Case Report

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High-grade Pulmonary Mucoepidermoid Carcinoma in a Child: A Rare Case Report

Pathology Section

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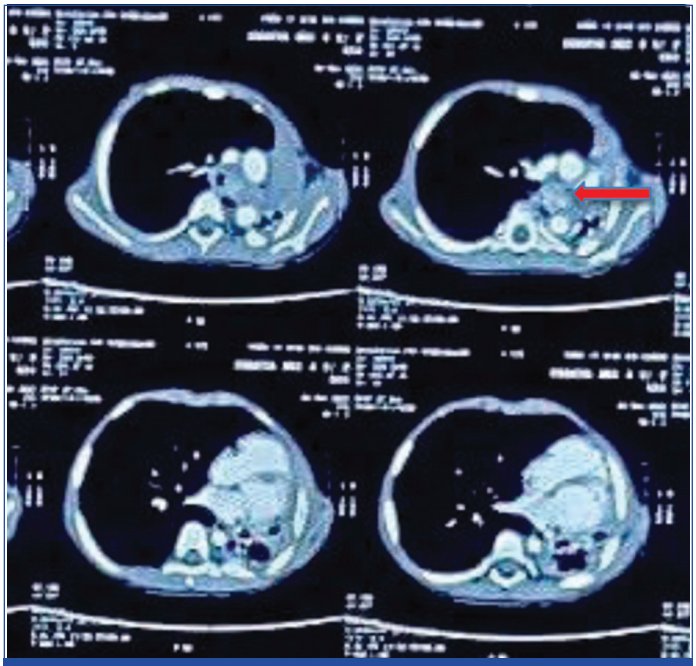
ABSTRACT

Mucoepidermoid Carcinoma (MEC) of the lung is a rare salivary gland-type tumour, accounting for <1% of primary pulmonary neoplasms, with female predominance in some series and a wide age range. This case reports a four-year-old girl with a four-month history of cough, haemoptysis, and breathlessness. Imaging revealed a 3.2×1.5×2.5 cm enhancing polypoidal mass arising from the left main bronchus, extending into the carina and lower trachea, with mediastinal shift and bronchiectatic changes. The patient underwent pneumonectomy and the histology showed Squamous metaplasia without dysplasia beneath that showed a neoplasm was composed predominantly of squamoid and intermediate cells, with cystic spaces lined by mucin-secreting cells and focal necrosis. Immunohistochemistry showed Tumour protein (p63) positivity in intermediate cells. Based on the Armed Forces Institute of Pathology (AFIP) and Memorial Sloan Kettering Cancer Center (MSKCC) grading systems, the diagnosis of high-grade MEC was confirmed. MEC involves the CREB Regulated Transcription Coactivator 1 (CRCT1 formerly known as MECT1) and Mastermind Like Transcriptional Coactivator 2 (MAML2) genes, located on chromosomes 19p13 and 11q21, respectively. Epidermal Growth Factor Receptor (EGFR) overexpression, observed in salivary gland MEC, is more sensitive to gefitinib than other wild-type EGFR non-small cell lung cancer cell lines and may offer potential for targeted therapy, though its role in pulmonary MEC remains uncertain. Each case of pulmonary MEC is inherently novel due to its rarity, presentation and histopathological diversity. Sharing these findings is essential for helping clinicians navigate diagnostic challenges, refine protocols, and predict patient outcomes for these uncommon lung tumours. Prognostic factors include histological grade, tumour, node, metastasis (TNM) stage, resection status, nodal metastasis, and patient age. Recognition of its molecular profile may guide future targeted therapies.

Keywords: Bronchial obstruction, Endobronchial tumour, Histopathology, Immunohistochemistry, Salivary gland type tumour

CASE REPORT

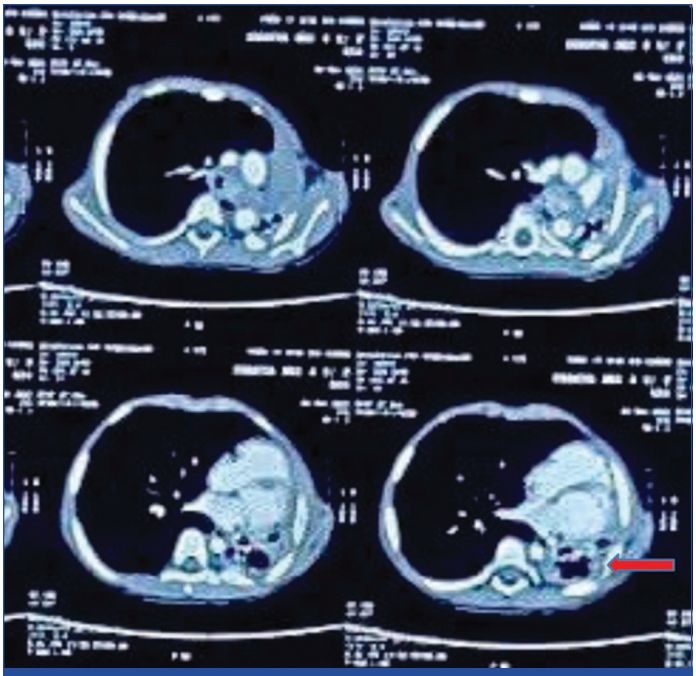
A four-year old girl was referred to Institute of Child Health (ICH) Pulmonology Department with the diagnosis of left main bronchus polypoidal mass with complaints of persistent cough and breathlessness with few episodes of haemoptysis in a span of four months. Her initial investigation showed X-ray with left lung opacities, investigation for tuberculosis including Mantoux test and sputum Cartridge-Based Nucleic Acid Amplification Test (CBNAAT), was negative. X-ray Imaging raised suspicion of left lung collapse or a foreign body for which fibreoptic bronchoscopy done outside revealed a left main bronchus polypoidal mass obscuring the lumen. She was then referred to the pulmonology department for further management. Contrast-enhanced computed tomography (CECT) of the chest revealed an intensely enhancing polypoidal mass arising from left main bronchus extending into carina and lower trachea [Table/Fig-1] with mediastinal shift to left with extensive bronchiectatic changes noted in left lower lobe [Table/Fig-2] and compensatory hyperinflation of right lung, in view of intensely enhancing mass lesion the possibility of vascular lesion was raised on imaging. A multidisciplinary team discussion involving paediatric surgery department, Cardiothoracic vascular department, ENT Department, Anaesthesiology decided for exploratory thoracotomy, excision of mass with ablation. The patient subsequently underwent a surgery. Intraoperative findings revealed that the left lung was completely collapsed, non ventilating, hard in consistency. On opening the bronchus, yellowish jelly-like mucoid material was seen extruding from the left main bronchus. Following drainage, whitish soft verrucous lesions were identified lining the interior of the left main bronchus, carina, and proximal right main bronchus, which bled actively on removal. In view of non viable nature of the left lung, a lung preserving strategy was abandoned and a left lung pneumonectomy was performed.



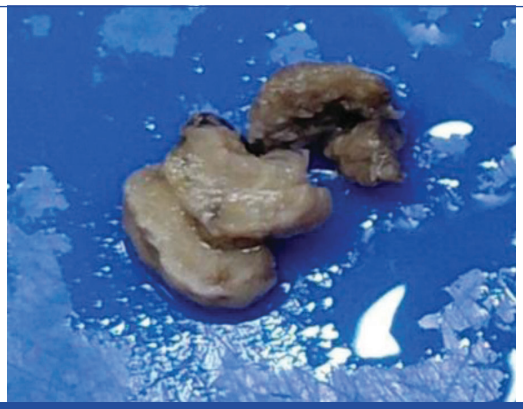
[Table/Fig-1]: Red arrow shows soft-tissue density lesion showing intensely enhancing polypoidal lesion obstructing the bronchus.

Then the polyp and pneumonectomy specimen was sent for histopathology.

The specimen sent in two formalin containers A and B. Sample A comprised of a single grey white polypoid soft tissue fragment measured 3.5×1.5×2.5 cm [Table/Fig-3] and Sample B contains single consolidated, atelectatic pneumonectomy specimen measured 8×6×5 cm, Cut surface showed multiple dilated bronchioles as cystic spaces filled with mucoid gelatinous material

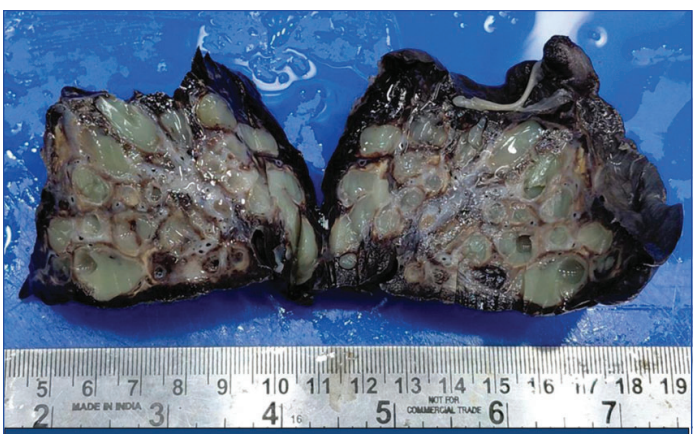


[Table/Fig-2]: Red arrow shows bronchiectasis changes noted in left lower lobe.

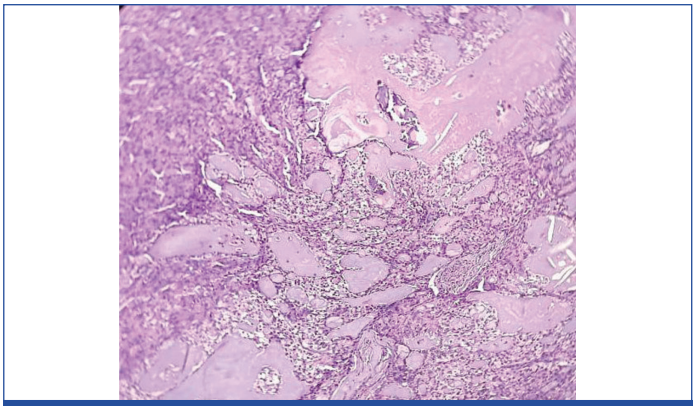


[Table/Fig-3]: Gross image A shows endobronchial polypoidal mass excision.

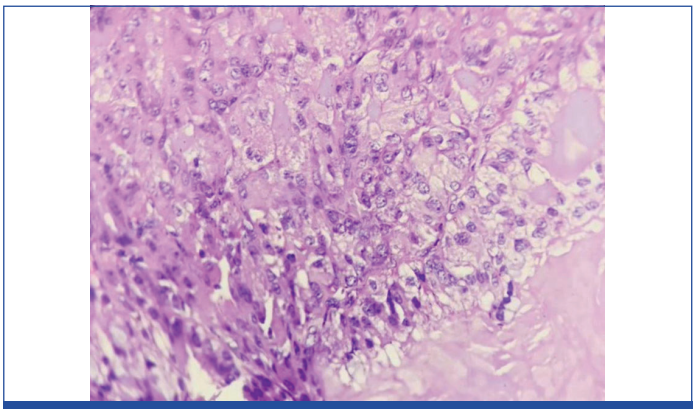
[Table/Fig-4], peripheral brown consolidated thin lung parenchyma noted. The histological examination of the polypoidal mass shows surface ulceration and focally lined by ciliated columnar epithelium with squamous metaplasia without dysplasia. Subepithelium showed a neoplasm composed predominantly of solid areas with squamoid and intermediate cells with eosinophilic to scant cytoplasm [Table/Fig-5] and pleomorphic nuclei. Adjacent areas showed cystic spaces lined by mucin-secreting cells [Table/Fig-6]. Microscopic examination with Hematoxylin and Eosin (H&E) stain showed these features. Focal area showed anaplasia [Table/Fig-7], and areas of necrosis also noted [Table/Fig-8]. No keratin pearls noted. The pneumectomy specimen showed bronchial dilatation with mucus



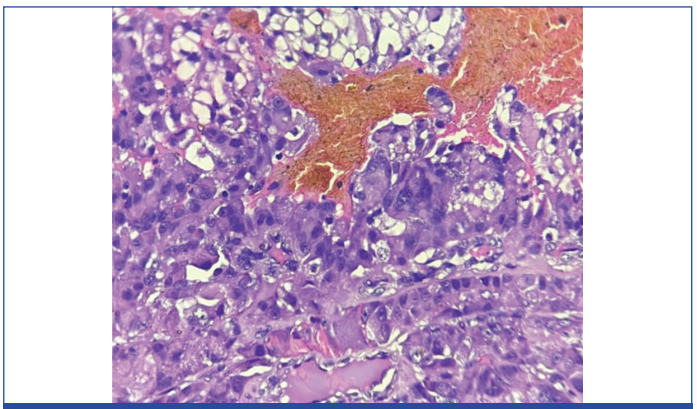
[Table/Fig-4]: Gross image B shows left Pneumectomy specimen cut surface shows bronchiectatic lung with dilated air spaces filled with mucoid material.



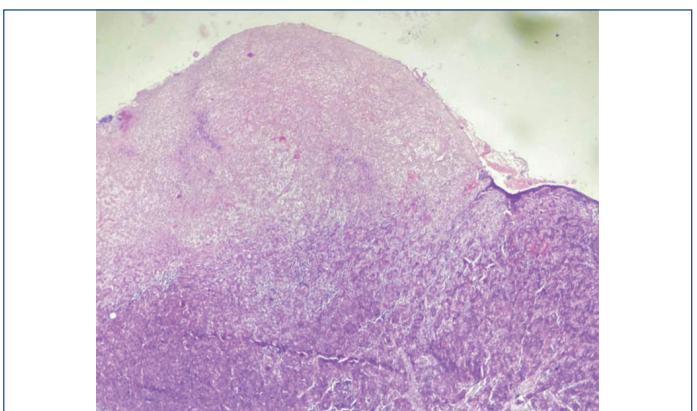
[Table/Fig-5]: Microscopic scanner view showing cystic and solid areas with cholesterol clefts, H&E stain (40×).



[Table/Fig-6]: Microscopic view showing cyst lined by clear mucin-secreting cells, adjacent to squamoid and intermediate cells with eosinophilic cytoplasm and pleomorphic nuclei, H&E stain (400×).



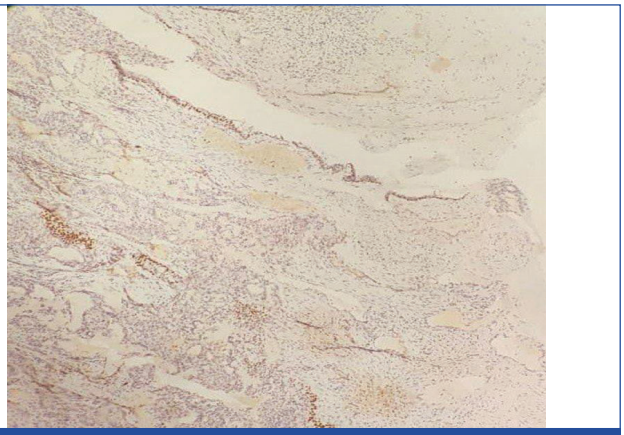
[Table/Fig-7]: Microscopic view showing areas of anaplasia, H&E stain (400×).



[Table/Fig-8]: Microscopic view showing focal areas of necrosis, H&E stain (40×).

plugging. The peribronchiolar areas shows chronic inflammatory cells infiltrates with lymphoid cells and fibrosis and the alveolar airspaces were collapsed. Based on the histomorphology, the diagnosis of MEC was made ruling out Squamous cell carcinoma, and Mucinous adenocarcinoma, Immunohistochemistry (IHC) for

p63 (tumor protein p63) was performed which showed scattered patchy nuclear positivity in intermediate cells and squamoid cells in the tumour [Table/Fig-9]. The tumour was classified as high grade under both major grading systems. According to the AFIP scheme by Goode RK et al., it fulfilled high-grade criteria due to prominent necrosis and anaplasia [1]. It was also graded as high-grade using the MSKCC qualitative system by Katabi N et al., based on the presence of necrosis [2]. Accordingly, a final diagnosis of high-grade pulmonary MEC was rendered. The postoperative period was uneventful, with the patient demonstrating satisfactory vital recovery. In view of the nature and aggressiveness of the lesion, a whole-body Positron Emission Tomography–Computed Tomography (PET-CT) scan was planned to evaluate for any additional lesions or distant spread. A referral to the radiation oncology and tumour board team was also scheduled, pending the PET-CT results, to determine the need for adjuvant therapy.



[Table/Fig-9]: Microscopic view showing scattered patchy nuclear positivity in intermediate and squamoid cells, favouring MEC, P63-IHC stain.

However, the patient was subsequently lost to follow-up before the planned investigations and oncological consultation couldnot be completed.

DISCUSSION

The MEC of the lung is an uncommon tumour derived from salivary gland-type cells, consisting of mucin-secreting, squamous, and intermediate cells, and they account for less than 1% of all lung cancers [3]. According to the World Health Organisation, these tumours are divided into low-grade and high-grade types. They mainly arise in the central bronchial region. Low-grade MECs generally have a favourable outcome with high survival rates, unlike high-grade MECs, which tend to behave aggressively similar to other non small cell lung cancers [4]. Diagnosis is primarily based on histopathological examination since clinical symptoms and imaging are not definitive [5].

Epidemiologically, these tumours represent 0.1% to 0.2% of primary lung tumour and can occur across all age groups, with about half presenting before age 30, affecting both males and females equally [6]. MEC tumours predominantly grow endobronchially in the central airways but can occasionally be found in peripheral lung areas [7], similar to the presentation of our case. The t(11; 19)(q21;p13) is the major chromosomal abnormality observed in salivary gland MECs, involving 2 genes: mucoepidermoid carcinoma translocated 1 (MECT1, now known as CRCT1) gene and a Mammalian Mastermind-Like 2 (MAML2) gene located at chromosomes 19p13 and 11q21, respectively [8]. The fusion protein MECT1-MAML2 has been implicated as a causative genetic event in salivary and BPMECs. Several studies have shown the impact of MECT1-MAML2 on the diagnosis and prognosis of salivary gland MEC [9]. The CREB-binding domain of CRTC1 (42 aa) is fused to the transcriptional activation domain of MAML2 (983 aa) to form the chimeric protein CRCT1-MAML2, which activates CREB target gene transcription and promotes MEC growth and survival [10]. CRTC1-MAML2 co-activates CREB to upregulate the Epidermal Growth Factor Receptor (EGFR) ligand, Amphiregulin (AREG). Aberrant AREG-EGFR signaling is critical for the oncogenic function of CRTC1-MAML2 [11].

Clinically, patients often present with symptoms related to airway obstruction [12] such as cough, haemoptysis, chest pain, wheezing, and recurrent pneumonia similar to the presentation of our case. They usually present late as this is a slow growing tumour [13]. Radiographically, chest X-rays and CT scans identify well-defined masses sometimes showing bronchial obstruction or atelectasis; however, imaging features lack specificity between tumour grades but high-grade tumour shows common signs of a lung carcinoma like invasive mass, pleural thickening and distant metastasis [14]. Most mucin-secreting areas of the tumours showed more densely distributed blood vessels, mostly capillaries, in between tumour cell nests, whereas other areas did less prominently in High-resolution Computed Tomography (HRCT) [15] similar to the presentation of our case.

Histologically, low-grade tumours are defined by a mix of mucin-producing, squamous, and intermediate cells. They typically present with a “cystic-plus-solid” architecture, these cystic areas lined by mild, columnar cells containing mucin; cell division (mitosis) is rarely seen while solid areas are composed of intermediate or squamoid cells that usually encircle the cystic structures. Intermediate cells appear as oval or polygonal cells with round nuclei and clear or eosinophilic cytoplasm [12], while high-grade variants are rare and significantly more aggressive, characterised by atypical intermediate and squamoid cells with high rates of necrosis and cell division. While mucin-secreting cells are present, they are less prominent which is also similar to the presentation of our case [16].

The Immunohistochemistry (IHC) is crucial to distinguish the different type of cells present in MEC and to rule out other lung carcinoma, MEC shows immunoreactivity for tumour protein p63 (p63), p40, cytokeratin 5/6 (CK5/6), and cytokeratin 7 (CK7) which confirms squamoid cells, and is positive for mucin 5AC (MUC5AC) highlights the mucin-producing component. MEC is negative for lung origin markers thyroid transcription factor-1 (TTF-1) and napsin A, myoepithelial markers smooth muscle actin (SMA), smooth muscle myosin heavy chain and calponin) and salivary mimic markers SRY-box transcription factor 10 (SOX10) and androgen receptor [14]. The expression of S100 protein (S100), carcinoembryonic antigen (CEA), and epithelial membrane antigen (EMA) is unpredictable and may vary from case to case [17].

Differential diagnosis for low grade includes mucous gland adenoma, carcinoid tumour, and high grade includes adenosquamous carcinoma, Necrotising sialometaplasia, Squamous cell carcinoma [7]. The lack of intermediate and squamoid cells discriminates mucous gland adenoma from low-grade MEC. The proximal exophytic endobronchial location; lack of individual cell keratinisation or squamous pearl formation; absence of tubular, acinar, and papillary growth pattern differentiates it from adenosquamous or squamous cell carcinoma.

The benefit of chemotherapy and radiation in advanced or high-grade MEC is uncertain [16]. Reports have shown that if EGFR mutation of the tumour is detected, tyrosine kinase inhibitors such as gefitinib may improve the prognosis in advanced pulmonary mucoepidermoid carcinoma (PMEC) patients [18]. Surgical removal is the preferred treatment offering excellent long-term survival in low-grade cases. The most common procedure for MEC is lobectomy, but surgical resection is still the mainstay of the treatment [19], whereas in our case where they chose pneumonectomy due to non-viable nature of the left lung. A few groups reported PMEC cases that were successfully treated with bronchoscopic neodymium-yttrium aluminium garnet (Nd-YAG) laser surgery [20].

Those were interesting attempts because the laser surgery was much less invasive than conventional surgical intervention, however

long-term follow-up information is needed for validating the effects of these treatments [5]. High-grade MECs have a higher likelihood of metastasis, mainly to lymph nodes, bone, and skin, and carry a worse prognosis [13]. Paediatric cases have shown comparatively better outcomes [21]. Prognostic outcomes differ substantially by grade: low-grade MECs have a five-year survival nearing 95% [22]. Poor prognostic indicators include incomplete tumour removal which shows positive surgical margins and the presence of lymph node metastases.

CONCLUSION(S)

The MEC of the lung is an exceptionally rare salivary gland–type malignancy, representing less than 1% of all primary pulmonary neoplasms. Despite its indolent growth, while low-grade MECs often yield excellent survival rates following surgical resection, high-grade cases behave more aggressively, akin to other non-small lung cancers and are prone to metastasis. This specific case highlights the aggressive nature of high-grade MEC in a four-year-old child, a population where this diagnosis is particularly uncommon and typically carries a poor prognosis despite aggressive surgical intervention. Current prognostic factors include the histological grade, TNM stage, and the status of surgical margins. Although the role of adjuvant therapies remains uncertain for high-grade pulmonary MEC, identifying molecular profiles like EGFR overexpression may provide future opportunities for targeted treatments.

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