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Ewing Sarcoma in the cervical trachea of a pediatric patient: A case report and review of the literature

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ABSTRACT

Malignant pediatric tracheal tumors are rare and often present with non-specific findings. Here we present the first case of a primary Ewing tracheal sarcoma in a pediatric patient. Initial presentation, surgical management, post-operative care, and a review of the literature are discussed.

1. Introduction

A 2 year old male with no significant past medical history was transferred to Cincinnati Children's Hospital to further evaluate a partially obstructing tracheal mass. Approximately six weeks prior to presentation, he developed asthma-like symptoms. Two weeks prior to presentation, he developed a non-productive cough, stridor, and increase work of breathing. His pediatrician diagnosed and treated him for croup. The following day, his breathing continued to worsen. The family sought treatment at an urgent care, and subsequently referred him to the local pediatric hospital. During the initial work-up, a lateral x-ray (Fig. 1) was performed that was suspicious for a foreign body. He underwent a microlaryngoscopy and bronchoscopy (MLB) that revealed a proximal tracheal mass isolated on the posterior wall. He was intubated with a 4.0 cuffed endotracheal tube distal to the lesion and transferred to Cincinnati Children's Hospital for further management. He subsequently underwent a repeat MLB which confirmed a posterior tracheal mass and normal distal anatomy, Fig. 2. A superficial biopsy revealed a high-grade small round cell sarcoma, not otherwise specified. A chest CT and MRI revealed an exophytic mass along the posterior wall of the trachea spanning C7/T1 without extension beyond the tracheal wall, Fig. 3. No evidence of distant metastases were identified by chest CT or PET/CT and bilateral bone marrow biopsy/aspirate was negative for malignant cells. He underwent primary resection with an oblique anastomosis (i.e. slide-tracheoplasty closure) achieving a complete resection with negative margins, Fig. 4. Tracheal resection showed a 1.0 × 1.0 cm circumscribed red-brown nodular lesion in the posterior wall which on histology demonstrated an infiltrating neoplasm in between tracheal cartilage composed of sheets of small blue cells with indistinct cell borders, minimal eosinophilic cytoplasm, and large nuclei with stipple chromatin. The tumors cells show diffuse membranous immunoreactivity to CD99, patchy CD56 cytoplasmic positivity, and immunonegative for PAX5, Fig. 5. Genomic profiling via Foundation One showed an EWSR1-ERG fusion. The histology and immunohistochemical and genomic profile are consistent with Ewing Sarcoma.

He was subsequently extubated on POD 4 to 12 L high flow nasal cannula and gradually weaned to room air. Serial MLBs were performed to assess healing (Fig. 6). At 4 weeks from surgical resection, the patient received adjuvant chemotherapy as per the now closed Children's Oncology Group study, AEWS0031. Fourteen cycles of dose dense every-other week chemotherapy was received (including standard dose vincristine, doxorubicin, cyclophosphamide, ifosfamide and etoposide with GCSF support) and tolerated with no excess or unexpected toxicity. Given his negative margins, no radiotherapy was required. His off therapy evaluations confirm continued remission to date.

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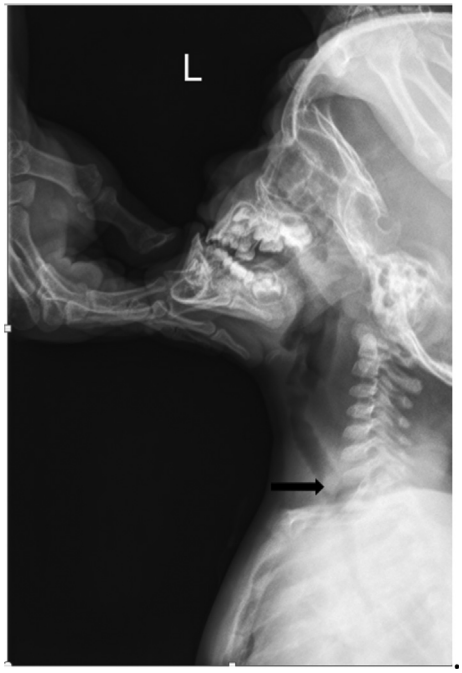

Fig. 1. Black arrow indicating tracheal mass on lateral x-ray.

2. Discussion

Primary tracheal tumors are very rare representing less 0.1% of tumors [1], Tracheal tumors in adults carry an 80–90% risk of malignancy whereas tracheal tumors in children are benign in 60–70% of cases [2]. Overall, the most common malignant tumors found in trachea include squamous cell carcinoma and adenoid cystic carcinomas [3]. Other, less common, malignant tracheal tumors include mucoepidermoid, carcinoid tumors, small cell carcinoma, adenocarcinoma, lymphoma, chondrosarcoma, large cell carcinoma, rhabdomyosarcoma, and leiomyosarcoma [1,3,4]. Primary tracheal tumors are more commonly found on the posterior tracheal wall [5].

To our knowledge, there are only a two case reports of a primary tracheal Ewing Sarcoma in the literature. In 2010, Elmi et al. described a 28 year-old-male who initially presented with stridor, recurrent cough, and progressive exertional dyspnea. A flexible bronchoscopy revealed an 80–90% obstructing mass of his distal trachea that was debulked and stented with a 14 mm silicon "Y" stent. He was treated with vincristine, doxorubicin, and cyclophosphamide, alternating with ifosfamide and etoposide every 2–3 weeks. After 5 cycles of chemotherapy, the tracheal mass regressed to 2 cm proximal to the carina and underwent a carinal sparing tracheal resection with post-operative radiation (50.4 Gy). He has remained disease free 7 months after finishing treatment [6]. The second case report published by Guzeloz et al., in 2014, described a 70 year-old-male who developed a lateral wall tracheal mass that underwent endobronchial excision with a laser with post-operative 50.4-Gy radiation therapy. Unfortunately metastatic disease was found 3 months after completing radiation therapy and ultimately died 8 months post-operatively from renal failure [7]. Surgical resection was performed in all reported cases.

Slide tracheoplasty has become the procedure of choice at Cincinnati Children's Hospital Medical Center for benign and malignant tracheal lesions. In the pediatric population, the upper two-thirds of the trachea can usually be accessed via a *trans*-cervical approach without the need of extracorporeal membrane oxygenation or cardio-pulmonary bypass. A planning micro-laryngoscopy and bronchoscopy is performed to assess accessibility as the light of the telescope *trans*-illuminates the anterior neck. Once the trachea is exposed and mobilized, the location of the proximal and distal incision is determined by a 27-guage needle while a rigid bronchoscopy is being performed. This allows precise control of the margins while the tracheal lesion is visualized within the trachea. Two tracheal retraction sutures are placed below the level of the lesion in the distal trachea. Once the tracheal incision is made, the oral endotracheal tube is converted to *trans*-cervical. The endotracheal is removed intermittently while the tracheal mass is removed en-bloc. For malignant lesions, additional margins are acquired for frozen pathological analysis. The proximal and distal incisions are made at a slight bevel. The anterior aspect of the proximal trachea is divided in the midline for several millimeters. The posterior aspect of the distal trachea is similarly divided in the midline for several millimeters. As the two segments are "slid" together, the overall lumen is expanded compared to a primary anastomosis thereby reducing the clinical impact of post-operative scarring [8,9].

The annual incidence of Ewing sarcoma is 1–3 cases per 1 million people per year [10]. Most patients are between 10 and 20 years at time of first diagnosis, although cases have been reported from birth to 80 years. Caucasians are nine times more likely to develop Ewing sarcoma than African Americans, and males are at a slightly higher risk than females [11,12]. The Ewing sarcoma cell of origin has recently been determined to represent either a mesenchymal stem cell or possibly a neural crest derived stem cell. Ewing sarcoma pathogenesis is driven by a pathognomonic chimeric transcription factor oncogene as a result of a somatic reciprocal

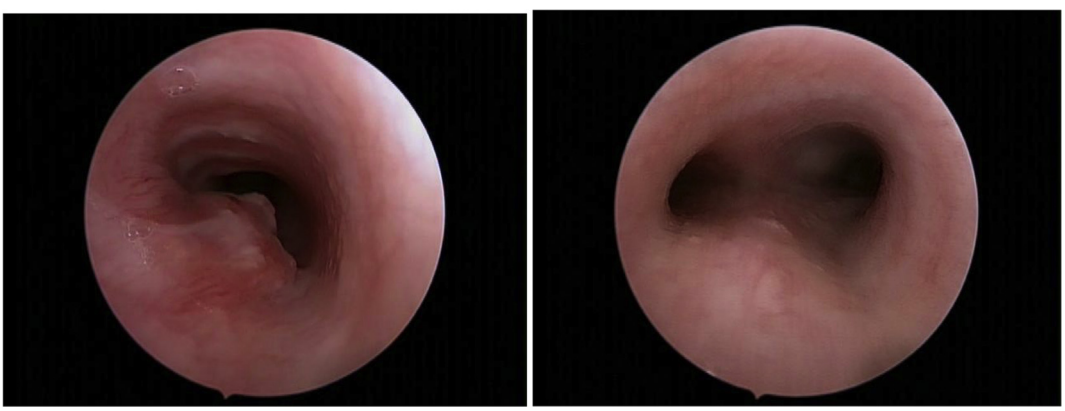

Fig. 2. Rigid bronchoscopy of the tracheal lesion and normal apperaing distal trachea and bronchi.