Myoepitheliomas are most commonly seen in the head and neck as benign salivary gland tumors; however, there have been limited numbers of reports occurring in the trachea and lung. Recently, a variant of myoepithelioma has been reported in the skin and soft tissue with syncytial growth pattern. We present the first case in the literature of a young adolescent to have a myoepithelioma of the tracheobronchial tree, and the first case of syncytial variant myoepithelioma of the tracheobronchial tree.

INTRODUCTION

Myoepithelial cells are contractile cells that are classically found in glandular epithelium above the basement membrane, assisting in glandular secretion. These cells are similar in structure to smooth muscle and have been described in salivary, lacrimal, mammary, prostate, and sweat glands. Myoepithelioma by definition is a benign growth of myoepithelial cells that lack a definitive ductal framework, without chondroid or myxochondroid stroma. Some pathologists allow up to 4% ductal framework in the whole section. These tumors have most commonly been reported in subcutaneous or deep fascial soft tissues of the extremities and in major and minor salivary glands in the head and neck. Myoepithelialomas have also been reported in the lung, prostate, and sweat glands.

Myoepithelialomas have a characteristic histology with nested or reticular growth within a collagenous or chondromyxoid stroma. Jo et al described a distinct type of growth pattern in cutaneous myoepitheliomas, the syncytial variant. This is characterized by syncytial growth with no stromal elements. In their series of 38 cases, the syncytial variant consistently stained positive for epithelial membrane antigen (EMA), p63, and S-100 with only 5 of the cases staining positive for keratin. 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 obstructing tracheal mass with histologic characteristics of 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.

CASE PRESENTATION

A 10 year old female with history of asthma presented in May, 2015 from an outside hospital after an episode of shortness of breath, tachypnea, accessory muscle use, and hoarseness. The patient’s condition worsened requiring intubation for 5 days. Following extubation the patient developed intermittent inspiratory stridor but was otherwise stable without difficulty breathing, tachypnea, or respiratory distress. Roughly six weeks later, the patient was seen by our 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 (Figure 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 x 1.9 cm that is hypointense on T1 and expresses intermediate signal on T2 (Figure 2).

Following MRI the patient was taken back to the operating room and the mass was excised using both a CO2 laser and microdebrider and a safe airway was established. Pathologic evaluation revealed fragments of spindle cell proliferation with epithelial cells arranged in syncytial sheets, atypia and pleomorphism were absent. Immunostaining showed strong positivity for EMA and multifocal positivity for S-100 protein. In addition, SMA, vimentin, and beta-catenin staining were positive; AE1/AE3, HM45, desmin, CD58, CD45, MyoD1, Myogenin, CAM5.2 and CD117 were all negative. Deep margins were positive for tumor. During the next 7 months the patient was re-evaluated in the operating room four times. The first two evaluations, and 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 antero-lateral trachea 2.5 cm below the glottis spanning from the second to the fourth tracheal ring (Figure 3). Decision was made to forego further endoscopic excision and plan for tracheal resection with end to end anastomosis at a later date.

The patient underwent excision with re-anastomosis in February 2016. 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. Follow up endoscopy and MRI show slight tracheal narrowing but no sign of recurrence (Figure 4).

Four different cell subtypes of myoepithelialoma have been described: spindle, plasmacytoid, epitheloid, and clear cells. These cells typically have a multinodular or lobular architecture with or without reticular growth within a collagenous or chondromyxoid stroma.

CONCLUSION

Myoepithelioma is a rare, benign entity that can occur in the trachea and respiratory tract. Treatment is best achieved with surgical resection. Both surgeons and pathologists need to be aware of this entity when a patient with a tracheal or lung mass is encountered.

REFERENCES