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Case Report

# Pediatric ethmoid sinus desmoplastic fibroma: Case report and review of pediatric bony sinus tumors

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ABSTRACT

While intraosseous tumors of the pediatric sinonasal tract are rare and tend to be slow growing, they can be locally aggressive and have a tendency to recur. Due to the possibility of devastating outcomes secondary to mass effect, it is important for physicians to promptly diagnose and properly manage these tumors. We report an extremely rare case of a desmoplastic fibroma of the ethmoid sinus in a pediatric patient and review its clinical findings, methods of diagnosis, and treatment.



## 1. Introduction

Jaffe first described desmoplastic fibromas in 1958. They are rare primary tumors of the bone that constitute only 0.3% of benign bone tumors. While the majority of patients are under 30 years of age, there are only a few cases of pediatric craniofacial desmoplastic fibromas described in the literature [1]. Despite its benign histological appearance and slow growth, the infiltrative nature can lead to frequent local recurrences. Clinical symptoms include headache, cranial asymmetry, orbital complaints, tenderness, and ear symptoms [2].

## 2. Case report

A previously healthy 11-year-old African American female was referred to the otolaryngology clinic after incomplete resolution from a presumed left ethmoid sinusitis. She reported a 6 month history of slowly increasing left eye protrusion and worsening tearing. Although her acute presentation included periorbital pain and fevers, she no longer complained of headaches, facial pains, purulent nasal discharge, or double vision. Ophthalmologic exam showed normal visual acuity and normal extraocular muscle movement without evidence of strabismus or diplopia. The only ophthalmologic findings noted were proptosis and epiphora. Nasal endoscopic exam revealed a yellow polypoid mass in the left middle meatus with associated mucosal edema. This ethmoid mass caused medial displacement of the uncinate process and middle turbinate. The exam failed to reveal the presence of mucopurulence.

Imaging studies were obtained, revealing an expansile osseous lesion in the left ethmoid sinus causing mass effect to the surrounding structures without evidence of local invasion. Figs. 1–3 demonstrate the characteristic findings of the lesion on CT and MRI.

In order to establish a diagnosis and recommend an appropriate therapeutic course of action, a surgical biopsy of the sinus mass was obtained. Pathology revealed normal bony architecture with copious numbers of spindle cells in a fibrous stroma as demonstrated in Figs. 4 and 5. The microscopic features were consistent with a desmoplastic fibroma.

The sinus tumor was excised in total via an external ethmoidectomy approach. An endoscopic approach was considered, but was abandoned due to the lateral extension of the mass and concerns about recurrence without a total excision. Intraoperatively, a left maxillary antrostomy, and nasolacrimal duct decompression were also performed. The child is doing well 5 months post operatively and has had no post-surgical complications. She is asymptomatic, the proptosis and epiphora have resolved and there is no evidence of recurrence on examination.

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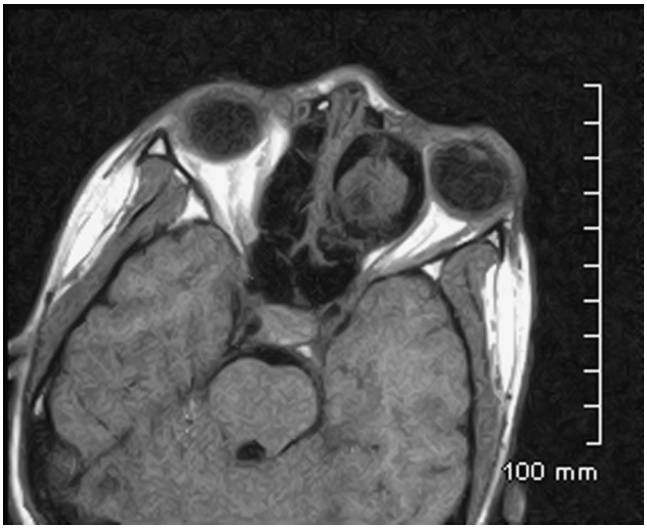


**Fig. 1.** Axial T1 weighted image demonstrates an expansile lesion within the left ethmoid air cells. The center of this lesion is isointense to skeletal muscle on T1 with a surrounding area of low/drop off signal.
*Source*: Image courtesy of Dr. Azita Khorsandi MD, Department of Radiology at New York Eye and Ear Infirmary.

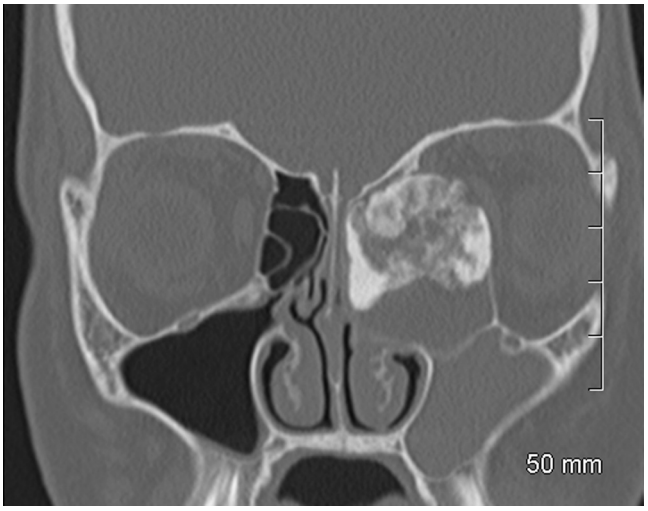


**Fig. 3.** Coronal CT images demonstrate the left ethmoid sinus lesion with significant mass effect to the lamina papyrecea and medial rectus muscle. Central mottled ossific changes are noted.
*Source*: Image courtesy of Dr. Azita Khorsandi MD, Department of Radiology at New York Eye and Ear Infirmary.

## 3. Literature review and discussion

Ossifying tumors of the pediatric sinonasal tract are rare and uncommonly reported in the literature. Desmoplastic fibroma reports are particularly and have been primarily described in the mandible and maxilla. To our knowledge, this is the first reported case of desmoplastic fibroma of the pediatric ethmoid sinus.

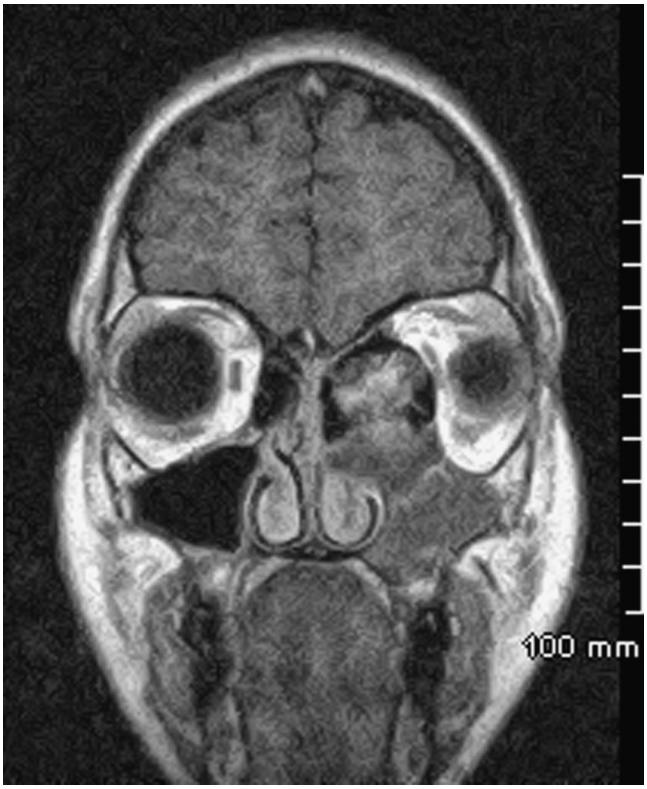


**Fig. 2.** Post contrast coronal T1 weighted images demonstrate central enhancement of the expansile left ethmoid sinus lesion with a persistent surrounding area of signal drop off.
*Source*: Image courtesy of Dr. Azita Khorsandi MD, Department of Radiology at New York Eye and Ear Infirmary.

Desmoplastic fibroma (DF) represents an extremely rare ossifying tumor of the pediatric sinonasal tract. It is generally characterized as a benign neoplasm of bone or connective tissue that can be locally aggressive, but rarely metastatic. It is more commonly found in children and is usually slow growing and painless until it begins to impinge on neighboring structures causing symptoms related to compression and mass effect [3].

Desmoplastic fibroma is more commonly found outside of the head and neck. It has been shown to involve the extremities, abdomen, pelvis, axial skeleton, gastrointestinal mesentery, and the mediastinal contents [4]. Jaffee described the first case of desmoplastic fibroma in the humerus, scapula, and tibia in 1958 [5]. In 1965 Griffith first described desmoplastic fibroma occurring in the gnathic apparatus [6]. According to Enzinger et al., approximately 23% of desmoid tumors found outside of the

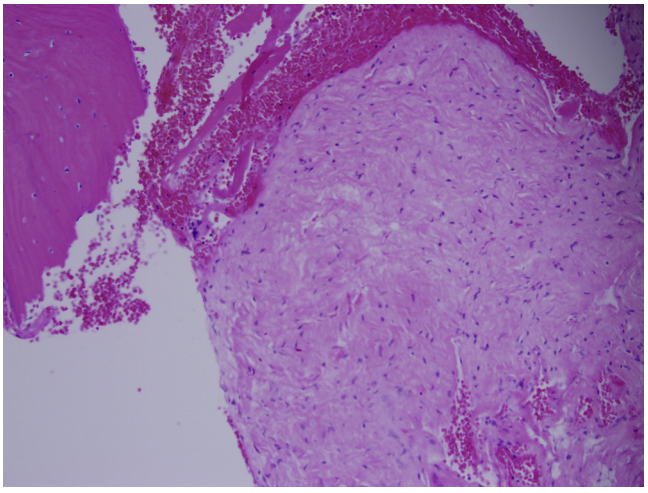

**Fig. 4.** Hematoxylin and Eosin stain of ethmoid sinus lesion at 20× magnification demonstrating normal bony architecture, but with a significant fibrous lesion with numerous spindle cells and collagen. Rare mitotic figures are noted. There is no evidence of bony invasion.
*Source*: Image courtesy of Dr. Iacob Codrin MD, Department of Pathology at New York Eye and Ear Infirmary.