

Pulmonary mucoepidermoid lung carcinoma in pediatric confused with asthma

Le Thuong Vu^{a,1}, Nguyen Minh Duc^{b,c,d,1,*}, Thieu-Thi Tra My^b, Luong Viet Bang^e, Le-Thi My^f,
Le-Tu Phuong Thuy^g, Hoang-Van Sy^{a,**}

^a Department of Internal Medicine, University of Medicine and Pharmacy at Ho Chi Minh City, Ho Chi Minh City, Viet Nam

^b Department of Radiology, Ha Noi Medical University, Ha Noi, Viet Nam

^c Department of Radiology, Children's Hospital 2, Ho Chi Minh City, Viet Nam

^d Department of Radiology, Pham Ngoc Thach University of Medicine, Ho Chi Minh City, Viet Nam

^e Department of Pathology and Cytology, Tam Anh General Hospital, Ha Noi, Viet Nam

^f Department of Radiology, Vinmec Times City International Hospital, Ha Noi, Viet Nam

^g Department of Internal Medicine, Pham Ngoc Thach University of Medicine, Ho Chi Minh City, Viet Nam

ARTICLE INFO

Keywords:

Mucoepidermoid carcinoma
Children
Pediatric
Lung cancer

ABSTRACT

Pulmonary mucoepidermoid carcinoma (PMEC) is an extremely rare tumor of the respiratory system. The clinical presentation of PMEC is variable and nonspecific, including cough, hemoptysis, and wheezing, and may mimic other symptoms of pneumonia or asthma. Here, we present a case of PMEC in a 12-year-old male who was diagnosed with and treated for asthma for 2 years. The patient presented with symptoms of respiratory failure that did not respond to steroids or bronchodilator medications. Chest computed tomography (CT) scans revealed an endotracheal tumor. The patient underwent complete tumor resection, with no signs of recurrence 6 months after treatment.

1. Introduction

Primary pulmonary mucoepidermoid carcinoma originates from the glands that line the tracheobronchial tree [1] and represents approximately 0.1%–0.2% of all primary lung tumors [2]. PMEC often affects younger patients compared with other, more common types of lung cancer [3]. Due to the tumor location, patients typically present with symptoms associated with bronchial obstruction and atelectasis [3]. The tumors can be classified as low-grade or high-grade based on histopathological results [3]. Complete surgical resection remains the primary therapy for PMEC [4]. This case emphasizes the roles of imaging and histopathology in the diagnosis, exclusion of other diseases, and avoidance of misdiagnosis and mistreatment.

2. Case report

A 12-year-old male patient who was diagnosed with asthma 2 years prior presented with increasing shortness of breath, wheezing, and

cough. The patient had no history of allergies. The patient had previously been hospitalized several times due to the same symptoms and was treated with bronchodilators and steroids; however, the symptoms did not improve and appeared to increase in severity. A blood test revealed increased neutrophil cell count (13 G/L) and C-reactive protein level (25 mg/L). A chest computed tomography (CT) scan was performed, which revealed an intratracheal mass. This mass was well-circumscribed with homogeneous enhancement (Fig. 1). The lung parenchyma was normal, and no mediastinal lymph nodes were observed. Bronchoscopy and tumor resection were indicated. The histological results demonstrated that the tumor cells included epidermoid, mucous, and intermediate cells without keratinization (Fig. 2). The final diagnosis was a low-grade PMEC tumor with negative surgical margins. The patient was not treated with any adjuvant therapy. After surgery, the symptoms of breathlessness and wheezing disappeared. Chest CT scans 6 months after surgery showed no signs of recurrence (Fig. 1).

* Corresponding author. Department of Radiology, Pham Ngoc Thach University of Medicine, Ho Chi Minh City, Viet Nam.

** Corresponding author.

E-mail addresses: bsnguyenminhduc@pnt.edu.vn (N.M. Duc), hoangvansy@ump.edu.vn (H.-V. Sy).

¹ Two authors contributed equally to this article as co-first authors.

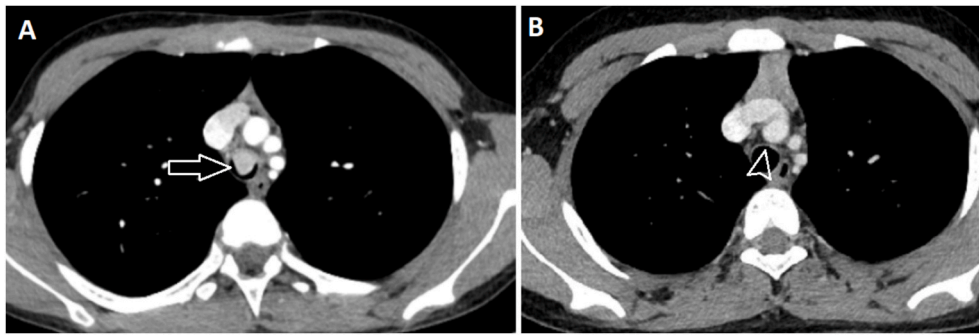


Fig. 1. Chest computed tomography (CT) scans for the patient. An intratracheal mass was observed, with well-defined borders and homogeneous enhancement (A, arrow). Six months after surgery, no signs of recurrence were observed (B, arrowhead).

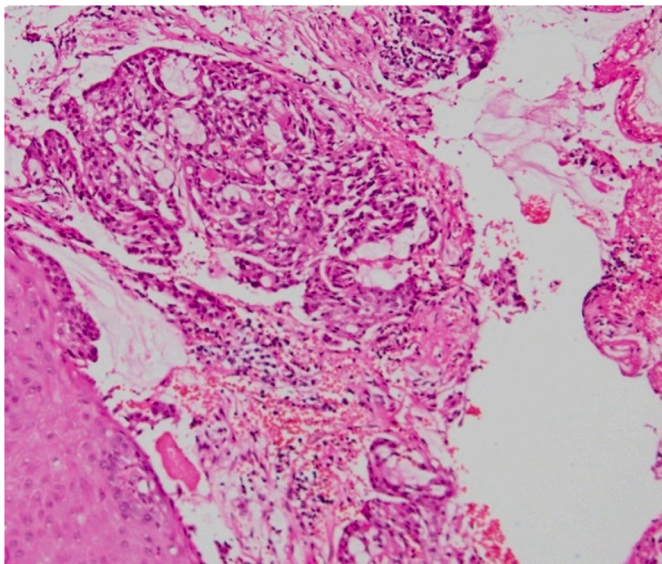


Fig. 2. Hematoxylin and eosin stain ($\times 100$) revealed that the tumor contained epidermoid, mucous, and intermediate cells.

3. Discussion

Lung cancer is quite rare in children. Smoking and asbestos exposure do not appear to be risk factors for P MEC [5]. P MEC affects male and female individuals equally and is primarily located in the trachea and bronchus [1]. Only 5% of P MEC cases are classified as high-grade, with the majority (95%) classified as low-grade [6]. Low-grade tumors often occur in young patients, whereas high-grade tumors are more likely to be observed in older patients [7].

The chest radiography may show consolidation, atelectasis, or a solitary lung nodule or mass; however, the chest X-ray may also appear normal in the case of a small endobronchial tumor without airway obstruction [8]. Chest CT scans typically show an endobronchial mass, with or without bronchial dilatation, and air trapping, obstructive pneumonia, or atelectasis [2,4]. Wang et al. [9] reported that low-grade P MEC is often located in the central bronchial or trachea, with smooth and well-defined margins, oval or lobular in shape, and markedly homogeneously enhancing; high-grade P MEC tends to be peripheral, with ill-defined margins, lobular, and heterogeneous, with reduced enhancement. High-grade P MEC can be difficult to differentiate from bronchial carcinoid tumors due to the hypervascularity of the tumor on CT images [6].

Bronchoscopy is commonly used to define the localization and obtain a biopsy for a definitive diagnosis. Macroscopically, P MEC cells include mucous, epidermoid, and intermediate cells, lacking in keratinization

[10]. The extracellular spaces are formed by the tumor cells and contain a mucoid substance [9]. High-grade tumors have increased nuclear pleomorphism, mitotic activity, and cellular necrosis but reduced mucoid substances and vessels compared with low-grade P MEC [9].

Surgical resection is the primary treatment option for patients with P MEC [11]. Multiple surgical approaches can be utilized, including lobectomy, segmental resection, or endoscopic removal, depending on the location and the extension of the tumor [11]. Adjuvant therapy is not indicated for cases of low-grade P MEC with complete resection [4]. No evidence currently supports the efficacy of chemotherapy or radiotherapy against high-grade P MEC, although epidermal growth factor receptor (EGFR)-targeted therapy has been suggested for unresectable or high-grade P MEC [12]. Low-grade P MEC is associated with a good prognosis and a 5-year survival rate of up to 95%, whereas high-grade P MEC is associated with a worse prognosis [3,10].

The patient in this article was a child who presented with respiratory tract obstruction symptoms and had been misdiagnosed with asthma for a long time. After complete tumor resection, the histological results revealed a low-grade P MEC; therefore, the patient was not indicated to receive adjuvant chemotherapy or radiotherapy. The symptoms of respiratory obstruction were completely solved by tumor removal.

4. Conclusion

Children who present with P MEC are rare, and the symptoms are often similar to other lung diseases, leading to delayed diagnosis. Chest CT scans may help determine the cause and exclude differential diagnoses, such as asthma and pneumonia. Most P MEC cases have a good prognosis, and a timely diagnosis and treatment may improve the overall survival rate of the patient.

Funding

Self-financed.

Author contribution

Le TV and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the content

Declaration of competing interest

Authors do not have any conflict of interests.

References

- [1] Z. Huo, H. Wu, J. Li, et al., Primary pulmonary mucoepidermoid carcinoma: histopathological and molecular genetic studies of 26 cases, *PLoS One* 10 (2015), e0143169, <https://doi.org/10.1371/journal.pone.0143169>.

- [2] Y.A. El-Sameed, S.H. Al Marzooqi, Primary mucoepidermoid carcinoma of the lung, *J Bronchol. Interv. Pulmonol.* 19 (2012) 203–205, <https://doi.org/10.1097/LBR.0b013e31825c6c30>.
- [3] S. Alsidawi, J.C. Morris, K.A. Wikenheiser-Brokamp, S.L. Starnes, N.A. Karim, Mucoepidermoid carcinoma of the lung: a case report and literature review, *Case Rep. Oncol. Med.* 2013 (2013) 1–5, <https://doi.org/10.1155/2013/625243>.
- [4] S. Jaramillo, Y. Rojas, B.J. Slater Bj, et al., Childhood and adolescent tracheobronchial mucoepidermoid carcinoma (MEC): a case-series and review of the literature, *Pediatr. Surg. Int.* 32 (2016) 417–424, <https://doi.org/10.1007/s00383-015-3849-y>.
- [5] S.R. Belgod, R.H.V. Reddy, S.P. Kumar, Mucoepidermoid carcinoma of the lung: a rare entity, *Oxford Med. Case Rep.* 201 (2015) 203–205, <https://doi.org/10.1093/omcr/omv012>.
- [6] B. François, M. Ammi, M. Rousselet, et al., Bronchial Mucoepidermoid carcinoma in a 14-year-old patient, *Arch. Pediatr. Surg.* 1 (2017), <https://doi.org/10.36959/472/346>.
- [7] N. Kalthor, C.A. Moran, Pulmonary mucoepidermoid carcinoma: diagnosis and treatment, *Expet Rev. Respir. Med.* 12 (2018) 249–255, <https://doi.org/10.1080/17476348.2018.1428563>.
- [8] P. Wildbrett, H. Lode, C.-D. Heidecke, N. Horras, R. Warzok, W. Barthlen, Mucoepidermoid carcinoma of the lung in a 6-year-old boy, *Afr. J. Paediatr. Surg.* 9 (2012) 159, <https://doi.org/10.4103/0189-6725.99406>.
- [9] Y.Q. Wang, Y.X. Mo, S. Li, R.Z. Luo, S.Y. Mao, J.X. Shen, Low-grade and high-grade mucoepidermoid carcinoma of the lung: CT findings and clinical features of 17 cases, *Am. J. Roentgenol.* 205 (2015) 1160–1166, <https://doi.org/10.2214/AJR.14.14153>.
- [10] A.C. Roden, J.J. García, R.N. Wehrs, et al., Histopathologic, immunophenotypic and cytogenetic features of pulmonary mucoepidermoid carcinoma, *Mod. Pathol.* 27 (2014) 1479–1488, <https://doi.org/10.1038/modpathol.2014.72>.
- [11] N.L. Vageriya, R.S. Shah, S. Prabhu, D. Naphade, H.R. Athawale, Intra bronchial mucoepidermoid carcinoma in an 8 year old girl: a case report of rare tumor with review of literature, *J. Pediatr. Surg. Case Rep.* 13 (2016) 41–44, <https://doi.org/10.1016/j.epsc.2016.07.002>.
- [12] J. Xi, W. Jiang W, S. Lu, C. Zhang, H. Fan, Q. Wang, Primary pulmonary mucoepidermoid carcinoma: an analysis of 21 cases, *World J. Surg. Oncol.* 10 (2012) 232, <https://doi.org/10.1186/1477-7819-10-232>.