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Sinonasal Myxoma With Intraorbital Expansion: A Rare Case

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A 9-month-old male presented for evaluation of rapidly progressing right-sided facial swelling, nasal obstruction, and epiphora. There was no associated pain, fever, chills, feeding difficulty, or weight loss. The examination was remarkable for a large right facial mass extending to the medial lower lid, nasal asymmetry, and near complete anterior nasal obstruction with septal deviation. Infraorbital nerve involvement could not be assessed due to patient age; facial nerve function was grossly intact.

Magnetic resonance imaging showed a minimally rim-enhancing cystic lesion (2.2 cm) in the region of the right nasolacrimal duct with T2 hyperintensity and no solid enhancement (**Figure 1a**). Imaging was most suggestive of a large right dacryocystocele.

The patient was taken to the operating room with plans for nasolacrimal duct stenting and nasal endoscopy with marsupialization of the presumed dacryocystocele. The right nasal cavity was found to be almost completely obstructed secondary to medialization of the right inferior turbinate. The nasolacrimal duct was patent, and no fluid was encountered during probing and stent placement; intranasal and external aspiration was attempted, but no fluid was identified. The decision was made to obtain additional imaging and plan definitive resection.

Computed tomography imaging of the face with contrast demonstrated a unilocular cystic facial lesion extending up to the right medial canthus, with adjacent remodeling of the inferomedial right orbit, right nasal bone, and right upper maxilla (**Figure 1b, 1c**). The decision was made to proceed back to the operating room for definitive excision of the facial mass. A 3-dimensional model was made and definitive surgical excision planned (**Figure 2a**).

A modified lateral rhinotomy approach (**Figure 2b-f**) afforded optimal exposure for soft tissue dissection. The lesion extended to the medial aspect of the orbit and directly involved the orbital rim. Medially, the nasal bone and the lateral nasal wall were intimately involved with the capsule of the tumor. Laterally, there was notable remodeling of the anterior maxilla with disruption of the anterior aspect of the maxillary sinus inferiorly. The mass was elevated off the underlying bone, and the opening into the maxillary sinus was carefully examined so that all abnormal tissue could be removed. The remainder of the tumor was then excised with preservation of the outer capsule. The lesion measured 3.5 × 2.5 cm. Histopathology and immunochemistry analysis revealed a myxoid neoplasm composed of small spindle cells with some enlarged nuclei and prominent nucleoli in a background of myxoid material, consistent with a sinonasal myxoma. There was no evidence of necrosis or odontogenic epithelium in the examined tissue. The patient had an uneventful postoperative course with an excellent cosmetic outcome and no sign of recurrence. A single case report does not fall within the Common Rule definition of research; therefore, we did not require Lifespan Institutional Review Board exemption or approval.

Discussion

Sinonasal myxomas are benign, locally invasive mesenchymal tumors described almost exclusively in children, with an average age of 15.4 months. They generally require excision, as they cause painless swelling followed by nasal congestion, obstruction, and epistaxis.[1] Sinonasal myxomas are often diagnoses of exclusion, with the broader differential for this type of mass including dacryocystocele, dermoid/epidermoid

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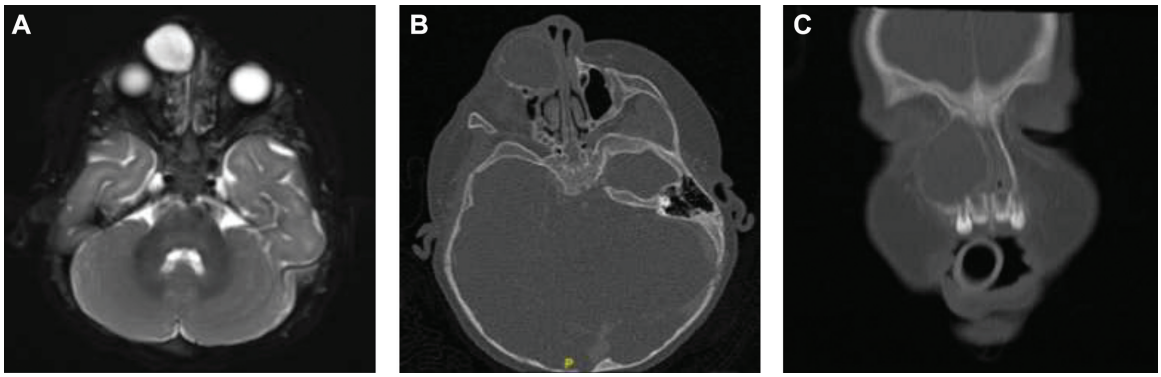

Figure 1. Magnetic resonance imaging: (a) axial T2 postcontrast image of right facial mass shows no solid enhancement. Computed tomography of the face with intravenous contrast: (b) axial and (c) coronal images show adjacent remodeling.

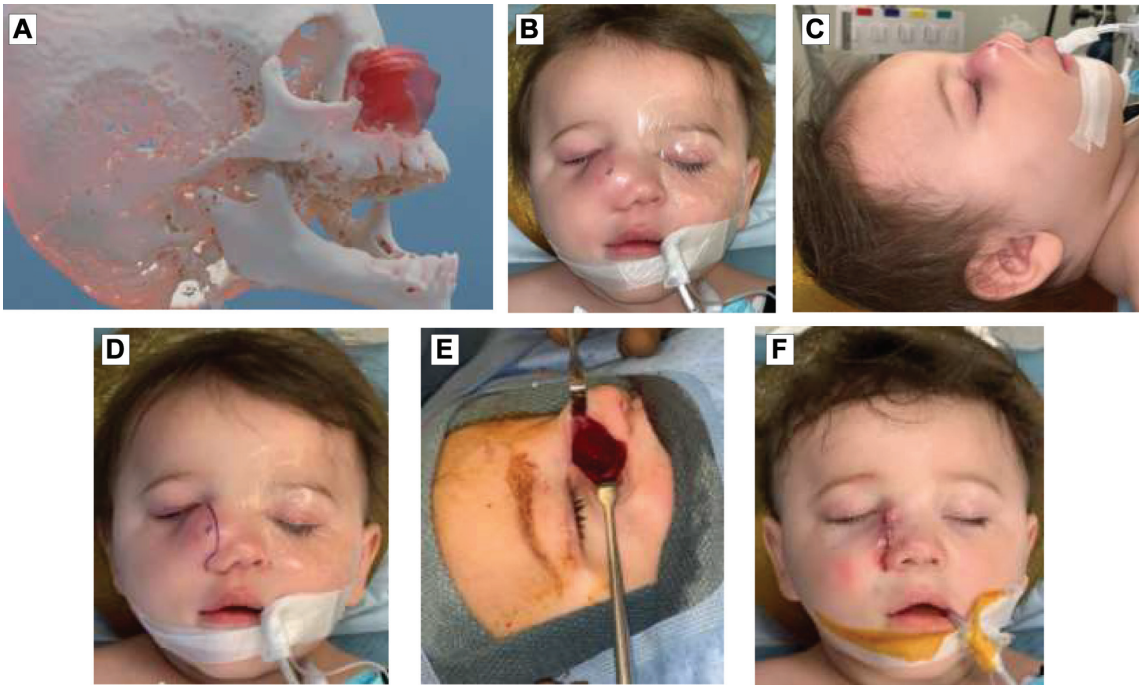

Figure 2. (a) Lateral right 3-dimensional model assisted in definitive surgical excision. (b-d) Pre-, (e) peri-, and (f) postoperative images show a right lateral rhinotomy approach and primary closure with minimal anatomic distortion.

cyst, encephalocele, nasal glioma, hemangioma, rhabdomyosarcoma, neuroblastoma, and lymphoma.[2] Although these lesions are uncommon, they are generally contained within the sinonasal complex or maxilla. Remodeling or invasion of the orbit is rare. This is the youngest case of a sinonasal myxoma with intraorbital expansion reported in the North American otolaryngology literature. The youngest patient with sinonasal myxoma reported to this audience was 5 months old and presented without bony lysis or intraorbital expansion.[2]

The radiologic characteristics of a sinonasal myxoma can mimic a cystic lesion. In this case, the proximity of the lesion to the nasolacrimal duct initially led to the presumptive yet incorrect diagnosis of dacryocystocele. The patient therefore required 2 procedures for definitive management. Our experience suggests that we maintain a high suspicion for sinonasal myxomas when assessing a child with a rapidly growing facial mass. In these cases, computed tomography is complementary to magnetic resonance imaging, providing enhanced diagnostic information and allowing for more optimal preoperative planning. Further studies are warranted to define treatment algorithms with respect to these rare tumors.

Disclosures

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Author Contributions

Neil S. Kondamuri, conception and design of this work, statistical analysis, and drafting the manuscript; analysis and interpretation of

the data; critical revision of the manuscript for important intellectual content; **Ramya Bharathi**, conception and design of this work; analysis and interpretation of the data; critical revision the manuscript for important intellectual content; **Albert S. Woo**, analysis and interpretation of the data; critical revision the manuscript for important intellectual content; **Jan C. Groblewski**, conception and design of this work; analysis and interpretation of the data; critical revision the manuscript for important intellectual content.

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