Cranial Nodular Fasciitis: Case Report and Systematic Review of the Literature

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INTRODUCTION

Cranial fasciitis (CF) is a rare, benign, fibroproliferative condition predominantly affecting the pediatric population. Initially classified in 1980, CF typically presents as a rapidly expanding, asymptomatic subcutaneous mass, most often located in the parietotemporal region of the calvarium. Although the exact etiology remains unclear, proposed factors include trauma, radiation therapy, hereditary influences, and idiopathic origins. Diagnosis relies on imaging, histological evaluation, and complete excision, with Gross Total Resection (GTR) yielding excellent prognoses and minimal recurrence rates.

This study presents a case of CF in a 20-month-old girl with a 4-month history of a progressively growing, painful temporal mass, accompanied by a systematic review of clinical features, management strategies, and outcomes in CF.

CASE REPORT

A 20-month-old girl presented with a 4-month history of a progressively enlarging, painful and firm right temporal mass, with no other symptoms or history of trauma. Imaging studies revealed a lytic lesion in the right temporal bone extending into adjacent soft tissues and minimally into the epidural space.

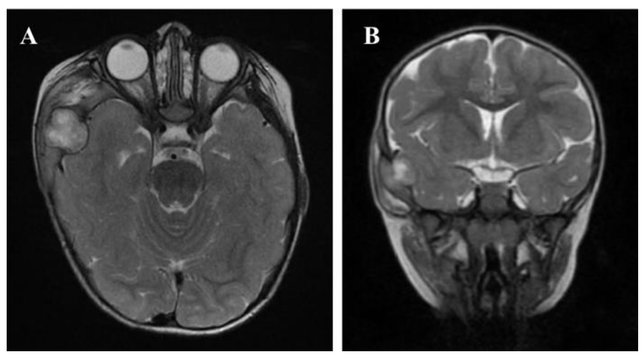

Figure 1. Preoperative imaging. (A-B) MRI showing an expansive lesion in the right temporal diploic region, measuring 20x24x27 mm

The patient underwent an open right temporal craniectomy with Gross Total Resection (GTR). Histopathology confirmed cranial subtype nodular fasciitis, with immunohistochemical positivity for α-SMA and calponin and benign features. Postoperatively, a contained cerebrospinal fluid (CSF) fistula was managed conservatively. At follow-up, she remained asymptomatic, with no evidence of recurrence.

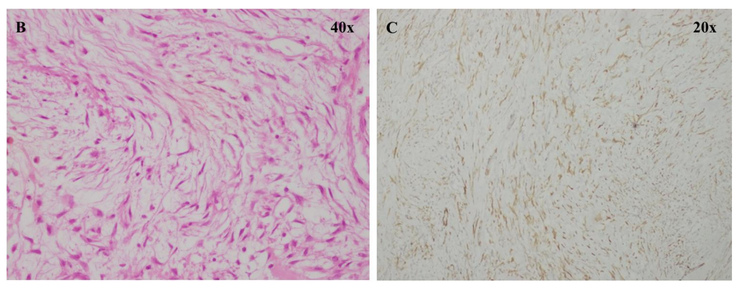

Figure 2. Histopathology (B) Hematoxylin and eosin stain showing spindle-shaped fibroblasts. (C) Immunohistochemically positive for alpha-smooth muscle actin (α-SMA).

RESULTS

A total of 149 cases of CF were analyzed, with a predominance of pediatric patients (88.8%) and an average age of 8.14 years. The cohort included 44% females. Trauma, prior radiotherapy, and previous surgeries were reported in 11.48%, 4.05%, and 7.43% of cases, respectively. Bone invasion was observed in 73.64% of cases, and dural involvement was present in 21.6%.

Table 1. Tumor locations of cranial fasciitis among all articles.

Anatomical location	Frecuency
Temporal	41(27.70%)
Frontal	23 (15.54)
Occipital	21 (14.18%)
Parietal	17 (11.48%)
Frontal - parietal	7 (4.72%)
Frontal- temporal	5 (3.37%)
Temporal - parietal	6 (4.05%)
Occipital – parietal	2 (1.35%)
Orbit	8 (5.40%)
Mandible	6 (4.05%)
Maxillar	4 (2.79%)
Ethmoid	1 (0.67%)
Others (nasal cavity, scalp, nasolabial)	6 (4.05%)

Surgical intervention was the primary treatment approach, with gross total resection (GTR) performed in 87.73% of cases, subtotal resection (STR) in 6.75%, and near-total resection (NTR) in 2.02%. Bone-specific procedures included craniectomy (16.51%), curettage (15.59%), and lesion resections without craniotomy (44.95%).

Biopsies were performed in some cases, with incisional biopsy in 4.05% and excisional biopsy in 1.35%. Other treatments included intralesional corticosteroid injections.

Table 2. Clinical presentation of cranial fasciitis among all articles.

Features	Frequency
Growth characteristics	
Rapidly enlarging mass	70 (42,29%)
Slowly growing mass	18 (12,16%)
Unspecified mass	24 (16.21%)
Pain and sensation	
Painless and/or nontender mass	26 (17.56%)
Painful and/or tender mass	3 (2.02%)
Asymptomatic (incidental diagnosis)	3 (2.02%)
Symptoms and associated issues	
Swelling of the area	2 (1.35%)
Blurred vision, diplopia, proptosis, hypoglobus	9 (6.08%)
Neurologic deficit	2 (1.35%)
Headache	3(2.02%)
Otalgia, otorrhea, otitis media	3(2.02%)
Nasal blockage and discharge	1 (0.67%)
Trismus and pain in the temporomabdibular joint	1 (0.67%)
Vomiting	1 (0.67%)
Sleep disturbances	1 (0.67%)

Recurrence occurred in 3.37% of cases. All recurrences were successfully managed with additional surgical interventions, and no further relapses were reported.

CONCLUSION

Cranial fasciitis (CF) is a rare benign condition, primarily affecting children, characterized by painless, rapidly growing masses, often in the temporal region. Its causes include trauma, radiation, and genetic factors. Diagnosis combines imaging and histology.

Treatment involves gross total resection (GTR), with a low recurrence rate. Bone curettage is advised for osseous involvement. Prognosis is generally favorable, but follow-up is necessary for recurrence detection. Early diagnosis and surgical treatment are crucial, with further research needed for a deeper understanding.

REFERENCES

1. Hussein MR. Cranial fasciitis of childhood: a case report and review of literature. J Cutan Pathol. 2008;35(2):212–4.
2. Dai R, Pien IJ, Brown DA, Marshall A, Fuchs HE, Marcus JR. Multiple recurrent fibromatosis with cranial fasciitis characteristics in a pediatric patient. J Craniofac Surg. 2017;28(7):1821–3.
3. Lauer DH, Enzinger F, Xiang Y, He S, Zhou Z, Gan Q. Cranial fasciitis in children: clinicoradiology features and management. BMC Pediatr. 1980;45(2).
4. Xiang Y, He S, Zhou Z, Gan Q, Jin K. Cranial fasciitis in children: clinicoradiology features and management. BMC Pediatr. 2022;22(1).
5. Paulson VA, Stojanov IA, Wasman JK, Restrepo T, Cano S, Plunkitt J, et al. Recurrent and novel USP6 fusions in cranial fasciitis identified by targeted RNA sequencing. Mod Pathol 2020;33(5):775–80.

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