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Primary tracheobronchial tumors in children

Patricio Varela, MD[a,*], Luca Pio, MD[b,c], Michele Torre, MD[c]

[a] Department of Pediatric Surgery, Clinica Las Condes Medical Center, Hospital de Niños Calvo Mackenna, University of Chile, Camino el tucuquere 4080, Santiago, Chile
[b] DINOGMI, University of Genoa, Genova, Italy
[c] Airway Team and Pediatric Surgery Unit, Istituto Giannina Gaslini, Genova, Italy

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ABSTRACT

Primary tracheobronchial tumors are rare lesions that can be benign or malignant, with different location along the airway tree. Symptoms may include wheezing, chronic pneumonia, asthma, chest pain, recurrent cough, atelectasis, haemoptysis, and weight loss. Due to the heterogeneity of symptoms, diagnosis can be difficult and the airway involvement can lead progressively to a bronchial or tracheal obstruction. Due to the rarity of primary tracheobronchial tumors in children, there are not any oncological guidelines on pre-operative work-up, treatment, and follow-up. Only few reports and multicentric studies are reported. In most cases, surgical resection seems to be the treatment of choice. Brachytherapy, endoscopic treatment, and chemotherapy are rarely described. In this article we present an overview on these rare tumors, including pathological aspects, clinical presentation, imaging assessment, and endoscopic or open surgical treatments. We discuss different surgical approaches, according with tumor location.

Introduction

The most common airway tumor in children is papillomatosis, a well known viral infection disease, described in many reports, especially from otorynolaryngologist groups, both in pediatric and adult age. Another relatively frequent and well known airway tumor is infantile hemangioma. Conversely, the group of the other primary airway tumors in children includes very rare diseases whose treatment often requires a multidisciplinary approach and major surgery. Over the years, some clinical case series were published[1–4] in which the patients were treated heterogeneously (by endoscopic or surgical approach). In other few larger studies published,[5–7] details on type of surgical procedure, exact tumor location, and other data were lacking.[5–7] Primary airway tumors have a wide range of histopathologic types and their long-term outcomes remain still unknown (Figure 1).

Non-specific symptoms as haemoptysis, chronic pneumonia, asthma, chest pain, wheezing, recurrent cough, and weight loss can delay the diagnosis and children may develop tracheal or bronchial obstruction.

Neville et al.[8] reported an overall incidence of 0.049 per 100.000 children, but the exact incidence of every single primary tracheobronchial tumor (PTT) type in pediatric population is not provided, due to the rarity of the singular entities. Epidemiology and End Results (SEER) registry, due to the rarity of the PTT, justified the absence of prospective studies comparing different treatments as well as the absence of guidelines for diagnosis and treatment.[8]

The aim of our article is to present an overview of rare pediatric airway tumors (excluding metastasis, papillomatosis, and vascular tumors), including pathological aspects, clinical presentation, imaging assessment, and endoscopic or open surgical treatment.

Pathology

Tumor histology is the main factor determining the survival rate as reported by Rojas et al.,[7] with a better survival reported for carcinoids and mucoepidermoid carcinoma. The better prognosis of these tumors was also related to the intraoperative findings of negative lymph nodes and survival is higher when lymphadenectomy and a lesser extensive surgery were performed.[7]

Carcinoid tumors (CaT) are the most frequent PTT in children with an overall incidence of 3–5 cases per million people per year[8] and represent among the 80% of PTT.[9,10] CaT have an endodermal origin from Kulschitzky cells, generally located in the basal layer of bronchial epithelium. CaT are classified as typical (10%) and atypical (90%), according to mitotic index and presence or absence of necrosis.[11]

* Corresponding author.
E-mail address: pvarelachile@yahoo.com (P. Varela).

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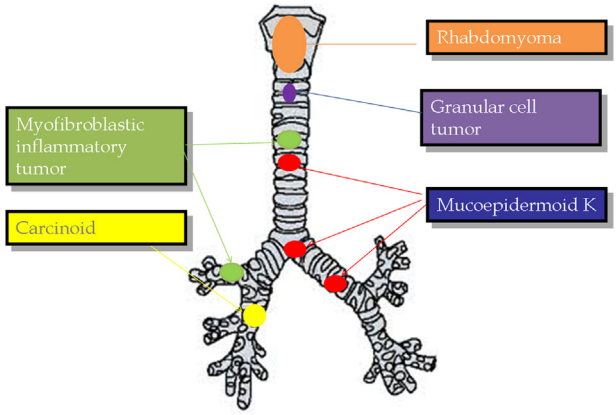

Fig. 1. The most representive primary tracheobronchial tumors are described with their more frequent location.

In association with airway symptoms, clinical presentation of CaT may include carcinoid syndrome (hypotension, diarrhea, and vasomotor flushing) in 10–30% of cases.[12]

As CaT can be characterized by lymph nodes metastases, lymph node sampling is recommended[13] during surgery. A complete surgical resection remains the treatment of choice—despite CaT can present as intraluminal or pedunculated, in case of a bronchial wall infiltration an endoscopic approach does not allow a complete resection.[14] The overall survival of CaT is good (95%) in the largest reported series of PTT.[7]

Mucoepidermoid carcinomas (MC) are the second commonest PTT in children. They take origin from salivary gland and are characterized by mucinous, intermediate, and squamous cells.[15] MC represent 10% of PTT and are classified in low, intermediate, and high grade, based on the mitotic index.[16] MC usually present as an airway (trachea or main bronchus) slow-growing vascular polypoid mass. Despite some endoscopic resections were attempted,[17] the possibility of recurrence, as reported by Romão et al.,[2] confirms that open surgical resection and lymph node sampling remain the treatment of choice.[18]

Similar to CaT, the overall survival for MC is excellent (87–100%).[9]

Granular cell tumors (GCT) take origin from Schwann cells and are frequently located in tracheobronchial lumen, only the 10% occurring in the larynx.[3,19] Whereas malignancy transformation was never documented in pediatric population, surgical resection must be considered, based on the morbidity risk.

Inflammatory myofibroblastic tumors (IMT), also named pseudotumors, are characterized by myofibroblastic spindle-shaped cells, lymphocytes, plasma cells, and eosinophils. They represent around 1% of PTT,[20] and are commonly located in upper trachea (Figure 2). Despite being considered as low-grade benign-tumor, IMT may give occasionally metastatic spread.[21] Corticosteroid treatment could favor proliferation of the tumor.[22] After the recent identification of ALK gene mutations in myofibroblasts on 50% of IMT patients, the utilization of ALK inhibitor as crizotinib was introduced in selected cases as complementary treatment and founded to be useful in preliminary studies.[23–25] Compete surgical resection actually remains the treatment of choice and endoscopic treatment must be reserved to those cases at high risk of surgical morbidity.[20,21]

Rhabdomyosarcoma (RMS) is a rare entity in children. In pediatric patients, RMS can be located in the larynx as an embrional botryoid variant. A previous radiation therapy for a pre-existing papillomatosis is the main recognized risk factors for larynx RMS, but there are also implications between genetic predisposition and immunological factors.[26]

Mutilating surgery is not indicated, as multiagent chemotherapy associated with radiotherapy allows an excellent overall survival even in cases of lymph nodes metastasis with survival of all cases after a follow-up between 13 and 17 years in the largest reported series.[27,28] Surgery must be reserved to the purpose of diagnostic biopsy or debulking for emergency condition.

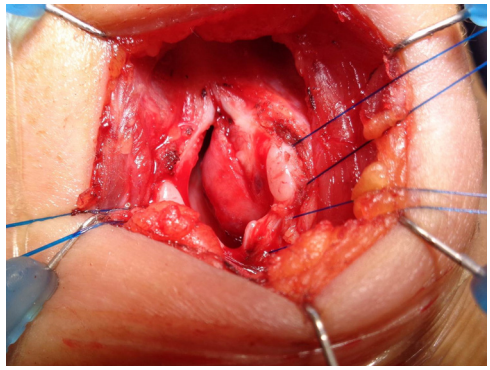

Fig. 2. Miofibroblastyc tumor: upper airway localization.

Other very rare tumors are the laryngotracheal chondromas (LTC). Surgical resection remains the treatment of choice and the rate of local recurrence seems to be low but still unclear, due to the slow growth of this PTT and the relatively short follow-up of reported cases in literature.[29]

In the literature, there is only 1 case of tracheal lipoblastoma (TL) in children, described by our group[30]—we described a posterior tracheal wall tumor with obstructive symptoms at presentation and a rapid growth with esophageal involvement (Figure 3). A partial cricoid tracheal resection was required, as well as a strict follow-up, due to the high local relapse risk in the early postoperative time.

Sign and symptoms

Clinical presentation depends on the site and type of the tumor. Obstructive symptoms are frequent in case of severe (> 50% of the lumen) upper airway obstruction with stridor, wheezing, and dyspnea; cough is very common and is expression of mucosal irritation or poor clearance with accumulation of airway secretions distal to the stenosis; haemoptysis is not a frequent symptom and is observed in case of mucosal ulceration. Commonly, clinical presentations of PTT are misdiagnosed as bronchitis, pneumonia, or asthma episodes. Complete or partial lung atelectasis is another frequent occurrence in these patients with bronchial localization. Neuroendocrine tumors as CaT can present with carcinoid syndrome as described above.

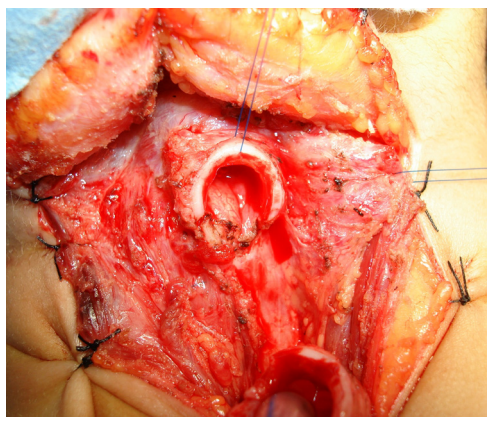

Fig. 3. Tracheal lipoblastoma. A partial tracheal cricoid resection was neccesary.