Primary mucoepidermoid carcinoma of the trachea in a child

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Abstract

Mucoepidermoid carcinoma of the trachea is a rare tumour, especially in the paediatric population. We report the case of a 9-year-old boy with mucoepidermoid carcinoma of the trachea that was preoperatively diagnosed as an intraluminal polypoid mass arising from the trachea and extending into the right main bronchus. A complete resection of the tumour with reconstruction and end-to-end anastomosis of the trachea was performed. The patient is now, 24 months after surgery, free of disease.

Keywords: Trachea • Mucoepidermoid carcinoma • Child • Tracheal resection • End-to-end anastomosis • Reconstruction

INTRODUCTION

Primary malignant tracheal neoplasms in children are rare. The presenting symptoms are generally dyspnoea, coughing, wheezing, haemoptysis or obstructive pneumonia. There have only been a few cases, predominantly of boys ranging in age from 4 to 9 years, described in the literature [1–5]. We present a 9-year-old previously healthy child with a tracheal tumour in whom the symptoms were considered to be caused by asthma. The child underwent emergency preoperative and intraoperative diagnoses of the tumour and successful resection of two tracheal rings together with the tumour followed by end-to-end anastomosis.

CASE REPORT

A 9-year-old boy was referred to our hospital from the Gaza Strip in the Palestinian Authority. Six months prior to admission, the child was diagnosed with asthma because he had dyspnoea and was coughing, and treatment with bronchodilators was started without any improvement in his respiratory status. One week prior to admission to our hospital, the patient was hospitalized in the local hospital with progressing dyspnoea and was intubated for 24 h with the existence of a tracheal tumour not being known. After successful extubation, the child was still dyspnoic, and due to suspected heart disease, was transferred to our centre for further evaluation.

On admission to the emergency room the child was dyspnoic, with 36 breaths a minute, and diaphoretic. His head was sunken into his shoulders as in the case of heavy-smoking chronic obstructive pulmonary disease patients. A chest X-ray did not reveal any abnormality, and cardiac problems were excluded by echocardiography. A computed tomography (CT) scan of the chest and upper abdomen was then carried out, which surprisingly revealed a tracheal orifice completely blocked by a tumour that was above the level of the carina and also extended into the right main bronchial orifice (Fig. 1). CT three-dimensional reconstruction of the trachea revealed a large broad-based tracheal tumour originating from the lower tracheal rings.

The patient was immediately transferred from the emergency room to the operating theatre where a flexible bronchoscopy, carried out under general anaesthesia, revealed a large red sessile tumour in the lower trachea, completely blocking the tracheal orifice. The option of endoscopic resection or biopsy of the tumour prior to surgery was discarded due to the immobility of the tumour and difficulty ventilating the child. Due to the existence of immediate cardiopulmonary bypass facilities at our institution, we decided that the optimal surgical approach was via a sternotomy rather than via a right thoracotomy. Following a sternotomy, the anterior and posterior pericardia were opened, the trachea with bifurcation mobilized and, under direct vision of the bronchoscope, the tumour was excised en-bloc with two rings of the trachea. Intraoperative frozen examination revealed a mucoepidermoid tumour of the trachea with tumour-free surgical margins. End-to-end anastomosis was carried out using absorbable 4.0 polydioxanone sutures. Continuous sutures were made on the posterior membrane and interrupted sutures on the cartilaginous part of the trachea. The mediastinum was drained by two 24 French chest tubes, the chest was closed and the child transferred to the paediatric intensive care unit.
Mucoepidermoid tumours represent ≏ 1% to 5% of bronchial adenomas, including also adenoid cystic carcinomas (cylindromas), mucous gland adenomas and carcinoids. The term “bronchial adenomas” is not correct because it refers to a benign process, and experience has demonstrated that these tumours have a broad spectrum of biologic activity and can sometimes have malignant behaviour. The term “bronchial adenomas” has therefore been recently replaced by “bronchial gland tumours”. These slow-growing tumours usually manifest with irritation and obstruction of the tracheobronchial tree and symptoms are related to their site [3]. When these masses are not resolved, a more scrupulous workup is usually performed and intratracheal tumours diagnosed. The most common malignant tracheal tumours are squamous cell and adenoid cystic carcinomas. Bronchoscopy with biopsy remains the main diagnostic modality in non-emergency cases.

The clinical course of these tumours correlates with the histological grade of the tumour. Low-grade tumours generally grow locally and are amenable to complete surgical resection with normal life expectancy, whereas patients with high-grade lesions have been reported to have a poor prognosis. Complete resection of the tumour en-bloc with the tracheal rings combined with reconstruction of the trachea is the mainstay of treatment [1–5]. The role of oncological treatment is unclear because of the small number of patients reported in this age group, who are usually treated successfully by surgery [1–5]. There are no reports in the literature of palliative treatment for these tumours in children.

We have discussed the case of a tracheal mucoepidermoid carcinoma in a 9-year old boy that was successfully treated with tracheal resection and reconstruction. At present, 24 months after surgery, the child is in good general health and free of disease.

Conflict of interest: none declared.

REFERENCES


eComment. Mucoepidermoid carcinoma: common findings and surgical treatment

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We read with interest the paper by Papiashvili and co-authors, reporting on an interesting case of mucoepidermoid carcinoma of the lower trachea in a 9-year-old boy [1].

Mucoepidermoid carcinomas are rare neoplasms [2] of the bronchial tree arising from mucous and serous glands of the epithelium. They are rare and represent about 1% to 5% of bronchial adenomas, including also adenoid cystic carcinomas (cylindromas), mucous gland adenomas and carcinoids. The term “bronchial adenomas” is not correct because it refers to a benign process, and experience has demonstrated that these tumours have a broad spectrum of biologic activity and can sometimes have malignant behaviour. The term “bronchial adenomas” has therefore been recently replaced by “bronchial gland tumours”. These slow-growing tumours usually manifest with irritation and obstruction of the tracheobronchial tree and symptoms are related to their site [3]. When these masses are located in the trachea, as with the case presented by Papiashvili et al, they produce upper respiratory tract occlusion with cough, wheezing and dyspnea.

In this young patient, respiratory distress was interpreted as asthma until a CT scan showed the tracheal obstruction. Tracheal obstruction must always be suspected in intractable asthma, also in young patients. In cases of localization in the proximal or segmental bronchi, symptoms are usually related to progressive occlusion of a bronchus and therefore are characterized by relapsing pneumonia. Complete surgical resection is the treatment of choice for mucoepidermoid carcinoma. Endoscopic resection has been attempted, but the incidence of local relapse is high. It has a role only as a temporary option in order to plan and facilitate surgery. The principles of surgery are a clear margin at frozen section, complete locoregional lymph nodes removal and maximum sparing of lung parenchyma with tracheobronchial resection and reconstruction techniques.

In the case reported by Papiashvili and co-workers they performed an excellent and successful operation through a sternotomy, which is an unusual access to the lower trachea. As reported by the authors, the choice was based on “difficulty ventilating the child” and “the existence of immediate cardiopulmonary by-pass facilities”. Being general thoracic surgeons, we would have made the same operation through a right thoracotomy, and would have been ready to use a partial by-pass with peripheral cannulation, if required. We appreciate the report of such an unusual case successfully treated in a young boy.