Primary intratracheal neurilemmoma in a ten-year-old girl: case report and literature review

Lei Wu, MD [a,b*], Micun Sha, BD [b,d*], Xiling Wu, MD [a], Jing Bi, MD [b,c], Min Yang, MD[e], Zhimin Chen, MD, PhD [a,#]

[a] Department of Pulmonology, The Children's Hospital, Zhejiang University School of Medicine, National Clinical Research Center for Child Health, Hangzhou, Zhejiang 310052, China

[b] Department of Endoscopy Center, The Children's Hospital, Zhejiang University School of Medicine, National Clinical Research Center for Child Health, Hangzhou, Zhejiang 310052, China

[c] Department of Otorhinolaryngology head and neck surgery, The Children's Hospital, Zhejiang University School of Medicine, National Clinical Research Center for Child Health, Hangzhou, Zhejiang 310052, China

[d] Department of Pediatrics, Suqian People's Hospital of Nanjing Drum-tower Hospital Group, Suqian, Jiangsu 223800 , China

[e] Department of Pathology, The Children's Hospital, Zhejiang University School of Medicine, National Clinical Research Center for Child Health, Hangzhou, Zhejiang 310052, China

[*] These authors contributed equally to this work.

[#] To whom correspondence should be addressed: Zhimin Chen, MD, PhD, Department of Pediatric Pulmonology, The Children's Hospital, Zhejiang University School of Medicine, National Clinical Research Center for Child Health, 3333 Binsheng Road, Hangzhou, Zhejiang 310052, China; Tel: +86 571 8887 3731; Fax: +86 571 8703

3296; Email: zmchen@zju.edu.cn

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Abstract

Primary intratracheal neurilemmoma is rare in children. Here, we reported a case of tracheal neurilemmoma in a 10-year-old girl. She had recurrent cough, dyspnea, and tachypnea. Magnetic resonance imaging revealed a soft-tissue mass was located on the right lateral posterior wall of the trachea. A multidisciplinary operation was performed. Immunohistochemical analysis demonstrated a benign neurilemmoma. Her clinical symptoms and signs improved. We reviewed the clinical features and surgical procedures of ten children with intratracheal neurilemmoma reported in literature. Radiographic techniques and bronchoscopy are helpful for diagnosis of the disease. Long-time follow-up is required.

Introduction

Tracheal tumors are relatively uncommon in adults, and less frequently observed in children[1]. About two thirds of these tumors are malignant or of intermediate malignancy while the remaining third are benign lesions[2]. Tracheal neurilemmomas, also referred to as a schwannoma, were first described in 1951 by Straus and Guckien, mostly in middle-age adults, but also in children[3]. The patients were present with symptoms of cough and wheezing or stridor (especially positional). Several patients have been mistakenly treated for asthma. Here, we report a case of tracheal neurilemmoma in a 10-year-old girl.

Case presentation

A 10-year-old girl was admitted to our unit because of recurrent cough, dyspnea, and tachypnea for 2 months. Her condition became worse after exercise. There were two fever episodes during the course of the disease. She was hospitalized twice in the local unit and diagnosed with bronchitis and asthma. Physical examination revealed that expiratory wheezing could be heard in both upper segments of the chest. Suprasternal, subclavicular, and intercostal retractions were very obvious. Enhanced magnetic resonance imaging (MRI) revealed a soft-tissue mass was located on the right lateral posterior wall of the trachea at the level of upper part of the thyroid gland (Fig 1A). Pulmonary function studies demonstrated severe inspiratory obstructive ventilation defects, without a significant response to bronchodilators. Electronic laryngoscopy showed a red, smooth and round mass like mushroom with several small, discrete vessels on the surface, the base was located at the 4 to 7 o'clock position of the trachea about 1 cm below the vocal cords. The mass was occupied about 90% of the tracheal lumen (Fig 2A). There was only a small gap between 7 and 10 o'clock for ventilation. A multidisciplinary operation was performed for the patient. The electronic bronchoscope entered the trachea from 7 to 10 o'clock and guided the endotracheal intubation for 4.5mm tube under general anesthesia. Then tracheotomy was performed, and the anesthesia ventilator controlled breathing through tracheotomy cannula. The electrocautery snare easily removed the spherical part of the mass (Fig 2B). Because the base of the tumor is too wide to be removed by the electric

snare, it was ablated using a low-temperature plasma（Smith & Nephew，EIC7070-01）under a laryngoscope (Fig 2C). In order to prevent tracheoesophageal fistula, the base of the new organism was not removed completely (Fig 2D). The ECMO team was present for giving emergency treatment once intubation was difficult or asphyxia. Tracheal intubation was removed after a week. The diameter of the mass is about 1 cm. Hematoxylin and eosin staining showed the tumor was comprised of bundles of spindle cells with elongated palisading nuclei (Fig 3A). Immunohistochemical analysis showed that the tumor was positive for S-100 and negative for smooth muscle actin (SMA), desmin, CD117, CD34 (Fig 3B).

Her clinical symptoms and signs disappeared after the tumor was removed. Tracheotomy cannula was successfully removed one week after operation. The patient returned to the hospital for repeat chest computed tomography (CT) examination and bronchoscopy after six months (Fig 1B). There was slight protrusion at the position of incision (Fig 2E).

Discussion

Schwannomas originate from Schwann cells of the peripheral nerve, and they are benign lesions usually. They were usually reported in the head, neck, retroperitoneum, extremities and lungs[4]. Children tracheal neurilemmomas is rare. We searched the PubMed and Embase databases for articles published until September 30, 2020, using the following search terms: (tracheal OR intratracheal) AND (neurilemmomas OR schwannoma). We only found ten cases of pediatric tracheal neurilemmomas (Table 1). The age ranged from 6.5 to 17. The incidence was higher in girls than boys. The progress is poor for the young children. Both of the two youngest patients died.

Table 1

Case	Reference no	Sex/age	Syptoms	Treatment	Histology	Follow-up	Year
1		F/10 Y	Cough, dyspnea, and tachypnea	Flexible bronchoscopic resection	Benign	1 year, well	2019
2	4	M/17 Y	Recurrent cough for 10 months	Flexible bronchoscopic resection	Benign	2 years, well	2017
3	3	F/9 Y	Cough, wheezing for 3 months	Rigid bronchoscopic resection	Benign	2 months, well	2005
4	1	F/9 Y	Paroxysms of dry coughing, shortness of breath for a month	Partial tracheal resection With primary anastomosis	Benign	4 months, well	2005
5	5	M/17 Y	Airway obstruction symptom	Not recorded	Not recorded	Not recorded	2004
6	6	F/7 Y	Hoarseness mass for 6 years	Intubate and tracheostomy	Benign	Died of local recurrence	2004
7	7	M/16 Y	Hypoxia	Endoscopic bra	Benign	Not recorded	1998 age

			induced by airway obstruction	polypectomy and laser therapy			
8	8	M/16 Y	Cough, wheezing for 11 months	Rigid bronchoscope using CO2 laser	Not recorded	Not recorded	1998
9	9	F/14 Y	S t pneumomediastinum	Intubate and segment t tracheal excised	Benign u	Not recorded s	1993
10	10	F/8 Y	Progressive stridor		Benign	Not recorded	1992
11	11	F/6.5 Y	Dyspnea, cough	Surgical transaction at base	Benign	Died	1964

Generally, neurilemmomas have a slow growth rate. Because the clinical presentation is nonspecific and insidious , diagnosis is often delayed. Until the tumor have attained a large size, the patients will be diagnosed. The main symptoms include cough, wheeze and dyspnea[12]. Hoarseness, hemoptysis, sudden acute respiratory distress, subcutaneous emphysema, and fever rarely occurred.

Due to the non-specific nature of these symptoms, those patients were usually treated for bronchial asthma mistakenly prior to evaluation of the airways. Our patient was treated for asthma at first. Pulmonary function is an effective method to distinguish tracheal neurilemmomas from asthma[8,13]. Pulmonary function of tracheal neurilemmomas usually revealed that a fixed upper airway obstruction with no response to bronchodilators. CT of chest or MRI is also helpful in assessing the location, size, and degree of intra and extratracheal extensions of the tumor.

Bronchoscopy is very useful in the diagnosis, biopsy and resection of the tumor. Microscopically, the tumor cells are long spindle. The nature of tumor is diagnosed by histological analysis of the biopsy. Usually, a neurilemmoma stains positive for S-100 protein, vimentin, and CD56, while negative for other markers including cytokeratin AE1/AE3, desmin, smooth muscle myosin (SMM), SMA, CD34, and CD117.

The treatment for primary tracheal schwannoma should consider the size, location, the depth of tracheal wall invasion and extratracheal involvement[14]. Surgical resection and endoscopic excision are utilized in the treatment of tracheal schwannoma mainly[13]. Traditionally, rigid bronchoscopy is probably the best conduit for managing endotracheal tumors[15]. In our case, since the tumor was in the upper trachea and occupied most of the airway, we chose electronic bronchoscope under laryngeal mask airway. We used low-temperature ion probe to remove the base of the tumor and reduce the probability of recurrence after electrocautery snare. ECMO team was deployed during the operation.

After endoscopic resection recurrence of the tumor is common and has been previously reported. Long-time follow-up is required to evaluate outcomes and complications, such as recurrence and airway stenosis.

Conclusion

Children intratracheal neurilemmoma is extremely rare. The main symptoms include cough, wheeze and dyspnea. CT of chest is helpful in assessing the location, size, and degree of intra and extratracheal extensions of the tumor. Surgical resection and endoscopic excision are utilized in the treatment of tracheal schwannoma mainly. Long-time follow-up is required.

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Figure legend

Fig 1 Result of preoperative and postoperative imaging. (A) Enhanced magnetic resonance imaging (MRI) reveals a soft-tissue mass occates on the right lateral posterior wall of the trachea. (B) CT shows there is slight protrusion at the position of incision .

Fig 2 Fiberoptic bronchoscopy findings. (A) A mass was located at the 4 to 7 o'clock position of the trachea. (B) The spherical part of the mass was removed by the electrocautery snare. (C) The base was ablated by a low-temperature ion probe.(D) Most of the base was removed. (E) There was slight protrusion at the position of incision.

Fig 3 Pathological pictures of tumor cells. (A) Histologic section from tumor shows that tumor cells are spindle with elongated palisading nuclei (hematoxylin-eosin stain, ×400). (B) Immunohistochemical stain with S-100 protein antibody reveals these tumor cells are diffuse and positive (Streptaridin Peroxidase Method, ×200).