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Case Report

Tracheal paraganglioma presenting as stridor in a pediatric patient, case report and literature review

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ABSTRACT

Objective: To review tracheal paragangliomas and describe the clinical presentation, radiologic findings, operative management, and histologic findings of a pediatric patient who presented with stridor refractory to traditional asthma therapy.

Methods: Chart review of an 8-year-old male who presented to a tertiary care pediatric hospital and literature review of tracheal paragangliomas.

Results: We present the case of an 8-year-old male who presented with new-onset of wheezing and dyspnea on exertion. He was given a new diagnosis of asthma and treated with bronchodilators that failed to improve his symptoms, which progressed over 3 months until he presented urgently with biphasic stridor. Bedside flexible laryngoscopy failed to reveal an etiology. Computed tomography (CT) imaging demonstrated 17 × 12 × 16 mm exophytic mass arising from the posterior membranous trachea with extension of the mass to the border of the thyroid gland and separate from the esophagus. Magnetic resonance imaging (MRI) angiography confirmed vascular supply from the right thyrocervical trunk and inferior thyroid artery. Rigid microlaryngoscopy revealed a friable vascular polypoid mass 2 cm distal to the vocal folds with 75% obstruction of the airway from which a small biopsy was taken. Pathology confirmed paraganglioma with neuroendocrine cells arranged in "zellballen" architecture and strong immunopositivity for chromogranin and synaptophysin in the neuroendocrine cells and S100 immunopositivity in the sustentacular cells. The patient underwent complete open resection of the tumor including three tracheal rings with primary anastomosis. Final pathology confirmed paraganglioma and negative margins. Genetic screening revealed a succinate dehydrogenase complex subunit C (*SDHC*) germline mutation, confirming hereditary paraganglioma/pheochromocytoma syndrome. He remains well at 3 month follow up without dyspnea or stridor.

Conclusion: Tracheal paragangliomas are exceptionally rare, with 12 reported cases. This is the only pediatric case reported. In pediatric patients with persistent airway complaints, subglottic and tracheal masses and obstruction should be considered. Due to the vascularity and endotracheal component of tracheal paragangliomas, a detailed surgical plan should consider embolization, endotracheal laser photocoagulation and electrocautery, and open surgical resection. Additionally, pediatric patients benefit from a multidisciplinary approach including radiology, endocrinology, and genetic counseling.

Introduction

Paragangliomas arise from neural crest elements in the parasympathetic nervous system [1]. In the adrenal system, these lesions are referred to as pheochromocytomas. In the head and neck, these lesions are called paragangliomas, and primarily arise from the carotid body, jugulo-tympanic region, and vagus nerve [2]. Other head and neck locations are infrequently described and paragangliomas of the trachea are exceptionally rare, with 12 reported cases [3,4]. Presenting symptoms of tracheal paragangliomas include hemoptysis, dyspnea, and cough. Full evaluations of patients with paragangliomas include whole body imaging with computed tomography (CT) and/or magnetic resonance imaging (MRI) with possible vascular evaluation and magnetic resonance angiography (MRA) of the site of the lesion. Other studies typically include biochemical testing for catecholamines. The role for genetic evaluation is increasing due to recent developments in the understanding of familial paragangliomas and the presence of inheritable germline mutations. The treatment of tracheal

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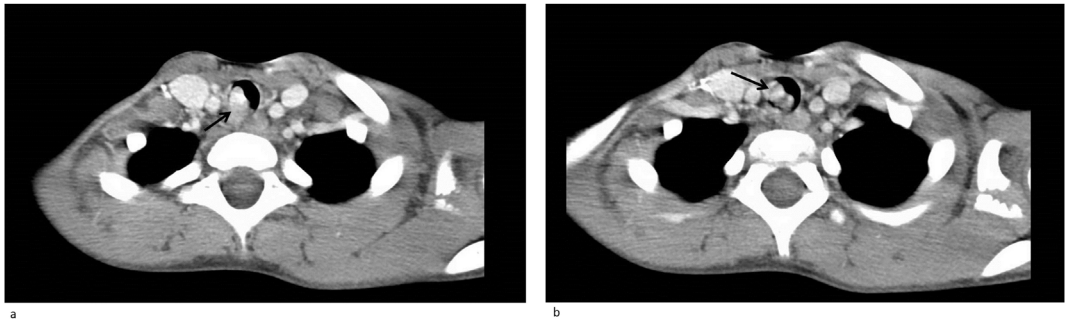

Fig. 1. Axial CT contrast neck indicating tumor arising from membranous trachea with intraluminal exophytic and extratracheal components.

paragangliomas most commonly includes open surgical excision, endotracheal excision and cautery with consideration of preoperative embolization. Radiation therapy and stereotactic radiosurgery have been used in other head and neck paragangliomas but have not been reported in tracheal paragangliomas [2]. We report the first pediatric case of a tracheal paraganglioma.

Case report

An otherwise healthy 8-year-old male presented to his primary care physician with new-onset wheezing and dyspnea on exertion. He was given a new diagnosis of asthma and treated with bronchodilators that failed to improve his symptoms, which progressed over 3 months until he presented urgently to otolaryngology with biphasic stridor. Bedside flexible laryngoscopy noted normal vocal cord mobility and no etiology for the patient's stridor. Computed tomography (CT) with contrast imaging demonstrated a 17 × 12 × 16 mm exophytic mass arising from the posterior membranous trachea in the right tracheoesophageal groove nestled posteroinferior to the right thyroid lobe (Fig. 1a–b). The mass appeared separate from the esophagus. Operative rigid microlaryngoscopy revealed a friable vascular polypoid mass 2 cm distal to the vocal folds with 75% obstruction of the airway (Fig. 2). A small biopsy was taken with significant bleeding controlled with topical epinephrine and pressure. The patient was left intubated for airway protection pending final pathology. Due to the vascularity of the mass, magnetic resonance imaging (MRI) angiography was obtained to identify the vascular supply which arose from the right thyrocervical trunk and inferior thyroid artery (Fig. 3a–b). Pathology confirmed paraganglioma with nests of tumor cells with typical neuroendocrine features and intervening vascular sinusoids between nests, consistent with a "zellballen" architectural pattern (Fig. 4). Immunohistochemistry was strongly positive for chromogranin and synaptophysin within the tumor nests with S100 highlighting surrounding sustentacular cells (Fig. 5a–c). Pancytokeratin was negative, ruling out a carcinoid tumor (Fig. 5d). All findings were characteristic for paraganglioma. Subsequent evaluation included normal chest and abdominal CT imaging and normal catecholamines and metanephrines.

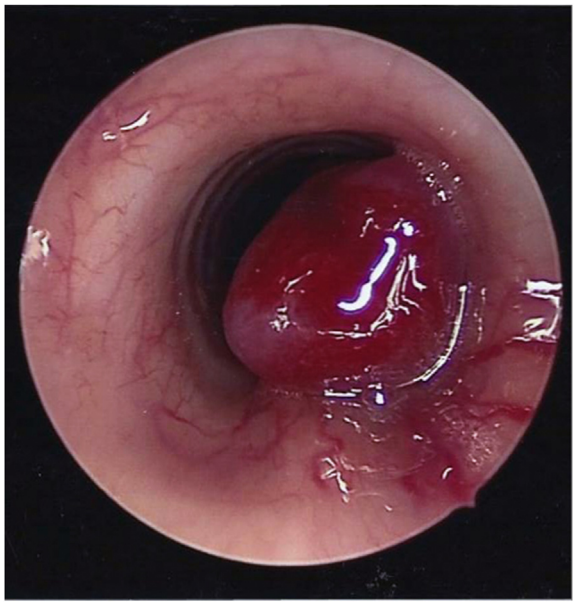

Fig. 2. Intraoperative telescopic examination of the tumor with exophytic, friable, vascular intraluminal component.

The patient underwent open resection of the mass through a transverse cervical incision. The right thyroid lobe was elevated and right recurrent laryngeal nerve was identified. At this point, there was a 2 cm friable polypoid mass arising from the right membranous trachea with external and internal extension (Fig. 6). The recurrent laryngeal nerve and esophagus were noted to be uninvolved by tumor and the inferior thyroid artery was suture ligated (Fig. 7). Complete en bloc excision of the mass included tracheal resection of three tracheal rings and primary anastomosis with negative margins on frozen and permanent pathology (Fig. 8). Final pathology confirmed paraganglioma with features identical to the previous biopsy. He was discharged home on postoperative day 10. Genetic analysis revealed a succinate dehydrogenase complex subunit C (*SDHC*) FrenchCanadian founder mutation (c.397C > T, p.R133*) and the patient was diagnosed with hereditary paraganglioma/pheochromocytoma syndrome. CT imaging of the whole body failed to reveal any other tumors. He was doing well at three months postoperatively without evidence of residual suture or stenosis on rigid bronchoscopy (Fig. 9).

Discussion

Paragangliomas of the carotid body, jugulotympanic region, and vagus nerve typically present with symptoms based on their anatomic location: an asymptomatic neck mass, pulsatile tinnitus, or dysphonia. These tumors are confirmed with anatomic imaging to evaluate size of tumor extension and functional imaging with angiography to determine vascular supply. Head and neck paragangliomas are typically managed with a multidisciplinary approach for preoperative embolization and resection using advanced microsurgical techniques [2]. The goals of surgery are complete excision, preservation of neurovascular anatomy, and acceptable soft tissue reconstruction [5]. Histopathology can confirm the diagnosis due to organized nests of chief cells surrounded by sustentacular cells in a "zellballen" pattern. Immunohistochemistry demonstrates positivity with neuroendocrine markers such as synaptophysin, chromogranin, neuron specific enolase (NSE) and CD56 within the chief cells while S100 will highlight the surrounding sustentacular cells. Cytokeratins are negative, helping to differentiate paragangliomas from carcinoid tumors. External beam radiation may be considered in unresectable tumors, recurrences, or patients with elevated perioperative risk.