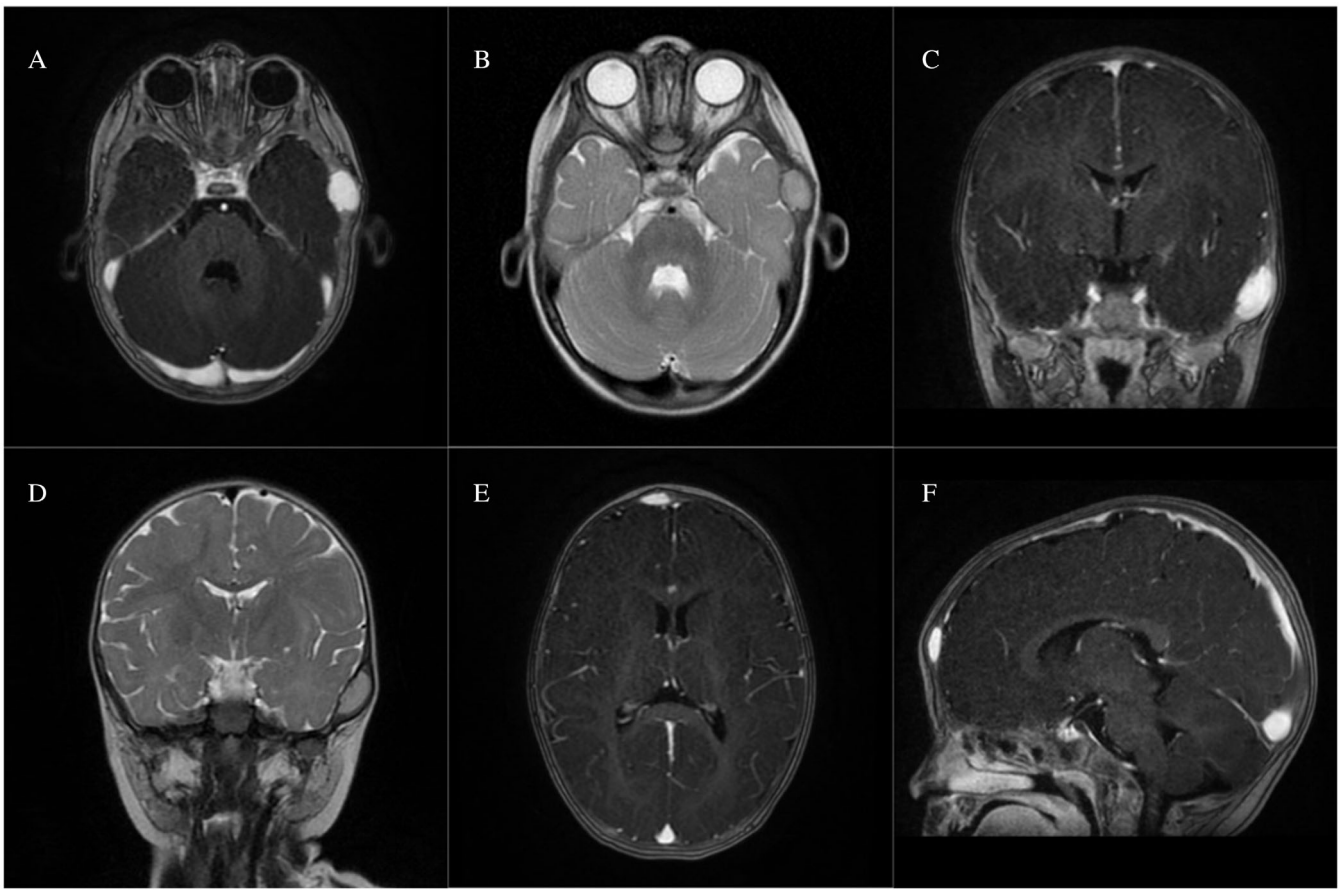


Fig 1 Preoperative magnetic resonance images (MRIs) of the presented case. (A) Axial contrast-enhanced T1-weighted, (B) axial T2-weighted, (C) coronal contrast-enhanced T1-weighted, and (D) coronal T2-weighted MRIs of the left temporal mass. (E) Axial and (F) coronal contrast-enhanced T1-weighted MRIs of the mass in the frontal bone, right adjacent to the sagittal sinus.

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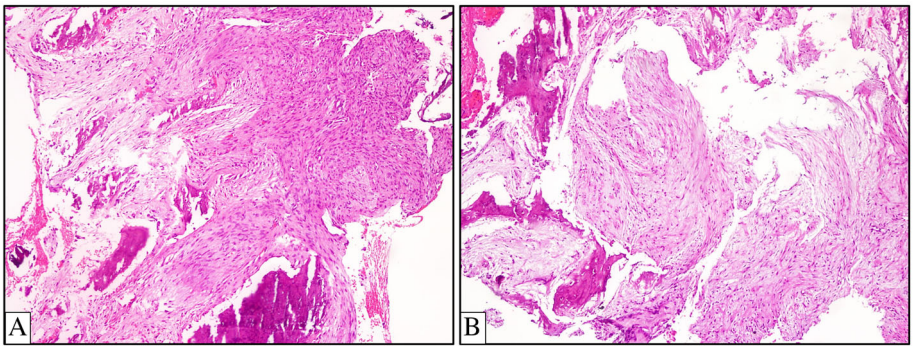


Fig 2 (A, B) Spindle cell mesenchymal lesion among destroyed cortical bone fragments (hematoxylin and eosin, × 100).

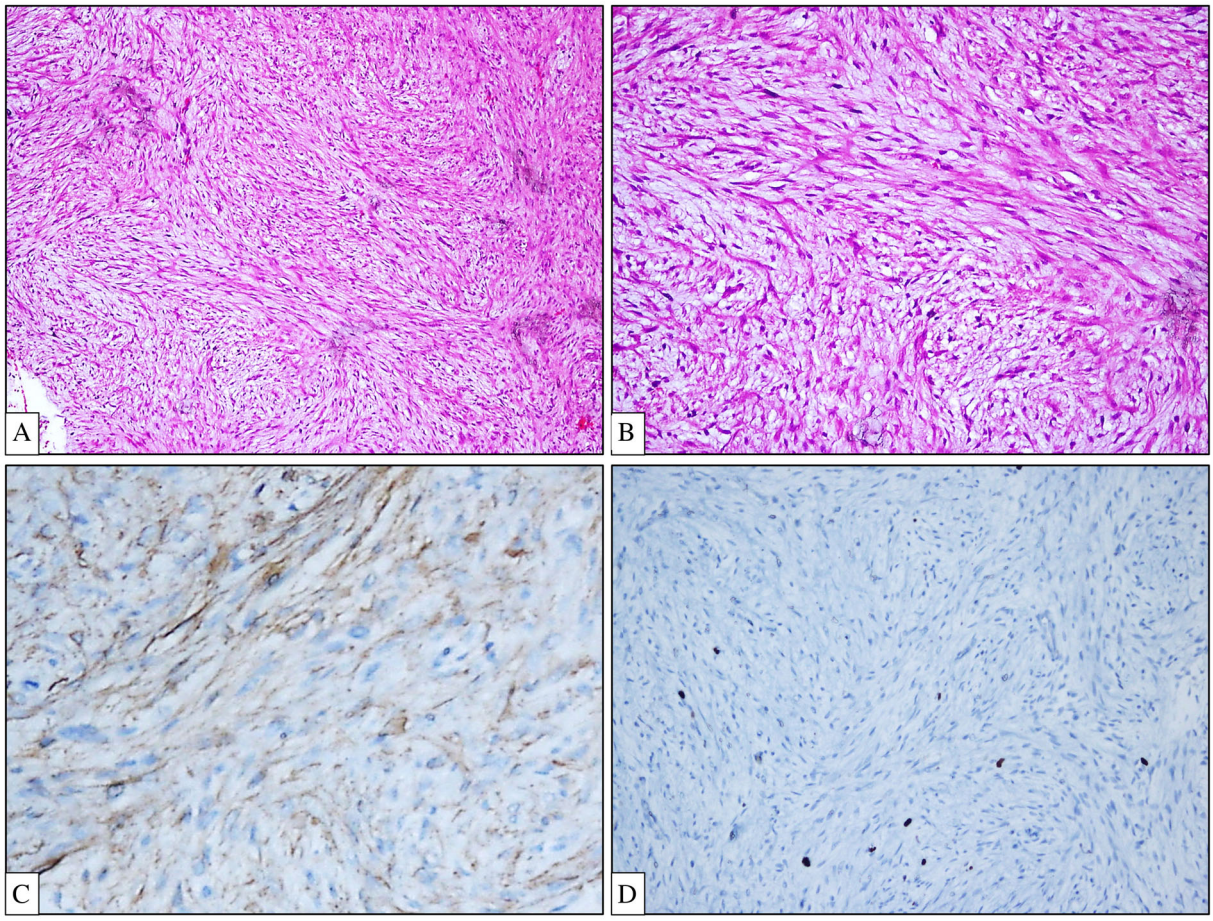


Fig 3 (A) Proliferation of fibroblasts and myofibroblasts with a storiform pattern, devoid of cytological atypia, arranged in short, intersecting fascicles in a fibromyxoid stroma (hematoxylin and eosin, × 100). (B) Higher magnification area of (A) (hematoxylin and eosin, × 200). (C) Positive staining pattern with beta-catenin localized in the cytoplasm in focal areas (beta-catenin, × 400). (D) Benign mesenchymal proliferation of spindle cells with low Ki67 labeling index (Ki67, × 100).

Neoplastic cells immunohistochemically showed a positive reaction with anti-vimentin antibody (clone V9; Dako, Glostrup, Denmark; 1:200), anti-beta catenin antibody (clone clone 15B8; Dako, Glostrup, Denmark; 1:200) and anti-smooth muscle actin (SMA) antibody (clone 1A4; Dako, Glostrup, Denmark; 1:200). Anti-progesterone antibody (clone PgR 636; Dako, Glostrup, Denmark; 1:200), anti-epithelial membrane antigen (EMA) antibody (clone GP1,4; Dako, Glostrup, Denmark; 1:200), anti-CD1A antibody (clone EP3622; Dako, Glostrup, Denmark; 1:400), anti-keratin antibody (clone AE1/AE3; Dako, Glostrup, Denmark; 1:50), anti-desmin antibody (clone D33; Dako, Glostrup, Denmark; 1:200), anti-CD34 antibody (clone QBend10; Dako, Glostrup, Denmark; 1:200) and S-100

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(Polyclonal; Dako, Glostrup, Denmark; 1:900) proteins were negative. This tumor had a low Ki-67 labeling index (2%; clone SP6; Dako, Glostrup, Denmark; 1:75; Fig. 3). Considering these findings, the case was diagnosed with CF.

DISCUSSION

Cranial fasciitis is a painless benign growing mass of the cranium, typically emerged as a single mass detected in the first years of life in pediatric patients. The median age of patients is 21 months.[7] Multiple masses in patients with CF are extremely rare in the literature. Up to date, only two reports of multiple CF cases have been published in the literature, presenting as distinct small scalp masses in the pediatric population.[5,6] The 12-month-old case with two distinct lesions of CF presented in this paper is the third case to be reported in the literature.

Cranial fasciitis is a non-neoplastic fibroblastic proliferation that can originate from the cranial structures including the fibromembranous membranes covering the sutures and fontanelles to the galea aponeurotica. The most commonly affected cranial region is the temporal bone,[8] as in our case. The lesions are typically rapidly growing, but tend to regress, and the recurrence after resection is very rare.[8] The mass typically arises as a single lesion from the deep scalp fascia, approximately 2.5 cm in median size.[7] A surgical excision with clear surgical margins is definitively therapeutic for CF.[6]

Histological examination of CF tissues has commonly contained loosely arranged, elongated, spindle and stellate cells in a myxoid background, all of which were best detected by hematoxylin and eosin staining.[9] When the benign spindle cell mesenchymal proliferation is observed, infantile myofibromatosis (IM), schwannoma and meningioma should be included in the differential diagnosis, especially for temporally located masses. In IM, they are centrally hyperintense on T2-weighted images and isointense on T1-weighted images. Target sign may be seen due to central necrosis. CFs appear as a heterogeneously increasing mass that is hypointense on T1-weighted images and has high signal intensity on T2-weighted images. In addition, no diffusion restrictions are observed on T1-weighted images.[10] Histopathologically, IMs have a biphasic pattern of polygonal/round cells with hyperchromatic nuclei. They consist of dark and light-stained areas scattered around blood vessels similar to hemangiopericytoma. In CF, short fascicles consisting of intersecting myofibroblasts with fine chromatin and small nucleoli are seen in loose collagenous and myxoid stroma. It is usually accompanied by extravasated erythrocytes and interspersed inflammatory cells. There are narrow, long and wavy cells in schwannoma that are immunohistochemically stained positive with S100. It has conical ends interspersed with collagen fibers. Tumor cells have poorly defined cytoplasm, and dense chromatin. Meningioma is spindle cell neoplasm with few or no meningothelial whorls or nests, positive for the meningothelial markers SSTR2A and EMA; usually they have thick collagen bundles. The CF lesions are immunohistochemically diffusely positive for HHF35, AML but negative for S-100 protein, EMA and progesterone. Beta catenin stain may show a cytoplasmic pattern in these lesions. Sometimes myofibroblasts may also be found in CF, so the SMA will also give a positive reaction.[1] Our case showed similar histopathological findings with the literature.

Electron microscopy may indicate the disclosing elongated spindle cells containing granular cytoplasmic material and myofibroblast cells with characteristically dilated rough endoplasmic reticulum, microfilament bundles and pinocytotic vesicles.[11] However, the electron microscopical examination was not available for our case.

A case series of CF demonstrated USP6 translocations by break-apart florescent in situ hybridization.[12] USP6 Gene Rearrangement by FISH Analysis was not available for our case. There are also recent case reports that suggest that CF may also harbor USP6 fusions.[2] In another study involving 6 cases, aberrant nuclear beta-catenin positivity was found in 3 patients. It has been reported that this indicates the dysregulation of the Wnt/beta-catenin pathway in a subset of CF, and this subset is pathobiologically related to desmoid fibromatoses rather than nodular fasciitis.[13] In the light of these findings, we applied beta-catenin staining to our case. We did not detect nuclear reactivity with this immunohistochemical staining. Further molecular genetic studies are needed to understand the genetic background of these rare multiple CF lesions.

In this report, a 1-year-old girl with multiple locations of CF was described, as the third case in the literature. The radiological and morphological findings of our case were comparable with the observations of the two previous reports. Histopathological examination remains to be the gold-standard for differential diagnosis of CF, as the treatment of this lesion differs from other malignancies of the skull in the pediatric population.

AUTHOR CONTRIBUTIONS

BYB, VE drafted the manuscript; BYB, HES, ATE, VE collected all medical reports of the patients, BYB, VE revised the manuscript critically. All authors read and approved the final manuscript.

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DISCLOSURE

No potential conflict of interest was reported by the authors.

DATA AVAILABILITY STATEMENT

All data generated or analyzed during this study are included in this published article.

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