Unilateral Periorbital Swelling in a Pediatric Patient

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ABSTRACT: Infratemporal fossa (ITF) tumors are rare in children and may present with a variety of symptoms. Teratomas are neoplasms derived from the 3 germ layers and approximately 6% to 10% are within the head and neck. Our study discusses one of the first reported cases of teratoma in the ITF in a pediatric patient. A 3-year-old girl presents with 2 years of recurrent monthly left periorbital swelling accompanied by fevers, skin discoloration, and pain. Prior episodes were treated with antibiotics with incomplete resolution. Imaging revealed a cystic lesion centered in the ITF. She was taken for endoscopic endonasal biopsy of the lesion and had no complications. Pathology revealed a mature teratoma composed primarily of pancreatic tissue. Providers should consider masses such as teratoma in the differential for ITF tumors and periorbital edema unresponsive to typical treatment.

KEYWORDS: Teratoma, infratemporal fossa, proptosis

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Introduction

Pediatric skull base tumors comprise around 5% of all skull base tumors.[1,2] They commonly involve the anterior and middle skull base. Individuals typically present with visual complaints, facial swelling, headache, epistaxis, nausea, and vomiting. Pediatric skull base tumors can be either benign or malignant and the differential diagnosis is broad for tumors specifically in the infratemporal fossa.[1,2] Due to the large and diverse differential diagnosis of infratemporal fossa masses, diagnosis typically occurs with histopathology. For adolescent males the most common tumors involving the infratemporal fossa would be a juvenile nasal angiofibroma; however other pediatric ITF tumors include primary yolk sac tumors, xanthogranuloma, lipoblastoma, Burkitt lymphoma, and teratomas [3,4-6].

Teratomas are neoplasms derived from 3 germ layers and are commonly found along the midline. Only about 6% to 10% of teratomas are identified in the head and neck.[7] Teratomas are divided into mature and immature teratomas. Immature teratomas are potentially malignant and often found in early infancy with embryonic elements within the tissue.[8] Mature teratomas are considered benign but they can cause symptoms and complications depending on their location of presentation. Prior studies by Barksdale et al and Kadlub et al have suggested that age of diagnosis is an important prognostic feature independent of tumor location. The risk of malignancy is higher as age increases as a greater percentage of adults have malignant teratomas diagnosed in comparison to children, though not applicable to neonates.[7-9] The mainstay for treating teratomas involves surgical management.[3,7-13]

Case

A 3-year-old girl who had recently migrated from El Salvador presented with 2 years of monthly recurrent swelling of the left lower eyelid. She had a history of a left traumatic eye injury at 8 months of age, asthma, and resection of a benign cystic neck mass during infancy. The recurrent swelling was often accompanied by subjective fevers, discoloration, and pain. Prior episodes were treated with courses of antibiotics with some associated improvement but incomplete resolution. The child was otherwise healthy with normal growth and development.

Physical exam demonstrated a mildly tender non-erythematous swelling of the left upper and lower eyelid without conjunctival injection. There was some proptosis of the left eye, however extraocular muscles were intact, visual acuity was normal, and pupils were round and equally reactive bilaterally. No other abnormalities were noted on physical exam. Labs obtained included a complete blood count, c-reactive protein, and erythrocyte sedimentation rate which were all normal except for an elevated peripheral white blood count which was found to be 15.4×10^9/L.

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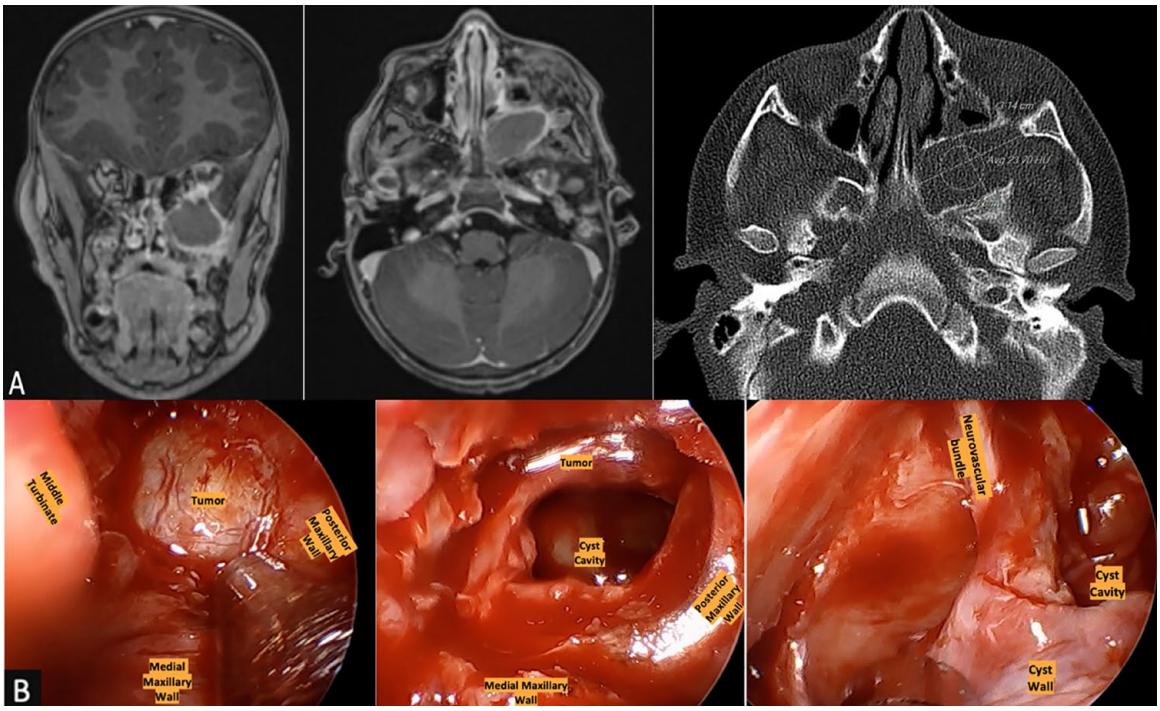

Figure 1. Large expansile mass in the infratemporal fossa and pterygopalatine fossa. (A) T1 weighted MRI with contrast showing a rim enhancing cystic lesion centered within the left pterygopalatine and infratemporal fossa and a non-contrasted CT sinus showing a 3.1 cm by 1.7 cm expansile lesion centered within the left pterygopalatine fossa without aggressive features. (B) Intraoperatively a large cystic mass was identified expanding the posterior wall of the maxillary sinus. The tumor can be seen on the posterior maxillary sinus wall, displacing it anteriorly and in close proximity of the neurovascular bundle.

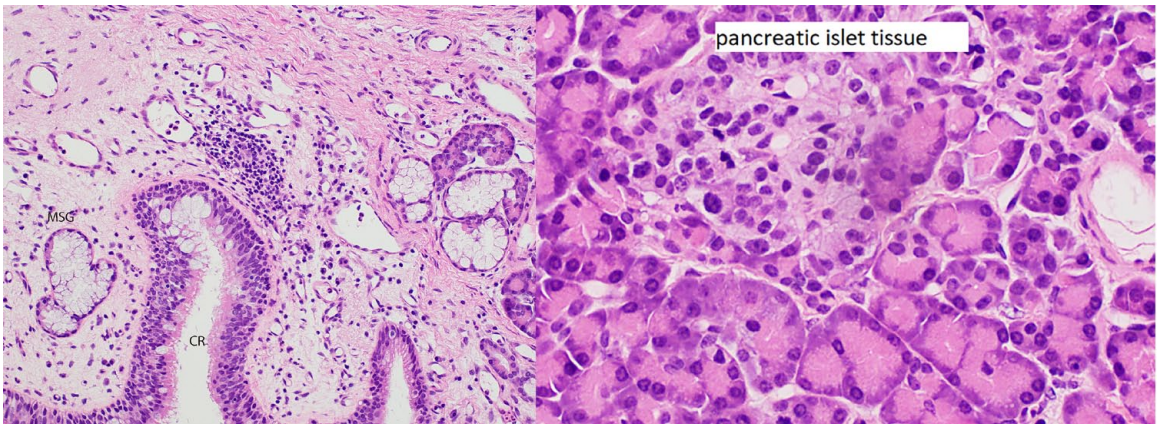

Figure 2. Histology reveals a mature teratoma. Pathology revealed a mature teratoma characterized by minor salivary glands, sino-nasal mucosa, and pancreatic tissue with exocrine cells and islets.

Imaging obtained included an MRI of the brain which revealed a T1 and T2 hyperintense, cystic lesion with an enhancing mural nodule centered in the left pterygopalatine fossa and infratemporal fossa. A sinus CT showed a $3.1 \times 1.7\,cm^2$ expansile lesion centered within the left pterygopalatine fossa without aggressive features (Figure 1).

She was taken for endoscopic endonasal biopsy of the lesion. The mass was on the posterior maxillary wall and was observed to displace it anteriorly. It was also in very close proximity to the left sphenopalatine artery and the maxillary branch of the trigeminal nerve as seen in Figure 1. The cyst was incompletely resected off the posterior infratemporal fossa, lateral pterygoid process, and posterior maxillary wall. A brown, oily fluid was encountered within the cyst. Frozen section was negative for malignancy. Fungal and bacterial cultures showed no growth. Permanent pathology revealed a mature teratoma characterized by minor salivary glands, sinonasal mucosa, and pancreatic tissue with exocrine cells and islets. The tumor tested negative for GFAP, and neurofilament as seen in Figure 2.

No complications were noted during the operation, and postoperatively she had improvement of eye swelling, no visual changes, and her extraocular movements were intact and without pain. Her postoperative course was uncomplicated, and she was discharged on postoperative day 1.

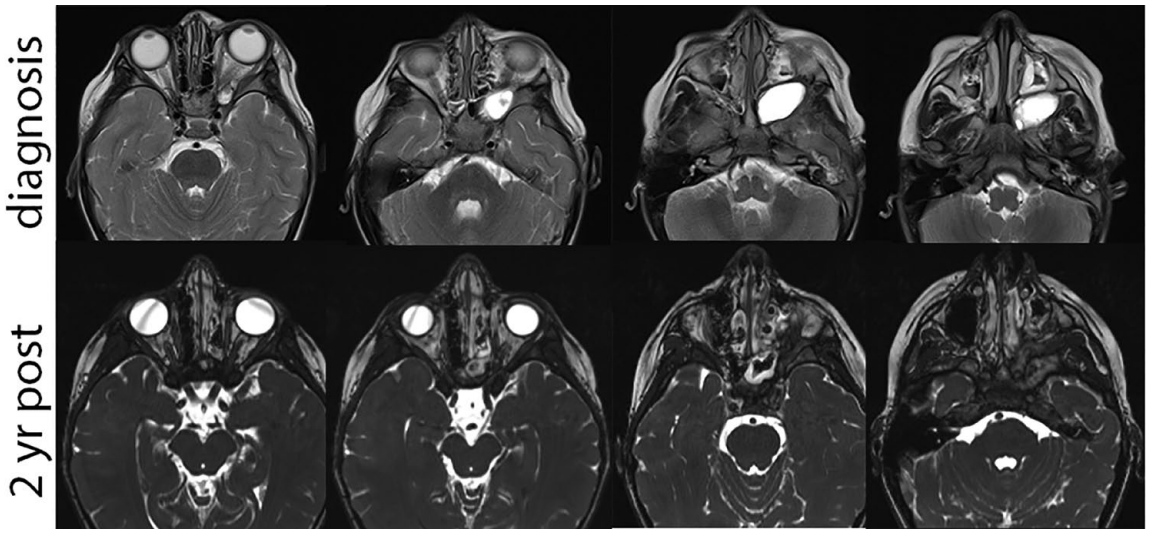

Figure 3. MRI (T2 weighted) at diagnosis demonstrates a cystic mass centered in the ITF extending into the inferior orbital fissure. Repeat MRI (2 years post-operative) demonstrates stable resection with minimal residual tumor in the infraorbital fissure. The sphenoid sinus is pneumatizing with circumferential edematous mucosa.

She was taken back to the operating room 2 months later for debridement of scar tissue and reevaluation which showed no evidence of tumor. No complications were noted at that time, and she has since been followed in clinic for a total of 22 months. Her latest MRI shows a small remnant of teratoma (Figure 3) that has been stable and will continue to be monitored with MRI imaging.

Discussion

Teratomas are rare but can present throughout the body. The incidence of pediatric cervicofacial teratomas is 1:40 000 to 80 000 live births and accounts for 2% of all neonatal tumors.[7] Mature teratomas of the head and neck, as in our case, have been reported but are unusual, and only 1 prior study reports a mature pancreatic teratoma in the infratemporal fossa.[9] The clinical presentation, as in our case, mimicked orbital cellulitis. Differentials to consider for periorbital swelling includes orbital and periorbital cellulitis, metastatic disease, ophthalmic neoplasms, nerve sheath tumors, other malignancies, autoimmune diseases, insect bite, trauma, allergies, conjunctivitis, ruptured dermoid cyst, vascular malformations, and more.[13] Ophthalmologic evaluation is an essential component in the care of such patients to evaluate visual acuity, extraocular movement, and other ophthalmic pathology. Radiology evaluation to best characterize the lesion pre-operatively and consider an extensive differential.

Potential etiologies for tumors in the ITF include pseudotumor, lymphangiomas, orbital cellulitis, ruptured dermoid cyst, optic nerve glioma, eosinophilic granuloma, leukemia, and metastatic disease.[13] Diagnosis generally requires surgical pathology. One pediatric study found that most tumors were of mesenchymal origin, including meningiomas, nerve sheath tumors, juvenile nasopharyngeal angiofibroma (JNA), and sarcomas.[2] Other differentials that are plausible, though not unique to children include schwannomas, fibromas, meningiomas, lipomas, hemangiomas, rhabdomyosarcoma, fibrosarcoma, adenocarcinoma, adenoid cystic carcinoma, osteosarcoma, and other rare tumors.[14]

Treatment for tumors in the ITF in the pediatric population is generally surgical resection or biopsy. However, there is debate about the surgical approach with some opting for an open external and others preferring an endoscopic endonasal approach. Surgical management should consider tumor location, age, and anatomical structures involved, preserving function, reducing morbidity, and strategic tumor approach.[1,2,11,14,15] In our presented case an endoscopic endonasal surgical resection was chosen to minimize long-term visual and neurologic sequalae (diplopia resolved after resection and patient reports intact sensation on her face). Given the benign nature of the tumor and the family's willingness to undergo annual surveillance imaging residual tumor was left in place to avoid injury to the inferior rectus and neurovascular injury. Had that tumor been malignant or routine medical care was not going to be feasible in the future, a more aggressive surgical resection would have been considered.

Literature on pediatric teratomas within the maxillary and infratemporal region is scarce and has been most frequently reported in neonates. We identified 1 prior case report of a teratoma in the infratemporal fossa in a neonate, 1 other report of a teratoma presenting in an infant with periorbital swelling and extensive involvement of the skull base including maxillary, ethmoid, and sphenoid sinuses, infratemporal fossa, and cavernous sinus.[11,13] One case of a mature pancreatic teratoma in the infratemporal fossa area was removed surgically via an

external approach.[9] All cases were managed surgically using a variety of approaches after imaging revealed a tumor, and diagnosis discovered with histopathology.

Our patient is one of the first reported cases of a mature pancreatic teratoma in the infratemporal fossa in a pediatric patient managed endoscopically. Barksdale et al suggested that malignant transformation of these masses has been observed previously, and semiannual imaging with serologies for 3 years is recommended.[7] In our case, the patient had a mature teratoma which is typically benign. Because this benign tumor extended into her orbital apex and abutted the inferior rectus and optic nerve, we elected to continue to monitor our patient with serial imaging. There is no consensus on the length of follow up or how to track early recurrence diagnostically though many agree that follow-up should be long term.[3,8-11] We are planning on annual follow up with MRI's for 5 years. In conclusion, pediatric providers should consider masses such as teratoma in the differential for periorbital edema that is not responsive to typical treatment. Management is dependent on many factors unique to each patient; however surgical management is recommended for tumors in the ITF.

Author contribution

AH, AP, MC: Conception and design of the work, initial drafts, and data analysis/interpretation. FA – data analysis/interpretation. AM, JS, BT, CE, BS: critical revision of the article, data interpretation. AK, KK: final approval of the version to be published, critical revision of the article, data analysis/interpretation.

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