**ABSTRACT**

**Educational objective** To describe the basic clinical presentation of an inflammatory myofibroblastic tumor and to understand that treatment is primarily surgical.

**Objectives** To report a case of inflammatory myofibroblastic tumor (IMT) of the maxilla occurring in a child and to summarize literature recommendations for management.

**Study Design** Retrospective case report.

**Methods** The medical records of a pediatric patient diagnosed with IMT of the oral cavity were reviewed.

**Results** A five-year-old male presented to the Head and Neck Surgery clinic with an ulcerated mass involving the entire left superior alveolar ridge. Imaging revealed an expansile lesion involving the left hard palate and alveolar ridge with extension into the maxillary sinus. En bloc resection was accomplished by performing an infrastructure maxillectomy, and primary closure was achieved using a contralateral palatal flap. Final pathological diagnosis was consistent with inflammatory myofibroblastic tumor (IMT). Post-operative imaging showed no evidence of residual or recurrent disease.

**Conclusion** IMT is a benign spindle cell proliferation with an inflammatory infiltrate that may mimic sarcoma. While IMT was previously classified as a subtype of inflammatory pseudotumor, it is now recognized as a distinct entity. The upper aerodigestive and gastrointestinal tracts are affected less commonly than the lungs. To our knowledge, this is the first report of IMT presenting in the oral cavity in a pediatric patient. The mainstay of treatment is complete surgical excision. In the pediatric population, it is particularly important to limit morbidity and to preserve function; therefore, primary reconstruction, as performed in the current presentation, is ideal. Otolaryngologists must remain cognizant of this entity, and complete resection remains the primary therapeutic modality.

**INTRODUCTION**

Inflammatory myofibroblastic tumors (IMT), also known as inflammatory pseudotumors or plasma cell granulomas, are rare benign neoplasms characterized by the proliferation of spindle cells with a pattern of persistent local growth and recurrence. They are most commonly found in the lung, but also affect a variety of other anatomic sites. In early reports, patients presented with constitutional symptoms of a systemic inflammatory process, with resolution following surgical resection. Children are preferentially affected, although these tumors present in all age groups.

Given the relative rarity of the disease, no definitive risk factors have been identified. More recently, genetic investigation has revealed overexpression of anaplastic lymphoma kinase (ALK) secondary to gene fusions involving ALK at chromosome 2p23 occurring in a majority of IMT.

Only a minority of cases—in one study, 14%—arise in the head and neck. Of the head and neck sites, the larynx is preferentially affected; cases arising in the oral cavity have also been reported. Regardless of the site of origin, the mainstay of treatment is surgical resection. Despite aggressive surgical management, recurrence is a threat, even many years following treatment.

Fewer than 10 cases of inflammatory myofibroblastic tumor affecting the gingiva have been reported. There have been no published reports of IMT involving the oral cavity in a pediatric patient. We present a case of IMT occurring in a child, treated with resection and local flap reconstruction, and summarize literature recommendations for management.

**CASE PRESENTATION**

A five-year-old male presented to the Head and Neck Surgery clinic complaining of episodic bleeding from the oral cavity. Examination was remarkable for an ulcerated mass involving the entire left superior alveolar ridge. Imaging revealed an expansile lesion involving the left hard palate and alveolar ridge with extension into the maxillary sinus (Figures 1 and 2). A prior incisional biopsy, performed by an oral maxillofacial surgeon, had proven inconclusive.

Because of the invasive appearance of the disease and a prior inconclusive biopsy, the decision was made to perform an en bloc resection with immediate local reconstruction. Under general anesthesia, an infrastructure maxillectomy was performed, and primary closure of the resultant defect was achieved using a contralateral palatal island flap.

The patient was admitted postoperatively, and an oral diet was started on post-operative day 3. Permanent section pathological analysis revealed an inflammatory myofibroblastic tumor (IMT). The patient was followed for 3 months post-operatively until he was lost to follow-up at one month post-operatively, imaging revealed no evidence of residual or recurrent disease.

**DISCUSSION**

Inflammatory myofibroblastic tumors (IMT), variously referred to as inflammatory pseudotumors or plasma cell granulomas, are benign but locally aggressive neoplasms that occur most commonly in the lungs. Of the extrapulmonary sites, the abdomen and pelvis are most commonly affected; less common sites include the head and neck, including the upper aerodigestive tract, and the extremities. Of the head and neck sites, numerous cases have been reported in the larynx, with fewer cases reported arising in the oral cavity.

The lesion has a predilection to arise in children, with one series of 84 cases reporting a median age at presentation of 9 years. The mainstay of treatment is complete resection, as local recurrence or persistence is the main cause of morbidity and mortality. In the pediatric population, however, wide local resection may be associated with consequences for future development, and some authors have accordingly advocated complete but judicious, resection.

In the case presented, an infrastructure maxillectomy was performed to extirpate disease. Immediate reconstruction was performed by using a contralateral palatal island flap, a well-described method of closure for lesions of the hard palate, soft palate, or retromolar trigone. At latest follow-up, the patient had no radiological evidence of residual or recurrent disease, and was tolerating an oral diet without symptoms of oroantral fistula.

**CONCLUSION**

Inflammatory myofibroblastic tumors are uncommon benign mesenchymal neoplasms with aggressive local behavior. While the lungs are most commonly affected, these tumors also originate in a variety of sites in the head and neck. Children are preferentially affected, and complete resection is the mainstay of treatment. The pediatric otolaryngologist must remain cognizant of this entity in the evaluation of a patient with a locally aggressive tumor.