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CASE REPORT

Cranial fasciitis: An unusual ethmoidal mass

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KEYWORDS
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Summary Cranial fasciitis is a rare benign tumour involving tissues in proximity to the skull that arises in children, predominantly male. As in the case presented here in a 2.5-year-old boy, it causes symptoms secondary to mass effect. This unusual presentation as an ethmoid mass presented at the medial canthus causing proptosis and epiphora. Whilst referred to sometimes as a pseudosarcoma due to its rapid growth and histological appearance, it is in fact a distinct benign lesion that is successfully treated by excision.

1. Introduction

Cranial fasciitis is an extremely rare benign tumour occurring superficial to or involving the cranial bones, first described in 1980. Little more than 40 cases have been reported in the literature. It is identical in pathological appearance to nodular fasciitis, but is distinct by virtue of its proximity to the skull and its propensity to arise in young children. It can arise from the deep fascial layer of the scalp or the underlying periosteum. Cranial fasciitis lesions have also been encountered in soft tissues of the head and neck, not directly related to the cranial bones, which some authors have termed extracranial fasciitis, though it is unclear if these represent a true separate entity [1].

The mean age of presentation is around 3 years [2] and there is a male—female ratio of 2:1. Rapid, painless growth is recognised [3] with a preoperative duration of around 2 months and lesions present with a median size of about 2.5 cm [4]. Clinically, it is a firm, non-tender mass with other symptoms secondary to mass effect. It is the rapid growth pattern that is likely to initially raise alarms and warrant prompt investigation and treatment. Accurate histological diagnosis may only be possible after complete excision of the lesion. After removal, no recurrences have been recorded, even in cases where there has been known incomplete excision. This favourable prognosis means that making the definitive diagnosis is imperative to distinguish the condition from more sinister pathologies.

2. Case report

A 2.5-year-old boy was referred with a 6-week history of epiphora and a 2-week history of proptosis with a mass palpable at the right medial canthus. The mass was asymptomatic, non-tender, firm to palpation and had normal overlying skin. There were no nasal or sinus symptoms and ophthalmology review showed no acute optic nerve compromise.

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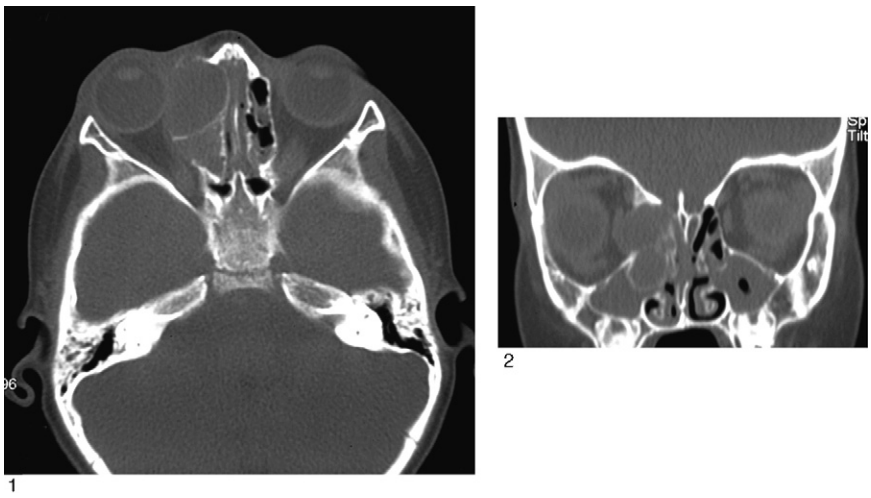

Fig. 1 CT images in the axial and coronal planes demonstrating a soft tissue mass replacing and expanding the right ethmoid complex, with medial orbital wall erosion and expansion with displacement of the medial rectus laterally.

Both computed tomography (CT) and magnetic resonance imaging (MRI) were obtained under sedation. The CT scan demonstrated a soft tissue mass replacing and expanding the right ethmoid complex. The medial orbital wall was eroded and expanded with displacement of the medial rectus laterally (Fig. 1). The MRI scan showed the right ethmoid complex to be filled with material of soft tissue density which expanded and destroyed the bone (Fig. 2). T2 weighted images were not the high signal expected of a mucocele. The possibility of rhabdomyosarcoma was raised. There was no evidence of intracranial extension and the brain itself was unremarkable.

Four days later an intranasal biopsy was performed. The mass was evident under the middle turbinate anteriorly, though the overlying nasal mucosa appeared normal. This mucosa was raised, the lateral nasal bony wall removed and biopsies of the firm grey-white mass obtained. Histological analysis showed the specimen to comprise of a spindle cell lesion of cells arranged in a vaguely storiform pattern. Focally, multinucleated giant cells were present. The overall features were suggestive of a 'fibro-osseous' lesion which was possibly benign, though a low grade malignancy could not be excluded. Lymphoma, rhabdomyosarcoma, carcinoma and melanoma were excluded as diagnoses.

Due to the lack of a conclusive diagnosis, excisional biopsy was performed just over a week later. This was performed externally, via a Lynch-Howarth incision. An approximately 3 cm mass was excised

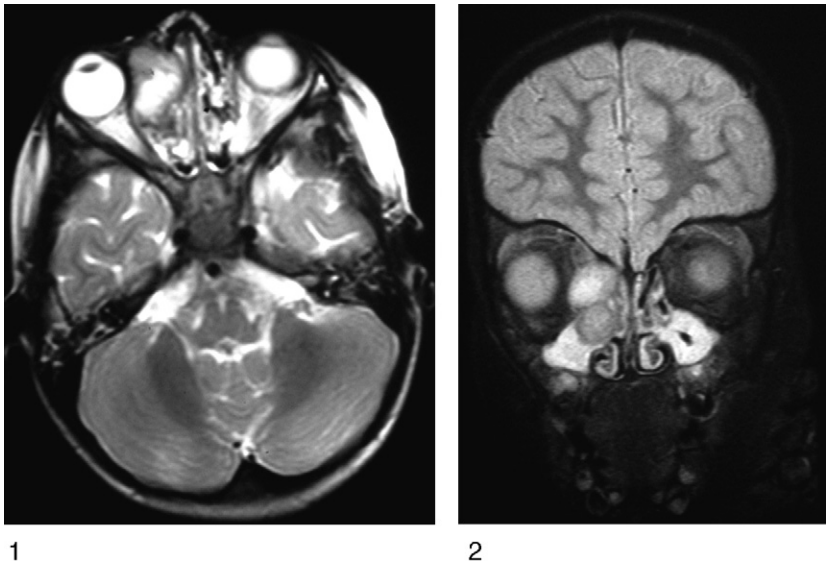

Fig. 2 MR images in the axial and coronal planes demonstrating the right ethmoid complex filled with material of soft tissue density which has expanded and destroyed the bone.