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Intra bronchial mucoepidermoid carcinoma in an 8 year old girl: A case report of rare tumor with review of literature

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ABSTRACT

Intra bronchial and intra tracheal tumors are very rare in pediatric age group. Most of the literature clubs primary pulmonary tumors with these; hence actual incidences are not well defined. Amongst this mucoepidermoid carcinoma (MEC), whether low or high-grade account for only 0.1–0.5% of lung carcinomas. We report such a rare case of an eight year old girl presenting with history of recurrent lower respiratory tract infections with respiratory distress. CT scan of chest was suggestive of a mediastinal mass. She underwent rigid bronchoscopic biopsy, histopathology showed it to be mucus gland adenoma. She underwent right thoracotomy with complete excision of mass along with right lower lobectomy. Histopathology reported it to be a low-grade MEC with clear margins. She did not require any chemotherapy or radiotherapy and has no recurrence at 1-year follow up.

Primary lung tumors whether intra parenchymal or intra bronchial are rare in childhood and are often overlooked owing to its nonspecific presentation. Different nomenclature and origin of the tumor over the years have brought further confusion to the pathologists' interpretation of the lesion. Amongst the intra bronchial tumors, MEC is reported in only 55 cases in children as documented in literature [1,2]. Although MEC of the salivary gland is relatively common, MEC arising from the mucous gland of the bronchus is rare. Surgical resection is the primary treatment for low-grade MEC with excellent outcome without any need of chemotherapy or radiotherapy as observed in case reported herewith.

1. Case report

An eight year old girl was being treated for recurrent lower respiratory tract infections and breathlessness off and on for 3 months. She was investigated with a plain radiograph of chest, which showed collapse of right lower lobe with shift of trachea and heart shadow to right side (Fig. 1). CT scan of chest with contrast revealed presence of a mediastinal mass 6 × 4 × 3 cm in size sitting on right bronchus, compressing its lumen and leading to collapse of right lower lobe (Fig. 2).

A diagnostic bronchoscopy showed almost complete occlusion of right lower lobe bronchus with intraluminal mass. A needle biopsy was performed and it was sent for histopathological examination. However, there was severe bleeding immediately after the first bite and the procedure was terminated with endotracheal intubation with gentle sucking of the blood and child was transferred to critical care unit on ventilatory support. The oozing stopped in few hours and the patient was extubated on next day and discharged on post operative day 4. The biopsy was reported to be a mucous gland adenoma without atypical cell and immunohistochemistry was negative.

The parents were explained need of surgery along with the possibility of pneumonectomy. Right posterolateral thoracotomy was performed by selective intubation of left main bronchus. The mass was dissected all around carefully. Adhesions were divided all around and distally away from the helium. Initially the bronchii of the lower lobe were incised just distal to the mass and then the right bronchus was opened just proximal to mass and as soon as incision was taken on the bronchus the mass protruded though the lumen and it was excised (Fig. 3). It was not possible to anastomosis proximal bronchus with distal bronchii of the right lower lobe and

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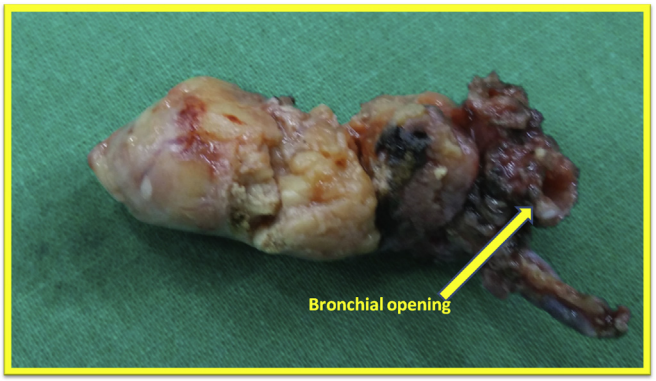

Fig. 1. Preoperative CT scan image, with mass marked with arrows obstructing the lumen.

decision was taken to repair proximal bronchus with right lower lobectomy, which was performed with uneventful post operative course. Intercostal drainage tube was removed on post operative day 2 and patient was discharged on day 3. Histopathology of the excised specimen showed it to be a low-grade MEC infiltrating up to the lamina propria but underlying muscle, cartilage and adventitia were free of disease. The bronchial margins were also free of disease without any lymphovascular or perineural invasion. The child is doing well and has no recurrence on 1-year follow up.

2. Discussion

Primary pulmonary malignancy is very rare in children; most of the literature is on adult patients. In pediatric age group only isolated case reports or small series has been reported and even they do not segregate the intarparenchymal from intra bronchial tumors. It is documented that 65% of these lesions are malignant [3]. As intra bronchial tumors are rare, often they are not considered in differential diagnosis of persistent pneumonitis and atelectasis [4]. In children, common causes of intra bronchial mass include

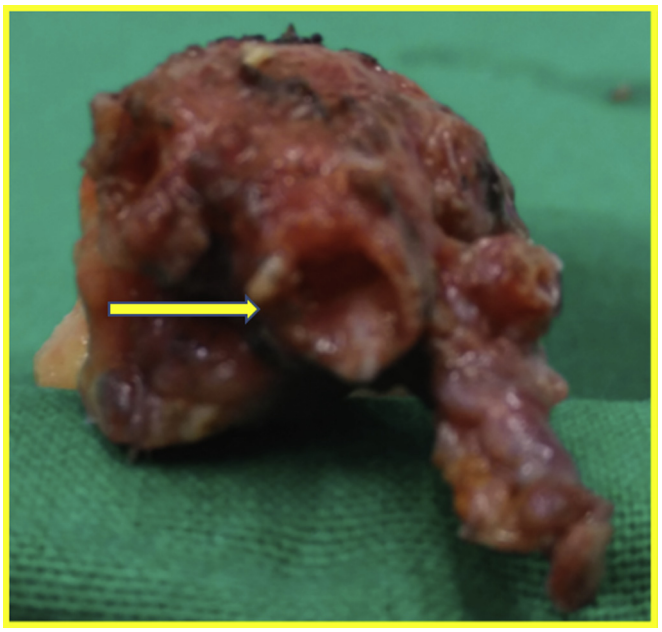

Fig. 2. Picture of specimen with opening of bronchus (arrow mark: bronchial opening).

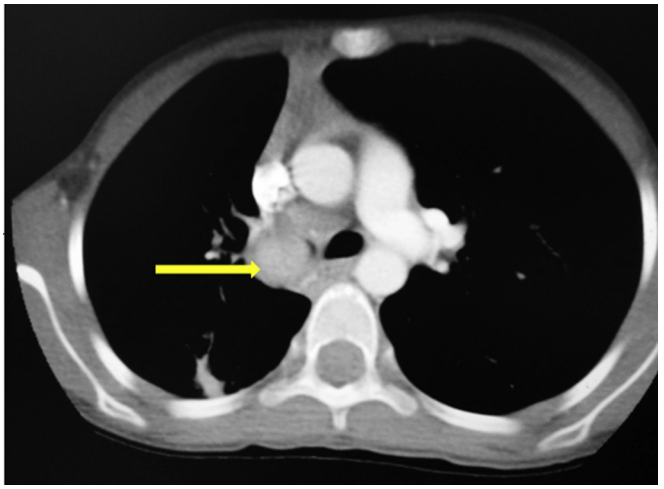

Fig. 3. Picture of specimen (arrow mark: mass within the bronchus).

aspiration of exogenous objects, respiratory tract papillomatosis, an inflammatory pseudotumor and very rarely, a metastatic or primary tumor. This often delays the correct diagnosis affecting prognosis of the disease.

The child with recurrent chest infections presents the clinician with a difficult diagnostic challenge. Does the child have a simply-managed cause for their symptoms, such as recurrent viral respiratory infections or asthma, or is there evidence of a more serious underlying pathology, such as bronchiectasis or tumors. Evaluation of children with chronic respiratory problems commonly includes blood tests, radiological analysis (thorax X-ray and occasionally high resolution computerized tomography) and lung function tests while bronchoscopy with or without intra bronchial biopsy is rarely needed in diagnostics. Only few studies have been published where a substantial number of children with different indications and symptoms were evaluated with bronchoscopy. Indications and results of 386 pediatric bronchoscopies were retrospectively investigated by Puhakka et al. There were 235 boys (61%) and 151 girls (39%). Eight percent of the girls were under 1 month of age, compared with only 4 percent of the boys. In children under 1-year of age, the most common indications were dyspnea (32%) and anomaly or suspicion of anomaly (20%). In children over 1-year of age suspected foreign body (32%) and recurrent respiratory infections (30%) were the most common indications. In 15 percent of patients, laryngomalacia and/or an abnormal epiglottis was confirmed. Subglottic stenosis, congenital or acquired, was confirmed in 9 percent and tracheal compression in 12 percent of cases. Seventeen percent of the children showed totally normal findings. When symptoms imply structural abnormalities, bronchoscopy provides the only reliable means to examine the anatomy of the larynx and main bronchi. Moreover, analysis of intra bronchial biopsies and bronchoalveolar lavage (BAL) can provide information about inflammation and infection [5,6].

In present literature, terms bronchial and pulmonary adenoma are rarely used because it comprises of both malignant and benign neoplasms including malignant carcinoid tumors, MEC, adenocystic carcinoma and benign mucous gland adenomas. However, in most of the reported literature 80%–90% of the pulmonary adenomas comprises of carcinoid tumors. Roby et al. suggested that in pediatric intra bronchial tumors; carcinoids may not out number MECs to the extent that it was reported in the past. The probable reason of this difference is that mucoepidermoid tumors arise in the minor salivary glands of the trachea and bronchi but are unlikely to arise from the lung parenchyma, where there are fewer minor salivary