INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare, mesenchymal neoplasm primarily occurring in the lung and abdominopelvic regions in children and young adults with a prevalence ranging from 0.04-0.7% [1-3]. IMT presents as a painless, enlarging mass and may show invasiveness and local recurrence but is rarely metastatic [4].

Histologic characteristics include a mixture of inflammatory and myofibroblastic spindle cells with hypocellular fibrous patterns that can be well-circumscribed to ill-defined on imaging [1,3]. Surgical intervention have been mostly noted as the mainstay treatment for IMTs, though chemotherapy agents and anti-inflammatory therapies such as steroidal and non-steroidal anti-inflammatory drugs have been reported, as well [5].

To date, there are published cases of IMT involving the head and neck, including the oral cavity, but to our knowledge, there are no reported studies of inflammatory myofibroblastic tumor with anaplastic lymphoma kinase (ALK) positivity of the tongue in a neonate [2,4,6-16].

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
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.

**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 hemangiopericytomatous vasculature with lymphocytic infiltrate and focal collections of osteoclastic 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-
CASE REPORT

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.

ARTICLE INFORMATION

*Correspondence: Maraya M. Baumanis, MD. Department of Otorhinolaryngology-Head & Neck Surgery, The University of Kansas Medical Center, Otolaryngology, 3901 Rainbow Blvd, Kansas City, KS 66160, USA. Email: mbaumanis@kumc.edu

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Outcomes

Outcomes seem to be substantial in surgical interventions, whether complete or partial resections, for treatment of IMTs [5]. Further, when observing the use of an ALK inhibitor agent in ALK positive IMTs, Thielen et al. reported achievement of partial to complete remission in 40% (12/30) patients with use of crizotinib [23]. Another study observed metastatic or inoperable ALK+ IMT reported the overall response rate for patients with IMT was 86% (N=14) with a partial response seen in 36% (5/14) [24]. The recurrence rate has been reported to be low with a 10-year survival rate at approximately 80% [1]. Our unique case showed complete clinical and radiological remission without recurrence at 8 months post-treatment with use of crizotinib.

CONCLUSION

Figure 4. (A) Pathology showing monotonous sheets of cells with ovoid to spindle-shaped nuclei, indistinct pale-gray cytoplasm with a delicate slightly vacuolated chromatin pattern, 200 x. (B) Other features include prominent hemangiopericytomatous vasculature with lymphocytic infiltrate and focal collections of osteoclastic giant cells, 200x. (C) Cells stained for anaplastic lymphoma kinase positivity, 400x.
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