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Case Report

# Sinonasal Myxoma Extending into the Orbit in a 4-Year Old: A Case Presentation

**Julian A Purrinos[1]* and Ramzi Younis[2]**

[1]University of Miami Miller School of Medicine, Rosenstiel Medical Science Building, Suite 1149, 1600 NW 10th Avenue, Miami, FL 33136, USA

[2]Department of Otolaryngology – Head and Neck Surgery, University of Miami Miller School of Medicine, Miami, FL 33136, USA

## Abstract

**Background:** Sinonasal myxomas are exceptionally rare benign tumors in pediatric patients. This report presents the case of a 4-year-old boy diagnosed with a sinonasal myxoma extending into the right orbit.

**Case presentation:** The patient's clinical presentation included moderate-angle esotropia and ocular torticollis. Advanced imaging revealed an expansile lesion in the right posterior ethmoid cavity with orbital involvement. The differential diagnosis considered included malignancies such as rhabdomyosarcoma and lymphoma, as well as benign neoplasms and inflammatory changes. A biopsy confirmed the diagnosis of sinonasal myxoma. The patient underwent a wide local resection performed by a multidisciplinary team, leading to a confirmed histopathological diagnosis of sinonasal myxoma.

**Conclusion:** This case highlights the diagnostic challenges and the importance of thorough clinical and radiologic evaluation in pediatric patients with unusual ocular symptoms. The report underscores the need for a multidisciplinary approach in managing rare neoplasms such as sinonasal myxomas.

**More Information**

***Address for correspondence:** Julian A Purrinos, BS, University of Miami Miller School of Medicine, Rosenstiel Medical Science Building, Suite 1149, 1600 NW 10th Avenue, Miami, FL 33136, USA, Email: jxp2025@med.miami.edu

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**Keywords:** Pediatric sinonasal myxoma; Pediatric orbital tumor; Differential diagnosis in pediatrics; Head and neck benign tumor in children; Pediatric rhinology

**Abbreviations:** MRI: Magnetic Resonance Imaging; ENT: Ear, Nose and Throat; CT: Computed Tomography; LEF-1: Lymphoid Enhancer-binding Factor 1; SMA: Smooth Muscle Actin; APC: Adenomatous Polyposis Coli; CTNNB1: Catenin Beta 1



## Introduction

Myxomas are benign neoplasms of mesenchymal origin, commonly found in myocardial tissue, but can also occur in the bone and soft tissues of the head and neck [1,2]. Pediatric sinonasal myxomas are exceptionally rare, slow-growing, but locally invasive tumors that pose unique diagnostic challenges [3]. Their histological appearance can resemble that of other tumors, leading to potential misdiagnosis and inappropriate or overly aggressive treatment [4,5].

In this report, we discuss a rare case of a 4-year-old boy presenting with esotropia and ocular torticollis, who was subsequently diagnosed with a sinonasal myxoma extending into the right orbit. This case underscores the importance of including sinonasal myxomas in the differential diagnosis for pediatric patients presenting with unusual ocular motility disturbances and highlights the complexities involved in managing these rare entities.

## Case presentation

### Initial presentation and history

A previously healthy 1-year-old boy presented to the ophthalmology clinic with moderate-angle esotropia, more pronounced in right gaze, suggestive of esotropic Duane syndrome. He also had associated ocular torticollis with a right-face turn. The child had no associated syndromes and was meeting growth and developmental milestones. Due to concerns about amblyopia, the patient was initially managed with intermittent patching of the left eye.

At a follow-up visit a few years later, subtle progression in his upward gaze limitation was noted, with a new onset of diplopia on upward gaze at home. Given the progression in deficits, imaging studies were ordered, including a Magnetic Resonance Imaging (MRI) scan of the face.

### Imaging and diagnostic findings

The MRI revealed a 1.8 × 1.8 × 0.8 cm expansile enhancing lesion occupying the right posterior ethmoid cavity with associated right orbital involvement. There was an abnormal appearance to the superior aspect of the right lamina papyracea/medial orbital wall secondary to pressure remodeling or lesion invasion. The lesion caused inferior

and medial displacement of the right superior oblique muscle and some effacement of the right medial rectus muscle. Differential considerations included malignancy (e.g., rhabdomyosarcoma, lymphoma, neuroblastoma), benign neoplasms, and inflammatory changes (e.g., mucocele). A CT of the orbits and sinus was recommended to better characterize the bony anatomy.

Subsequent CT of the orbits and sinus with contrast showed a similar-sized soft tissue density lesion centered in the right posterior ethmoid, which bowed and eroded the right lamina papyracea, extended slightly into the right extraconal fat, and inferiorly displaced the right superior oblique muscle. The lesion also superiorly bowed the right ethmoid roof where there was bony dehiscence but no evidence of intracranial extension (Figure 1). While the lesion did not significantly enhance on this exam, diffuse enhancement was present on the recent MRI. The differential again included malignancy versus mucocele.

**Findings:** A soft tissue density lesion centered in the right posterior ethmoid air cells which bows and erodes the right lamina papyracea, extends slightly into the right extraconal fat, and inferiorly displaces the right superior oblique muscle. The lesion also superiorly bows the right ethmoid roof where there is bony dehiscence but no evidence of intracranial extension.

### Further workup and management

The patient was referred to pediatric ENT, which performed a biopsy of the mass. The pathology report identified the lesion as a benign myofibroblastic spindle cell neoplasm, with a differential diagnosis including sinonasal myxoma and nodular fasciitis. Due to the local invasiveness of the sinus and orbital tumor, the patient underwent a wide local resection with 1 cm margins in a joint surgery performed by pediatric ENT and oculoplastics specialists. The final histopathological diagnosis confirmed a sinonasal myxoma. Histologically, the tumor exhibited a variably collagenous stroma with prominent stromal blood vessels and neoplastic cells showing fibroblastic morphology. Immunohistochemistry showed that the tumor cells were positive for LEF-1 and SMA, and negative for nuclear β-catenin. Next-generation sequencing did not identify mutations in beta-catenin or APC genes.

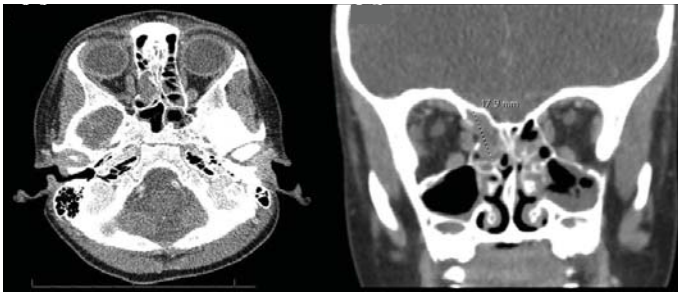

**Figure 1:** CT of the Sinus with IV Contrast.

The family was extensively counseled regarding the biopsy results and the planned management approach. Given the successful wide local resection with 1 cm margins, there is a low risk of recurrence; however, regular follow-up is still important due to the potential risk of recurrence. Discharge preparations included coordination for multidisciplinary follow-up involving ENT and Ophthalmology to monitor the patient's condition and manage any potential complications. Currently, the patient is being managed on an outpatient basis for his stable to improving esotropia and ocular torticollis, with plans for strabismus surgery in the future if alignment issues persist. The treatment plan emphasizes regular follow-up and compliance with prescribed therapies to ensure optimal outcomes.

## Discussion

Myxomas are benign, slow-growing, and locally invasive neoplasms derived from mesenchymal elements [3]. These tumors can develop in either bone or soft tissue within the maxillofacial region, with facial bones being the most common site. While the jaws are frequently affected and believed to have an odontogenic origin, rare instances of myxomas in the sinonasal area suggest a non-odontogenic origin [1,3]. Sinonasal myxomas primarily occur in the maxillary bone and paranasal sinuses. In children up to the age of 3 years, known as "infantile sinonasal myxomas," these tumors exhibit relatively indolent clinical behavior with a low recurrence rate of 5% to 10% [6]. This contrasts with other myxomas of craniofacial bones, such as odontogenic myxomas or fibromyxomas, which show a higher local recurrence rate of 20% to 40% [3,6,7].

In this case, a 4-year-old boy presented with esotropia and ocular torticollis, leading to the diagnosis of sinonasal myxoma extending into the right orbit. The rarity and indolent nature of this lesion often result in missed or delayed diagnosis. Symptoms of nasopharyngeal myxomas vary based on the site of invasion and mass effect on adjacent structures. Advanced imaging techniques like CT and MRI are essential for diagnosing and assessing the extent of these tumors. Radiographically, sinonasal myxomas appear as well-defined, expansile, radiolucent lesions that may or may not infiltrate adjacent tissue [3,8]. In our case, MRI revealed an expansile enhancing lesion, and CT better characterized the associated bony erosion.

The differential diagnosis for such a mass includes malignancies (e.g., rhabdomyosarcoma, lymphoma, neuroblastoma), benign neoplasms, and inflammatory changes (e.g., mucocele). A biopsy confirmed the diagnosis. Histologically, infantile sinonasal myxomas show variably collagenous stroma and prominent stromal blood vessels, with neoplastic cells exhibiting fibroblastic morphology [8]. Immunohistochemically, these tumors typically express nuclear β-catenin, indicating Wnt pathway activation due to CTNNB1 or APC mutations [6,9]. However, our patient's

tumor cells were positive for LEF-1 and SMA but negative for nuclear β-catenin, with no mutations in β-catenin or APC genes, suggesting an alternative pathogenetic mechanism and highlighting the genetic heterogeneity of these rare tumors.

Despite being benign, sinonasal myxomas are locally invasive and can cause bony destruction. The treatment of choice is en bloc surgical resection with a 1-cm margin, as these tumors are not responsive to radiation therapy or chemotherapy [4,8]. Limited enucleation and curettage can result in a high recurrence rate. As noted in a review of 36 literature cases of infantile sinonasal myxomas, the average follow-up period was 43 months, with a recurrence rate of 11% at an average of 8 months post-surgery [8]. The true recurrence rate for infantile sinonasal myxomas is likely much lower if complete removal is accomplished at initial surgery with 1-cm margins, as would be expected in our patient. However, complete removal is often difficult due to the anatomic location of the tumor. Long-term follow-up is always recommended. While myxomas can show aggressive local growth, they have not been reported to undergo malignant transformation or metastasis [3,8].

## Conclusion

This case highlights the unusual presentation and complexities involved in diagnosing and treating pediatric sinonasal myxomas. The rarity and indolent nature of such tumors in children often lead to initial misdiagnosis or delayed consideration. Despite these challenges, early diagnosis and a comprehensive multidisciplinary approach are crucial. This report contributes to the growing literature on pediatric sinonasal myxomas and emphasizes the need for clinical vigilance and timely intervention in managing such rare neoplasms.

### Ethical considerations

The patient's parents provided informed consent for the case details and images to be published. All procedures performed were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

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