

Infant with nasolacrimal sinonasal myxoma: Diffusion MRI features

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We report the imaging features of a rare sinonasal myxoma situated over the right nasolacrimal duct in a 5-month-old male. We emphasize the importance of including sinonasal myxomas in the list of differential diagnostic possibilities when encountering a nasolacrimal gland mass in an infant, and describe the CT and MRI characteristics of this rare entity.

Introduction

Myxomas of the head and neck in infants are rare, probably representing less than 0.5% of all paranasal sinus and nasal tumors in the pediatric and adult population overall (1). Myxomas are benign, locally invasive mesenchymal neoplasms that are usually found in the mandible, representing 3-5% of all odontogenic tumors or cysts (2). Nonodontogenic myxomas arise in the nasolabial region and are termed sinonasal myxomas; these are more commonly found in the pediatric age group (3). Myxomas are believed to arise from an odontogenic, osteogenic, or soft-tissue origin; however, lesions in the sinonasal region outside of the odontogenic apparatus probably do not originate from the odontogenic mesenchyme (2). Sinonasal myxomas are exceedingly rare, and the few well-documented cases that have been reported were almost exclusively in children (3). In this report, we present the CT and MRI findings of a sinonasal myxoma in a 5-month-old boy. Imaging findings, particularly on the diffusion-weighted MRI sequence and associated ADC map, im-

proved our ability to accurately differentiate these rare tumors from other lesions that can arise within or around the nasolacrimal duct in an infant. The final diagnosis of sinonasal myxoma ultimately relies upon the microscopic

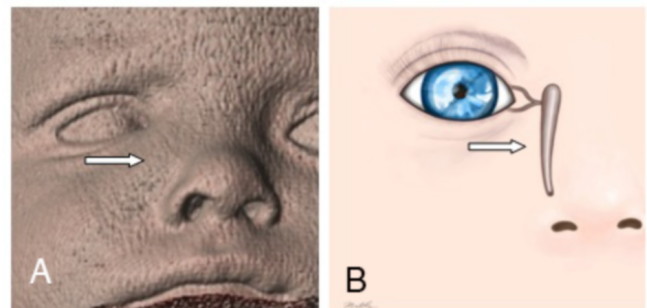


Fig. 1. **A.** 3D surface-rendered image from a CT image shows nasolabial swelling over the topography of the right nasolacrimal duct. **B.** Artist's illustration of the nasolacrimal duct corresponding to the area of swelling. Illustration courtesy of Mia S. Kelly, BA (copyright 2015).

analysis of the tissue, which is included in this report. Although rare, the possible diagnosis of sinonasal myxoma should be kept in mind when considering the usual differential diagnosis of an expansile mass over the nasolacrimal duct in an infant, especially when imaging features support this diagnosis.

Case report

A 5-month-old male developed asymptomatic focal right nasolabial swelling over the right nasomaxillary crease that extended over the right cheek (Fig. 1). The mass caused

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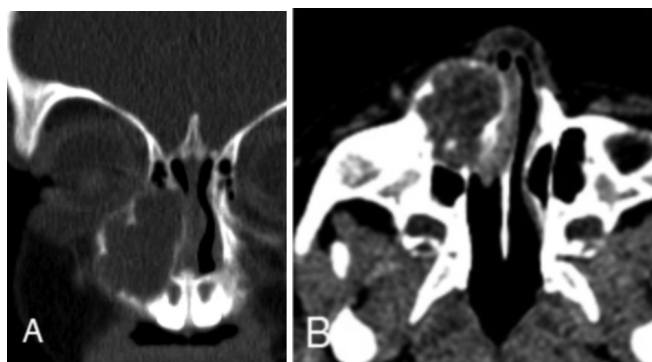


Fig. 2. Unenhanced CT (coronal and axial) images (A and B) reveal an expansile, lobulated, circumscribed, low-attenuation, soft-tissue mass centered over the right nasolacrimal groove and eroding into the left nasal cavity and inferomedial floor of the right orbit, without bony lysis.

nasolacrimal duct obstruction and epiphora on the right side. After treatment with antibiotics, there was no appreciable change in swelling. A CT scan revealed an expansile, low-attenuation, cystic-appearing mass eroding and protruding into the right nasal cavity and right inferior orbital wall, without bony lysis or intraorbital extension (Fig. 2). A followup MRI scan revealed a well-marginated, T1 hypointense (Fig. 3A), heterogeneously enhancing lesion (Fig. 3B) that was homogeneously T2 hyperintense (Fig. 3C). There was incomplete fluid suppression on the T2 FLAIR images (Fig. 3D). Small susceptibility artifacts within the mass were noted on the GRE (gradient echo) sequence, compatible with calcifications, with a corresponding punctuate density seen on the axial unenhanced CT image (Fig. 3E). Intermediate and bright signal were present on the apparent diffusion coefficient (ADC) map without restricted diffusion on the diffusion-weighted imaging (DWI) sequence, indicative of partial facilitated diffusion (Figs. 3F and 3G).

The general differential diagnosis of an expansile mass situated over the lacrimal gland includes a dacryocystocele, dermoid or epidermoid cyst, encephalocele, nasal glioma, hemangioma, rhabdomyosarcoma, neuroblastoma, lymphoma, or sinonasal myxoma. The presence of homogeneous enhancement did not support the possibility of a dacryocystocele, encephalocele, epidermoid, dermoid, or nasal glioma. There was no T1 shortening to suggest the characteristic fatty component of a dermoid cyst, and there was no restricted diffusion to suggest an epidermoid or dermoid cyst. There was no bony lysis to support the possibility of a rhabdomyosarcoma or other aggressive neoplasm. On the ADC map, there was no low signal or restricted diffusion on the DWI images that would correspond to a highly cellular lesion such as neuroblastoma or lymphoma. Furthermore, the bony lysis characteristically seen in non-Hodgkin's lymphoma of the nasal cavity was not seen (4). The cystic nature of the lesion did not comply with the imaging findings that are characteristic for hemangiomas. There was no connection with the intracranial

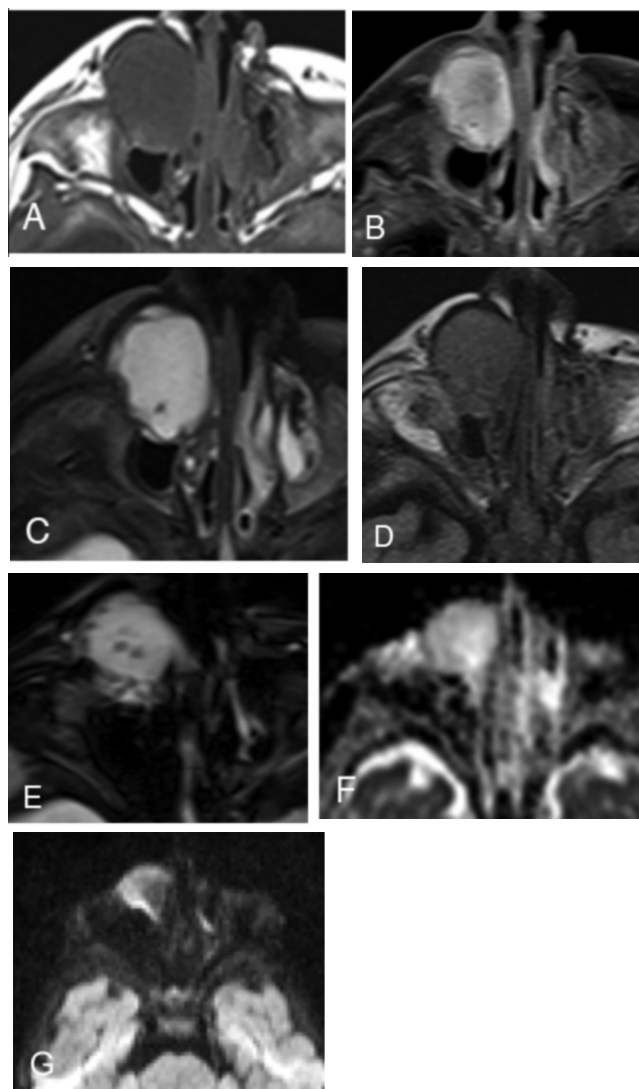


Fig. 3. **A.** T1W unenhanced axial image shows homogeneous T1 hypointensity in a well-circumscribed, low-attenuation mass over the right nasolacrimal gland. **B.** Avid homogeneous enhancement is seen on the post-contrast, fat-saturated T1 view. **C.** Homogeneous T2 prolongation is seen throughout the lesion except for a punctate focus of low T2 signal posteriorly. **D.** FLAIR-weighted images show intermediate signal (lack of fluid suppression), which suggests that the lesion is either solid or that it is a cyst containing proteinaceous material. **E.** GRE axial images reveal several small oval susceptibility artifacts that could represent degraded blood products or mineralization. **F.** ADC map shows high intermediate signal centrally and peripherally within the lesion with a posteromedial area of facilitated diffusion. **G.** There is no evidence of restricted diffusion on DWI.

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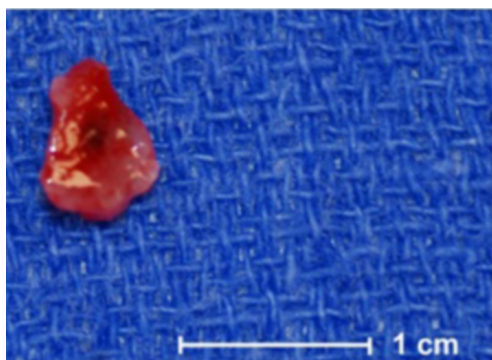


Fig. 4. Grossly, the biopsy and resection specimen consisted of pink, glistening, gelatinous soft tissue.

structures to suggest an encephalocele. Therefore, based on the CT and MRI findings of an avidly enhancing right nasolacrimal/sinonasal mass causing bony remodeling and increased diffusivity, sinonasal myxoma was favored as the most likely etiology in this case.

Right endoscopic sinus surgery was performed to open, debulk, and biopsy the right nasolacrimal mass. During surgery, bony expansion without osseous erosion was observed. The mass itself was gelatinous gray and located medial to and distinct from the the right nasolacrimal duct, which was preserved (Fig. 4).

Histological analysis revealed loose myxoid tissue interspersed with stellate and spindled fibroblasts, characteristic of a sinonasal myxoma (Fig. 5). There was minimal cytoatypia without mitotic activity consistent with a benign neoplasm. The tumor lacked the cellularity and other morphologic features of a rhabdomyosarcoma. Immunohistochemical staining for ALK1 was negative in the lesional cells, excluding an inflammatory myofibroblastic tumor. Scattered calcifications and acute (likely perioperative) hemorrhage were present in the subsequent resection specimens, but not in the initial small biopsy. Cytogenetic analysis performed on a portion of the resection specimen revealed a normal 46, XY male karyotype.

Discussion

A previous review of maxillofacial myxomas in infants reported from 1991 to 2010 revealed 14 cases in infants younger than 24 months, all of which spared the mandible. Of the 14 cases, only 8 cases were purely sinonasal. The age range was 11-20 months, with 9 boys and 6 girls (2). In addition, a case of a sinonasal myxoma was reported in a 12-month-old boy in 2010 involving the nasolacrimal duct itself (3). A maxillary myxoma was reported in a child of only 11 months of age (4). To our knowledge, the patient in our case is the youngest reported infant with a sinonasal myxoma. While CT and MRI results have previously been described in a few other similar case reports, we could find no prior description of the diffusion MRI findings in sinonasal myxomas.

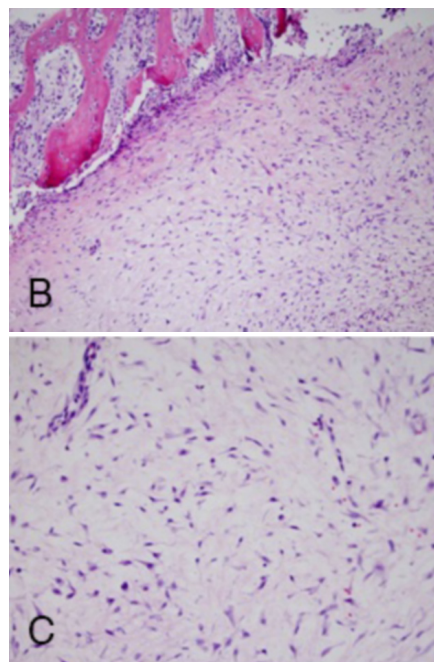


Fig. 5. **A.** Histologically, the tumor is composed of loose, myxoid tissue with numerous stellate and spindled fibroblasts. There was minimal cytoatypia, with small bland nuclei and no discernible mitotic activity. There was focal acute hemorrhage, but no necrosis was seen. **B.** The stroma was superficially infiltrated between bony spicules, eliciting relative changes that included osteoblastic rimming.

Presurgical diagnosis is important in the management of a sinonasal myxoma, because this type of lesion does not respond to chemotherapy and is poorly responsive to radiotherapy (6). Because of the unencapsulated and infiltrative nature of these tumors, extensive surgery is often required to achieve clear resection margins (7). In one unfortunate reported case, a myxoma of the nasal cavity and paranasal sinuses extensively invaded the brain and orbits, causing blindness (8).

Preoperative planning in this case report was facilitated by the CT and MRI findings. CT showed an expansile, erosive mass centered over the right nasolacrimal duct with bony remodeling, but no bony destruction. MRI showed homogeneous avid enhancement and increased diffusion, compatible with the myxoid, low-cellular tumor composition. Pathology revealed that the tumor was composed of stellate and spindled cells with wavy tapered ends scattered in a myxoid background.

Punctuate susceptibility artifacts were detected within the lesion on the GRE sequence, compatible with calcifications or hemorrhage. On unenhanced CT images, a punctuate calcification was demonstrated posteriorly within the lesion (Fig. 2b). Pathology revealed that the tumor imperceptibly merged with foci of calcification. The calcified areas tended to have a more fibrous and collagenous background than the myxoid background seen in the rest of the tumor. No hemorrhage was detected within the lesion itself.

Combined, these CT and MRI findings were unique features that appeared to be specific for the suspected sinonasal myxoma. No other type of lesion in this location should, in theory, exhibit these imaging characteristics. Minimally invasive endoscopic biopsy was performed to confirm the diagnosis. Our patient then required two addi-

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tional surgeries to achieve total tumor resection. He has since been stable without recurrence for 2½ years since his original surgery.

In summary, we have reported the unique MRI findings in a rare case of sinonasal myxoma in a 5-month-old male infant to emphasize the importance of including this entity in the differential diagnosis when encountering a nasolacrimal lesion. Furthermore, this case demonstrates that pre-surgical diagnostic accuracy can be improved by including DWI/ADC, as it can reveal facilitated diffusion due to the hypocellular myxoid composition of sinonasal myxomas when encountering an expansile sinonasal lytic lesions located in or around the nasolacrimal duct.

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