INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare, mesenchymal low-grade neoplasm characterized by benign histology but has the potential of showing aggressive clinical behavior with local invasiveness and recurrence. It primarily occurs in children and young adults in the lung and abdominopelvic regions, but there are reports of oral cavity IMTs. In this report, the authors present a case of a neonate born with an enlarging tongue mass whose biopsy revealed IMT with anaplastic lymphoma kinase (ALK) positivity. Treatment included tumor debulking and an ALK inhibitor, crizotinib, which resulted in complete remission. To our knowledge, this is the only reported case of oral cavity IMT with ALK positivity in a neonate.

CASE REPORT

A 0-day-old neonate, born without complication at 41 weeks, presented as a transfer from an outside facility to Children's Mercy Hospital (CMH) in Kansas City, Missouri for further evaluation and management of a large, bleeding tongue mass. Of note, there was reported comprehensive antenatal care without complication or detection of the tongue mass on three prenatal ultrasounds. At birth, the tongue mass was noted. The patient did not have significant airway obstruction with APGARs 8 and 9 at one and five minutes of life, respectively. On physical exam, there was a large, firm, oozing tongue mass with transition from normal appearing tongue mucosa to necrotic and atypical tongue mucosa roughly halfway posterior on the lingual surface (Figure 1). The patient’s airway was widely patent on flexible laryngoscopic exam, and she was saturating well on room air.

The patient then underwent transnasal intubation in the operating room and magnetic resonance imaging (MRI). MRI face with and without contrast showed a large mass that filled and distorted the normal anatomy of the oral cavity and floor of mouth with extent inferiorly and laterally to the level of the mandible without bony extension and superiorly to the level of the hard

ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare, mesenchymal low-grade neoplasm characterized by benign histology but has the potential of showing aggressive clinical behavior with local invasiveness and recurrence. It primarily occurs in children and young adults in the lung and abdominopelvic regions, but there are reports of oral cavity IMTs. In this report, the authors present a case of a neonate born with an enlarging tongue mass whose biopsy revealed IMT with anaplastic lymphoma kinase (ALK) positivity. Treatment included tumor debulking and an ALK inhibitor, crizotinib, which resulted in complete remission. To our knowledge, this is the only reported case of oral cavity IMT with ALK positivity in a neonate.
Figure 2. (A) MRI T2 sequence showing a large tongue mass that is heterogeneously hyperintense without vascular flow voids measuring 3.4 x 5.5 x 3.1 cm. (B) MRI T2 sequence 5 months post-treatment. MRI, magnetic resonance imaging.

Figure 3. Shrinkage of tongue mass after initiation of chemotherapeutic agent, crizotinib. (A) At 2 months of age. (B) At 18 months of age.

**CASE REPORT**

Biopsy of the tongue mass, tracheotomy, and gastrostomy tube placement occurred. Pathology, which was analyzed by CMH and an outside facility, was consistent with a congenital inflammatory myofibroblastic tumor with ALK, S100-protein, smooth muscle actin (SMA), desmin, and p53 positivity.

Traditional therapy would be complete resection, but in this patient would require subtotal glossectomy. Due to the high morbidity with this procedure, we received compassionate approval for targeted therapy with an ALK inhibitor, crizotinib. Prior to initiation of therapy, due to the necrotic nature of the tumor, dubulking was performed. Despite initial growth of the tumor, after twelve months of receiving crizotinib, there was good response with noticeable shrinkage of the tumor (Figure 3A). The patient did have prolongation of her QTc interval, though a normal baseline echocardiogram, which can be a side effect of crizotinib. She underwent serial electrocardiograms (EKGs) during treatment to monitor her QTc interval, which normalized after cessation of crizotinib. The patient was last seen at 18 months old, off of therapy for eight months without signs or symptoms of recurrence with no detectable tumor by MRI imaging (Figure 2B). She has been decannulated with removal of her gastrostomy tube without difficulties talking, swallowing, or breathing (Figure 3B).

**DISCUSSION**

**Overview**

Inflammatory myofibroblastic tumor is an overall rare, mesenchymal neoplasm that occurs primarily in the lung and abdominopelvic regions, with only 14-18% of the extrapulmonary IMTs occurring in the head and neck region [5]. Of that small subset of head and neck occurrences, the orbits and upper airways are most common [5]. Our study adds to the literature in that it is a tongue IMT, positive for ALK, in a neonate.

**Pathological, Histological, and Imaging Features**

Extrapulmonary cases of inflammatory myofibroblastic tumor show a combination of spindled fibroblastic and myofibroblastic cells with inflammatory infiltrate of lymphocytes, eosinophils and plasma cells [17]. The pathology in our patient showed relatively monotonous sheets of cells with ovoid to spindle-shaped nuclei, indistinct pale-gray cytoplasm with a delicate slightly vacuolated chromatin pattern (Figure 4). Mitotic figures and foci of necrosis visible. Other features are prominent hemangiopericytomaous vasculature with lymphoctic infiltrate and focal collections of osteoclast giant cells. Fluorescence in situ hybridization (FISH) was positive for ALK in our patient. Positivity for the ALK gene has been reported in approximately 50% of IMTs [1,18]. The diagnosis of IMT requires histo-
Inflammatory myofibroblastic tumor is a low-grade malignancy that rarely occurs in the head and neck. To date, there are no reportable cases of IMT with ALK positivity occurring in the tongue in a neonate. Though, surgical resection has been the mainstay of treatment for IMTs, if positive for ALK, crizotinib may be considered. This study shows complete remission in an enlarging tongue mass in a neonate treated with crizotinib with no signs of recurrence at eight months follow-up. Therefore, it is important for clinicians to consider other treatment options for IMT in select patient populations.

**REFERENCES**


