Unusual Tumors of the Jaw in Children

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ABSTRACT

Objective
Present three cases of unusual pediatric jaw tumors, including the first reported case in the literature of an immature teratoma of the maxillary sinus.

Methods
Retrospective chart review of three patients who presented to a tertiary care center with rare jaw tumors over 15 years. Cases include a desmoplastic fibroma of the maxilla, an immature teratoma of the maxillary sinus, and infantile fibromatosis of the mandible. Their clinical presentation, imaging, pathology, treatment and outcomes are reviewed.

Results
Three unusual pediatric jaw tumors were identified. An 18-month-old male with a desmoplastic fibroma of the left anterior maxilla underwent surgical excision and is disease-free after 12 months. A 10-year-old male presented with an immature teratoma of the right maxillary sinus and underwent wide surgical resection with primary reconstruction and postoperative chemotherapy. His tumor recurred requiring a maxillectomy after which he remained disease-free. A 10-month-old male presented with infantile fibromatosis of the right mandible and underwent surgical resection. His tumor recurred after four months requiring a hemi-mandibulectomy with reconstruction. He has been disease-free for five years.

Conclusion
Jaw tumors in children are relatively uncommon and have a broad differential diagnosis. We present three rare cases, including the first reported case in the literature of an immature teratoma of the maxillary sinus. This case series may help guide care for these complex patients given the paucity of literature.

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INTRODUCTION

Jaw tumors are rare entities that can be disfiguring in the Pediatric population and carry significant morbidity. We present three cases of rare jaw tumors which required multiple procedures and, in one case, adjuvant chemotherapy and proton beam radiation. All three patients had no evidence of disease at last follow up visit.

METHODS AND MATERIALS

This is a retrospective chart review of three patients who presented to a tertiary care center with rare jaw tumors over 15 years. Cases include a desmoplastic fibroma of the maxilla, an immature teratoma of the maxillary sinus, and infantile fibromatosis of the mandible. Their clinical presentation, imaging, pathology, treatment and outcomes are reviewed.

RESULTS

Case 1: An 18 month-old male presented with a two-month history of a painless left facial mass abutting the lateral nasal wall, associated with excessive tearing of the eye and left nasal obstruction. On examination the mass was bony-hard and non-tender. It had expanded the medial wall of the maxilla and obliterated the left naso-facial fold. His extraoccular movements were intact and his intraoral exam was normal. CT imaging demonstrated a well-circumscribed expansile, lucent lesion centered in the left maxillary bone measuring 2.6 x 1.2 cm and thinning the left inferior orbital rim (figure 1). He underwent wide local excision via a mid-facial degloving approach. The mass filled the maxillary antrum, and appeared to arise from the anterolateral wall of the maxillary sinus. Histopathology revealed a desmoplastic fibroma. He was imaged six months after the procedure and underwent a second-look procedure via a Caldwell-Luc approach. No residual disease was found. He was lost to follow up a year after the procedure.

Case 2: A 10 year-old boy presented with a one-month history of right jaw pain and mild ipsilateral facial weakness followed by the rapid appearance of a golf ball sized mass. CT imaging revealed a heterogeneous soft tissue mass in the right maxillary sinus measuring approximately 4.4 cm by 4.6 cm, invading the right osteomeatal complex, pterygopalatine fossa, masticator space, and eroding the right orbital floor. Incisional biopsy revealed an immature teratoma (figure 2). He underwent resection using a facial degloving approach with calvarial bone graft and a titanium mesh for reconstruction of the right orbital floor. Three months later he returned with a recurrence in the posterior maxilla and was treated with endoscopic excision and chemotherapy. Bleomycin, cisplatin and etoposide were used. The tumor did not respond. He therefore underwent revision maxillectomy and sphenoidotomy achieving negative surgical margins. He underwent postoperative proton beam radiation therapy in 33 fractions. The patient is now followed by serial endoscopic exams and MRIs with no evidence of recurrence two years after surgery. He developed slight hypophthalmos and loss of mid-facial bulk, and underwent reconstruction of the right orbital floor with a titanium implant and composite dermato-fat graft to the right cheek. He has healed well and achieved an acceptable cosmetic result.

RESULTS

Case 3: A 10 month-old male presented with a two month history of a right neck mass which was rapidly increasing in size. It was not associated with, pain, fever, weight loss or difficulty with feeding. Examination revealed a firm, non-tender mass which was confluent with the lower rim of the right mandible. The mass was bimanually palpable and had restricted mobility. He had no cervical lymphadenopathy. The remainder of his head and neck exam was normal. Fine needle aspiration cytology was suspicious for fibrosarcoma. CT images demonstrated a large 4x5 cm globular mass continuous with the lower margin of the mandible on the right (figure 3). He underwent wide surgical excision with stripping of the mandibular periosteum around the margins of the tumor. The pathology revealed aggressive fibromatosis otherwise known as a desmoid tumor. The tumor recurred two months later which required a segmental mandibulectomy. He remained disease free for 12 months after the second procedure and continues to be followed.

DISCUSSION

Desmoplastic fibromas are rare myofibroblastic intraosseous tumors that are benign but locally invasive. While their pathophysiology is unclear, genetic predisposition, endocrine disorders and trauma are thought to be contributing factors. They represent less than 1% of all osseous tumors and may occur in any bone in the body.[1] The mandible is the most common facial bone involved (80% of cases), followed by the maxilla (16%).[2] Most cases present with gradually increasing painless facial swelling. Other symptoms may include pain (15%), tooth mobility (7%) and proptosis (2.5%).[2] Desmoplastic fibromas have non-specific radiographic appearances with either a unilocular or complex multilocular appearance. The current accepted treatment of DF involves surgical resection, but there is controversy as to the most appropriate technique. Iwai et al reported 0% recurrence with resection, 20 to 40% recurrence with enucleation and 70% recurrence with curettage alone.[2] We used a combination of wide excision and enucleation due to the proximity of the orbit and infraorbital neurovascular bundle. Wide local excision with clear histologic margins is the most widely accepted surgical approach. After resection, all patients require close follow up.

While congenital teratomas occur in 1 in 4000 births, the head and neck are rarely involved, comprising less than 2% of cases.[3] This patient does not represent a congenital teratoma, and to the best of our knowledge represents the first reported case involving the maxillary sinus in a pediatric patient. There are approximately 50 known cases of adult sinonasal teratocarcinosarcoma reported, including one case where the patient developed regional and distant metastases and was treated with multiple resections and postoperative radiation.[4] One case reported of an aggressive teratocarcinosarcoma of the nasopharynx, nasal cavity and paranasal sinuses in a 25 year-old male required surgical resection and postoperative chemotherapy and radiation.[5] Despite the paucity of reported cases, most authors agree that this disease entity warrants extirpative surgical management, with consideration given to postoperative chemotherapy and radiation therapy in the adult population. In the case reported above, multimodality was felt to be warranted given the aggressive tumor biology and recurrent nature of the disease.

DISCUSSION

Infantile fibromatosis is a tumor characterized by proliferation of fibroblasts and/or myofibroblasts originating from skeletal muscle, fascia or periosteum. They are locally aggressive, infiltrative, and may recur after excision. They most commonly occur in the abdominal wall, with 10-45% occurring in the head and neck.[6] Genetics, trauma and endocrine factors have all been theorized to play a role in the pathogenesis. Over 90% of cases present as a firm, hard, non-mobile swelling adherent to underlying soft tissue or bone. They expand quickly and are rarely accompanied by pain or other symptoms.[7] The CT appearance of infantile fibromatosis is variable. Lesions often display a similar density to muscle but may be more hypo- or hyper-dense. The cortex of the bone is rarely breached and the mass may be surrounded by a rim of cortical bone.[6] Complete surgical excision is the preferred method of treatment, however this may often be difficult given the proximity of vital structures. Close follow-up with repeat imaging is recommended given the likelihood of recurrence.

CONCLUSIONS

Pediatric patients presenting with an enlarging facial swelling warrant prompt work-up with early imaging and tissue biopsy to limit morbidity. Many of the benign tumors in this age group are locally aggressive and rapidly invade adjacent vital structures making complete surgical excision hazardous. Malignant tumors may require multimodality therapy and patients should be followed closely to ensure that recurrence is not missed.

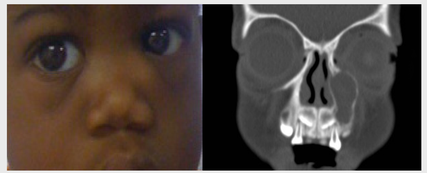

Figure 1

Figure 1. Photograph and coronal CT scan demonstrating a desmoplastic fibroma of the left maxillary sinus.

Figure 2. Photograph and coronal CT scan demonstrating an immature teratoma of the right maxillary sinus.

Figure 3.
Photograph and coronal CT scan demonstrating infantile fibromatosis of the right mandible.

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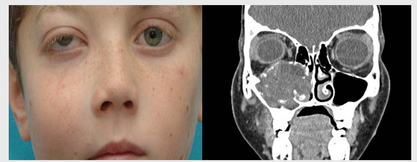

Figure 2

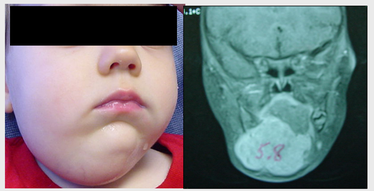

Figure 3