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Case Report

Treatment of an obstructive, recurrent, syncytial myoepithelioma of the trachea with tracheal resection and reconstruction

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ABSTRACT

Myoepithelioma is a rare occurrence in the trachea and respiratory tract with only 11 cases reported in the literature. We present a case report of a 10-year-old female who was found to have an anterior tracheal mass causing near total obstruction of the airway on bronchoscopy. Characteristics of the mass were consistent with syncytial myoepithelioma. The patient experienced multiple recurrences requiring tracheal resection with end-to-end reanastomosis. To date there have not been any reported cases of myoepithelioma of the trachea in a child and no reports of syncytial myoepithelioma in the trachea or respiratory tract.

1. Introduction

Myoepithelioma by definition is a benign growth of myoepithelial cells that lack a definitive ductal framework, without chondroid or myxochondroid stroma [1,2]. Some pathologists will consider up to 4.99% ductal framework in the whole section [2]. These cells are similar in structure to smooth muscle and have been described in salivary, lacrimal, mammary, prostate, and sweat glands [3]. Most lesions have been reported in subcutaneous or deep fascial soft tissues of the extremities and in major and minor salivary glands in the head and neck [1]. Myoepitheliomas have also been reported in the lung, trachea, breast, ovaries, nose, and bone [4–17]. Malignant tumors are differentiated from benign tumors by tumor necrosis, vascular and lymphatic invasion, higher mitotic rate, prominent cellular pleomorphism, and atypia [1,2].

Four different cell subtypes of myoepithelioma have been described: spindle, plasmacytoid, epitheliod, and clear cell [2]. These cells typically have a multinodular or lobular architecture with nested or reticular growth within a collagenous or chondromyxoid stroma [18]. One study described a distinct type of growth pattern in cutaneous myoepitheliomas, the syncytial variant. This is characterized by syncytial growth with no stromal elements. In this series of 38 cases, the syncytial variant consistently stained positive for epithelial membrane antigen (EMA) and S-100 with only 5 of the cases staining positive for keratin [18].

Myoepithelioma is a rare occurrence in the trachea and respiratory tract with only 11 cases reported in the literature [4–13]. We present a case report of a 10-year-old female who was found to have an obstructing tracheal mass with pathologic characteristics of syncytial myoepithelioma. To date there have not been any reported cases of myoepithelioma of the trachea in a child and no reports of syncytial myoepithelioma in the trachea or respiratory tract in the literature.

1.1. Case report

A 10-year-old female with history of asthma presented to our institution from an outside hospital after an episode of shortness of breath, tachypnea, accessory muscle use, and hypoxia. The patient's condition worsened requiring intubation for 5 days. Following extubation, the patient developed intermittent expiratory stridor but was otherwise stable without difficulty breathing, tachypnea, or respiratory distress. Approximately six weeks later, the patient was seen by our pediatric otolaryngology department and underwent direct laryngoscopy and bronchoscopy which revealed a white, pedunculated, anterior tracheal mass 2–3 cm below the glottis with near total obstruction of the airway (Fig. 1). Biopsy of the mass was performed, followed by endotracheal intubation and transfer to the intensive care unit.

MRI of the neck with contrast revealed a nodular, homogenously enhancing mass of the right trachea just inferior to the thyroid gland measuring 1.4 x 1.1 × 1.9 cm (cm) that was hypointense on T1

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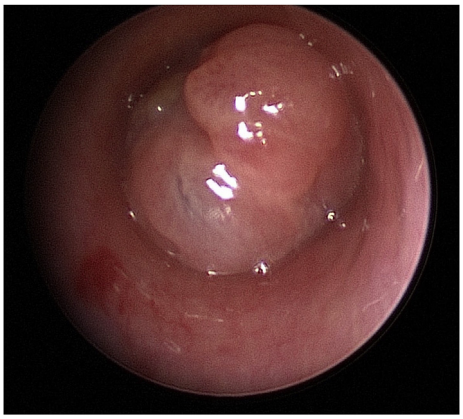

Fig. 1. Bronchoscopy image revealing a white, pedunculated, anterior tracheal mass 2–3 cm below the glottis with near total obstruction of the airway.

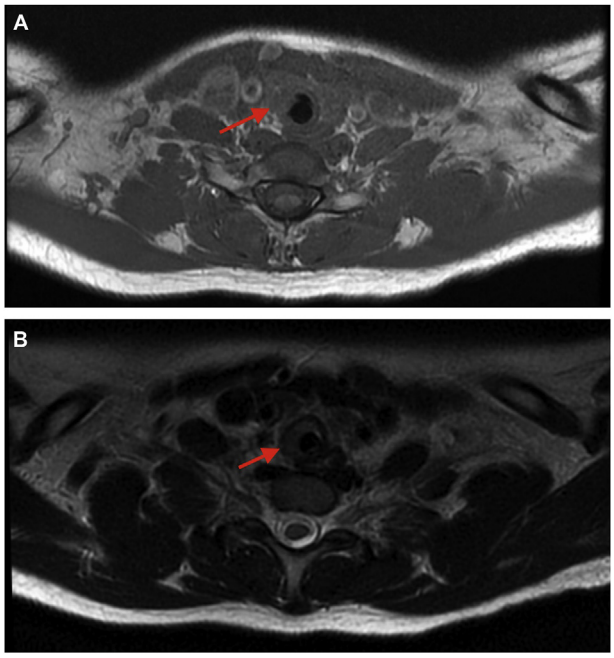

Fig. 2. MRI of the neck with contrast revealing a nodular, homogenously enhancing mass of the right trachea just inferior to the thyroid gland measuring 1.4 x 1.1 × 1.9 cm (cm) that was hypointense on T1 *(a)* and expressed intermediate signal on T2 *(b)*.

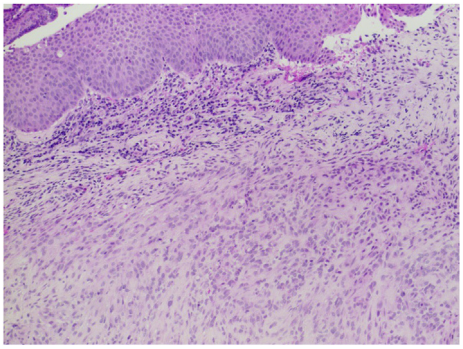

Fig. 3. Histologic examination of the tissue biopsied showing a relatively circumscribed submucosal proliferation of cytologically bland ovoid and epithelioid cells arranged in syncytial sheets.

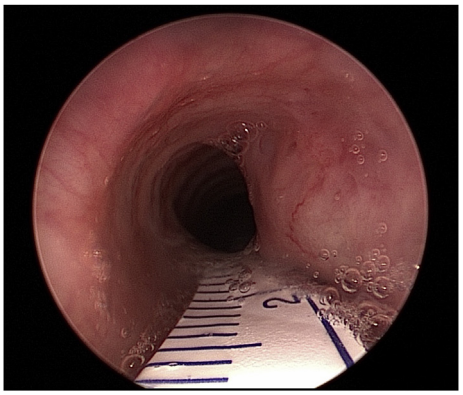

Fig. 4. Bronchoscopy image showing regrowth of the mass along the right anterolateral trachea 2.5–3 cm below the glottis spanning the second through fourth tracheal rings.

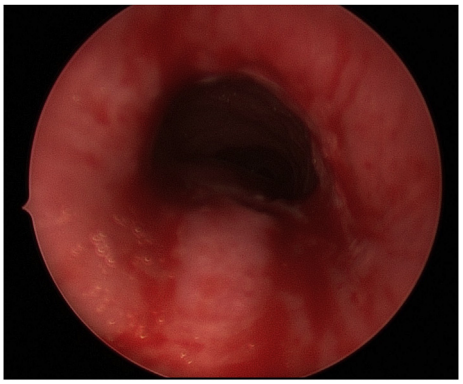

Fig. 5. Bronchoscopy image after tracheal resection with end-to-end anastomoses showing slight tracheal narrowing but no signs of recurrence.

(Fig. 2a). This mass expressed intermediate signal on T2 (Fig. 2b). Following MRI, the patient was taken back to the operating room and the mass was excised using both a carbon dioxide (CO2) laser and microdebrider and a safe airway was established. Histologic examination of the tissue biopsied showed a relatively circumscribed submucosal proliferation of cytologically bland ovoid and epithelioid cells arranged in syncytial sheets (Fig. 3). The cytoplasm was eosinophilic and the nuclei were uniform and vesicular without atypia or pleomorphism. Mild lymphocytic infiltrates were seen mostly in the periphery. The neoplasm was positive for EMA, S-100, SMA, and DOG-1. The histologic findings and immunohistochemistry staining pattern was consistent with syncytial myoepithelioma.

Over the next 7 months the patient was reevaluated in the operating room four times. The first two evaluations, two and three months post-initial excision respectively, showed minor regrowth of the mass and excision with the microdebrider was performed. The third evaluation, four and a half months post-excision, showed scar formation without regrowth. The fourth evaluation seven months post-initial excision showed regrowth of the mass along the right anterolateral trachea 2.5–3 cm below the glottis spanning the second through fourth tracheal rings (Fig. 4). Subsequently, the decision was made to forego further endoscopic excision and plan for tracheal resection with end-to-end anastomosis.

Intraoperatively, upon dissecting to the level of the trachea, the mass was noted to extend beyond the right lateral trachea but not into adjacent strap muscles or surrounding soft tissues. The trachea was resected below the fourth tracheal ring and also below the second tracheal ring. Resection the trachea was followed by end-to-end anastomoses. Post-operatively, she has been evaluated with an outpatient history and physical examination every 3 months for the first year and