Syncytial myoepithelioma of the trachea in a 10 year old female: case report and review of the literature

Michael Pfeiffer D.O.[1], Sri Chennupati M.D.[2]

[1]Department of Otolaryngology, Philadelphia College of Osteopathic Medicine & St. Christopher Childrens Hospital, Philadelphia, PA
[2]Department of Otolaryngology, Drexel University College of Medicine, Philadelphia, PA
Program Director: John McGrath D.O.

ABSTRACT

Myoepitheliomas are most commonly seen in the head and neck as benign salivary gland tumors; however, there have been a limited number 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.[1] These cells are similar in structure to smooth muscle and have been described in salivary, lacrimal, mammary, prostate, and sweat glands. [1] Myoepithelioma by definition is a benign growth of myoepithelial cells that lack a definitive ductal framework,[2] without chondroid or myxochondroid stroma.[3] Some pathologists allow up to 4.99% ductal framework in the whole section.[3] 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.[2] Myoepitheliomas have also been reported in the lung,[4,5,6,7,8,9, 10, 11] trachea,[12, 13] breast,[14] ovaries,[15] nose,[16] and bone.[17] Malignant tumors are differentiated from benign tumors by tumor necrosis, vascular and lymphatic invasion, higher mitotic rate, prominent cellular pleomorphism, and atypia.[3, 18]

Four different cell subtypes of myoepithelioma have been described: spindle, plasmacytoid, epitheliod, and clear cells.[3] These cells typically have a multinodular or lobular architecture with nested or reticular growth within a collagenous or chondromyxoid stroma.[19] 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) and S-100 with only 5 of the cases staining positive for keratin.[19]

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.

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 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. 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.

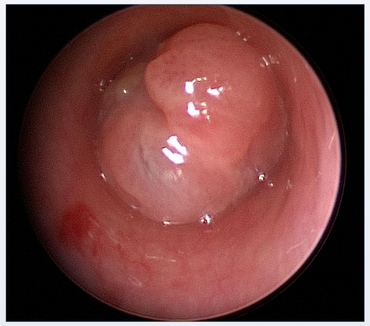

Figure 1: Initial bronchoscopy showing obstructive tracheal mass

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).

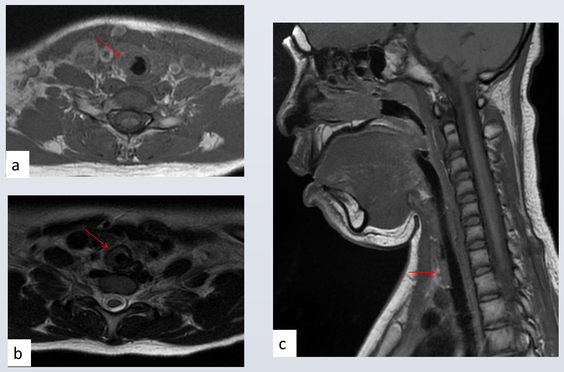

Figure 2: MRI of tracheal mass with patient intubated; arrows denote mass. a) Axial T1 weighted MRI; b) Axial T2 weighted MRI; c) Sagittal T1 weighted MRI.

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 epitheliod 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, HMB45, desmin, CD58, CD45, MyoDi, 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, 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 2.5-3cm 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.

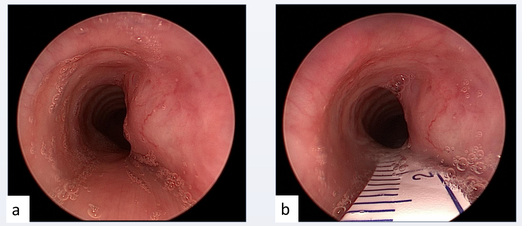

Figure 3: Residual right tracheal mass 2 cm below the glottis.

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).

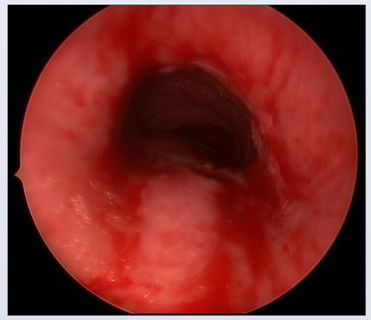

Figure 4: Post operative bronchoscopy status post tracheal resection and end to end anastomosis showing mild tracheal stenosis and no sign of recurrence

DISCUSSION

Tracheal masses are rare in general. Patients with tracheal masses can present with stridor and dyspnea and can be mistaken to have other more common diseases such as asthma, COPD, tracheomalacia, or congestive heart failure. Symptoms of stridor warrant fiberoptic nasopharyngolaryngoscopy performed by an otolaryngologist. If no obvious cause of stridor is seen on exam, bronchoscopy or computed-tomography of the neck and chest should be obtained to evaluate for airway narrowing or masses.

This case report describes a benign myoepithelioma of the trachea in a 10 year old girl. Upon performing a literature search, only 11 other cases of myoepithelioma of either the trachea or the respiratory tract have been reported (Table 1). The average age at presentation for the 12 cases reported in the literature, including this case report, was 46.9 (10-77) with a male:female ratio of 1:1.4. Of these 12 cases, 9 were located in the lung and 3 were in the trachea. Karthik et al. described four subtypes of myoepithelioma: plasmacytoid, spindle, epitheliod, and spindle cell.[3] All papers except one reported the predominant cell type of the tumor: five were mixed, four were spindle cell, and two were plasmacytoid.

Jo et al. described a variant of myoepithelioma that has thus far only been reported in cutaneous tissue known as syncytial myoepithelioma. These tumors are characterized by syncytial growth of ovoid, spindled, or histiocytoid cells with minimal to no stroma that stains positively for EMA and S-100.[19] This case presents a tracheal mass staining positive for EMA and had multifocal positivity for S-100 protein with syncytial growth of spindle cells consistent with syncytial myoepithelioma. This is the first reported case of syncytial myoepithelioma of the trachea and also the first reported case of myoepithelioma of the trachea in a child.

Treatment for benign myoepithelioma is surgical resection with negative margins. In this case, endoscopic excision was attempted multiple times with re-growth each time. Thus, the decision to perform tracheal resection with re-anastomosis was made. To date the patient has not had any recurrence. Surgeons that encounter myoepitheliomas of the trachea should consider en bloc resection of the mass with end-to-end anastomosis of the trachea.

Age	Gender	Location	Treatment	Histologic type	Reference
24	M	Anterior mediastinum adherent to parietal pleura	Thoracotomy and excision	Not specified	4
18	F	Endobronchial	VATS and inferior lobectomy	Plasmacytoid, spindle	5
60	F	Peripheral lung	VATS and wedge excision	Spindle, plasmacytoid	6
40	F	Endobronchial	Thoracotomy and lower lobectomy	Spindle	7
54	F	Peripheral lung	Wedge excision	Spindle	8
58	M	Endobronchial	Lobectomy	Spindle, plasmacytoid	9
58	M	Endobronchial	Lobectomy	Plasmacytoid	9
60	M	Peripheral lung	Thoracotomy and upper lobectomy	Spindle	10
37	F	Peripheral lung	Pneumonectomy	Plasmacytoid, spindle	11
67	F	Intrathoracic trachea	Endoscopic snare excision	Spindle	12
77	M	Trachea	Endoscopic snare excision	Plasmacytoid	13
10	F	Trachea	Tracheal resection and anastomosis	Spindle, epitheliod	Current case

Table 1: Literature summary of tracheobronchial myoepitheliomas

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.

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