Int J Clin Exp Med 2017;10(9):13906-13910
www.ijcem.com /ISSN:1940-5901/IJCEM0056003

Case Report

Bronchial mucoepidermoid carcinoma in a 12-year-old boy: a case report and literature review

Fengwei Kong[1*], Chunying Wang[1*], Heng Wang[2], Wenbin Wu[2], Miao Zhang[2], Dong Liu[2]

[1]Department of General Surgery, Xuzhou Infectious Disease Hospital, Xuzhou 221000, P. R. China; [2]Department of Thoracic Surgery, Xuzhou Central Hospital Affiliated to Southeast University, Xuzhou 221009, P. R. China. [*]Equal contributors.

Received March 10, 2017; Accepted September 10, 2017; Epub September 15, 2017; Published September 30, 2017

Abstract: Lung cancer in young patients is rare, which might be ignored or misdiagnosed empirically. Herein, a primary pulmonary mucoepidermoid carcinoma (MEC) was found in a 12-year-old boy, which was indicated pathologically through transbronchial biopsy. Single-stage bilobectomy of right middle and right lower lobes was performed, and a complete surgical resection of the tumor was achieved. The patient survived without local recurrence or distant metastasis during the follow up of 1 year. Because of its rarity, the diagnosis and therapy of lung cancer in very young patients might be delayed. Therefore, primary lung cancer should be kept in mind during the differential diagnosis of obstructive pneumonia or atelectasis in young patients to avoid empirical misdiagnosis. Besides, a timely aggressive resection of localized pulmonary MEC may lead to a satisfactory prognosis.

Keywords: Mucoepidermoid carcinoma (MEC), lung cancer, young patient

Introduction

It is reported that the misdiagnosis rate of lung cancer in young patient is 51.5%, and 11.8% of these patients are asymptomatic, while 85.4% of them are staged as III/IV on admission, besides, adenocarcinoma is the most common type [1]. Mucoepidermoid carcinoma (MEC) primarily occurs in salivary glands. Data from Surveillance, Epidemiology, and End Results Program reveals that 5.9% of MEC is originated from submucosal glands of the lung [2]. Biopsy through bronchoscopy is crucial for accurate diagnosis of this malignancy. Because of its rarity, the diagnosis and therapy could sometimes be delayed, thus, it is of vital importance for clinicians to consider the possibility of lung cancer in young patients, because a timely diagnosis might deliver single-stage radical resection and better prognosis, as surgery is the mainstay of therapy for MEC. Herein, a rare case of primary MEC in a very young patient is presented for discussion.

Case presentation

A 12-year-old boy was referred to Xuzhou Central Hospital, for persistent productive cough and intermittent high temperature lasting for nearly two months, without significant loss of weight or hemoptysis, on the suspicion of chronic pneumonia. However, the initially empiric antibiotic treatment did not work. His family history was unremarkable. He had no exposure history to tobacco, coal, cooking fumes, asbestos or heavy metals. This young boy was misdiagnosed as pneumonia empirically two months ago, without radiological examinations, with the aim to diminish radioactive damage to him because of his young age. He has suffered from recurrent productive cough every winter in the past 6 years, and he has been diagnosed and treated as chronic bronchitis during this period. Medical treatment was administrated routinely, as his pulmonary symptoms could be alleviated when the weather was getting warm.

Thorough physical examination of this boy including skin, oral mucosa and genital areas failed to identify any suspicious lesions, without signs of allergic pneumonia. To rule out special infections such as fungus, tuberculosis and viruses in this patient, blood and sputum cultures were performed for several times. However, the results from laboratory tests regarding

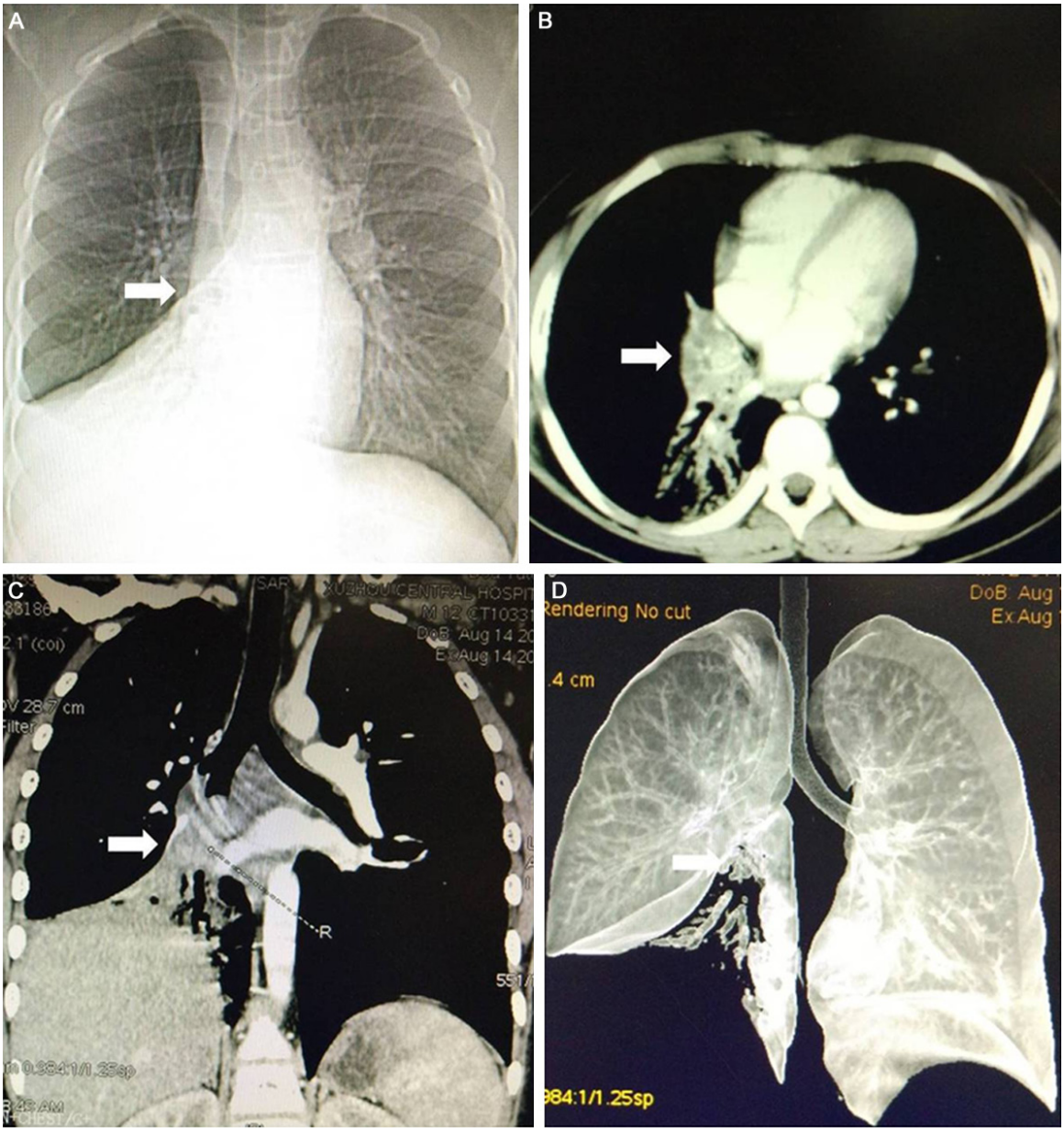

Figure 1. Chest X-ray and CT of the patient on admission revealed an intraluminal lesion in the right main bronchus, with atelectasis of the right middle and lower lobes.

special infection were normal. Besides, examinations for hepatitis and human immunodeficiency virus were negative. Then a chest X-ray was carried out, which indicated atelectasis of the right middle and lower pulmonary lobes. Additionally, three dimensional chest computed tomography (CT) reconstruction revealed an irregular pulmonary mass which blocked the brouchus of the right middle and lower lobes, without significantly enlarged mediastinal or celiac lymph nodes (**Figure 1**). Based on the radiological findings, the diagnosis of this boy was corrected as pulmonary tumor. Serum tumor markers including carcinoembryonic antigen, cytokeratin 19 fragment, squamous cell carcinoma, neuron specific enolase, carbohydrate antigens (CA) such as CA125 and CA19-9 were all in normal range. The score of Eastern Cooperative Oncology Group Performance Status of the patient was 1. Further abdominal CT, enhanced cranial magnetic resonance images (MRI) and bone emission computed tomography (ECT) excluded detectable distant metastasis. Positron emission tomography was not per-

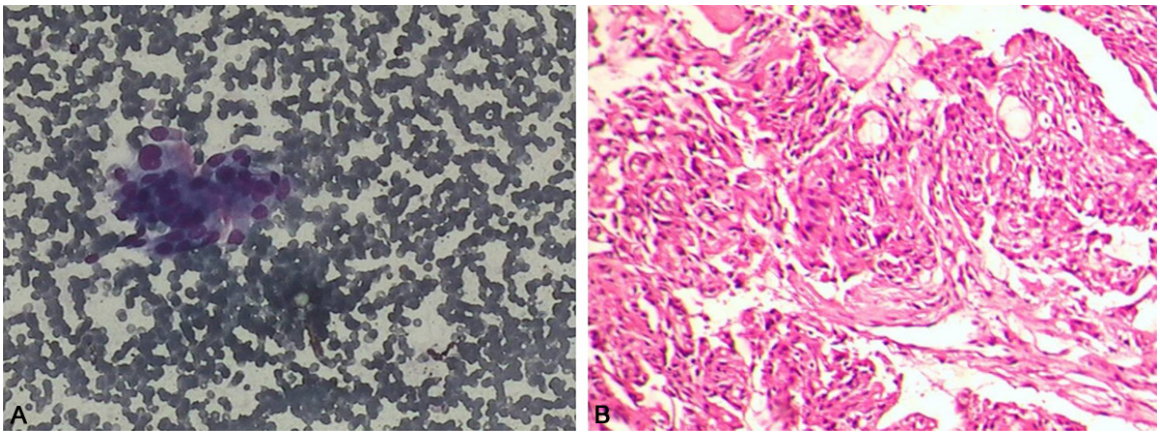

Figure 2. Biopsy of the mass located in bronchus displayed atypical malignant cells (A), and the histopathological examination revealed pulmonary mucoepidermoid carcinoma (B), HE staining (200 ×).

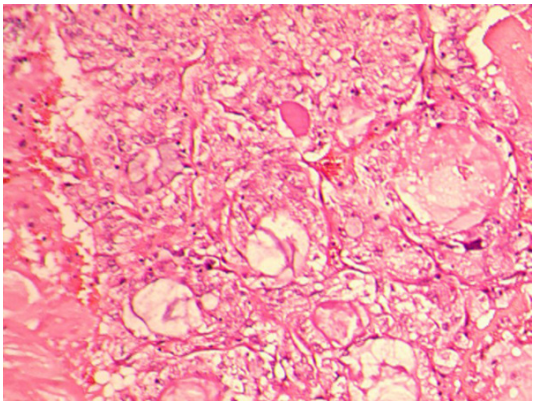

Figure 3. Postoperative pathological tests further confirmed the accurate diagnosis of pulmonary MEC in this patient, HE staining (200 ×).

formed because it was not covered by his health insurance.

Biopsy of the intraluminal lesion was performed through bronchoscopy to gain pathological diagnosis, and an irregular mass was observed in the right middle bronchus, which was confirmed as primary pulmonary MEC pathologically (**Figure 2**). Further immunohistochemical staining of the specimen revealed positive expression of cytokeratin 7, cytokeratin 5/6, P63 and Ki67+ (2%), and negative expression of cytokeratin 20, thyroid transcription factor 1 (TTF-1), epidermal growth factor receptor, thyroglobulin, cluster of differentiation 56 and chromogranin A. Then the patient was clinically staged as cT2N0M0 according to the 8th edition of American Joint Committee on Cancer TNM staging system for lung cancer.

As single-stage radical resection of the tumor was expected to be feasible after multidisciplinary consultation, aggressive surgery for this boy was approved by Ethical Committee of Xuzhou Central Hospital. Then bilobectomy of the right middle and lower lobes was carried out successfully, followed by mediastinal lymph nodes dissection via thoracotomy, under general anesthesia with double-lumen endotracheal intubation, in accordance with the principles of oncological surgery.

Postoperative pathological staining further confirmed his accurate diagnosis of primary pulmonary MEC, without visceral or parietal pleural invasion (**Figure 3**). The resected mass was 4 cm × 2 cm × 2 cm in size, meanwhile, the resection margins and dissected lymph nodes were tumor-negative. Therefore, the patient was pathologically staged as II A (pT2bN0M0). The postoperative recovery of this boy was uneventful, and he discharged 10 days after the surgery, without adjuvant chemotherapy or radiotherapy. Thereafter, whole-body CT, cranial MRI and bone ECT were carried out every 3 months for this patient, which excluded local recurrence or distant metastasis during the follow up of 1 year up to now.

Discussion

Little is known about the incidence, characteristics and outcomes of non-small cell lung cancer (NSCLC) in the young patients. The median age of lung cancer at diagnosis is 65-71 years [3-5]. It is reported that female gender, never-smokers, adenocarcinoma and distant metas-

tasis are more frequent in young lung cancer patients (age under 45 years), while the EGFR mutation rate is lower, as compared with older patients [3, 4]. However, the young patients have better overall and cancer-specific survival than the old cases [4, 5].

Bronchial MEC is exceedingly rare, accounting for 0.1-0.2% of primary lung cancers, which often presents as intraluminal masses, causing airway obstruction and recurrent pneumonia [6]. Pulmonary MEC demonstrates remarkable resemblance to MEC of the salivary glands [7]. Though its rarity, distant metastasis to brain has been reported [8]. The increased frequency of MEC in the pediatric population suggests a genetic abnormality, and recent genetic studies have demonstrated reciprocal chromosomal translocations [9]. Driver oncogenes including epidermal growth factor receptor mutation and echinoderm microtubule associated protein like 4-Anaplastic lymphoma kinase fusion gene are identified in 75% of the young lung cancer patients [10], however, risk factors of young patients are relatively uncertain. Exposure to tobacco smoking and mutagenic xenobiotics such as occupational exposures can lead to lung cancer [11]. On the contrary, tobacco control could efficiently reduce state-wide lung cancer morbidity in young adults [12]. Molecular features of pulmonary MEC include p63 expression and lack of keratinization, while TTF-1 and napsin are typically negative. Lastly, endobronchial cytology or CT-guided biopsy of the lesion is essential for accurate diagnosis and staging of MEC [13].

The therapeutic mainstay of pulmonary MEC is lobectomy with tumor-negative margin, followed by sampling or dissection of mediastinal lymph nodes, which might deliver cure of the disease, because a timely complete surgical resection is effective for pulmonary MEC, with favorable prognosis of the patients [14, 15]. Radiotherapy may be indicated for high grade MEC patients with positive surgical margins [16].

In summary, tracheobronchial MEC are rare in pediatrics, but it should be included in the differential diagnosis in a child with repeated respiratory symptoms [17], and a complete resection of the lesion is essential for long-term survival in MEC patients [18]. We reported a rare case of primary lung MEC in a 12-year-old patient, who was misdiagnosed as pneumonia empirically before admission, and a single-stage surgery showed a satisfactory oncological outcome.

Acknowledgements

This study was supported by Jiangsu Province Innovative and Entrepreneurial Talent Introduction Plan (Wenbin Wu, 2016), and Xuzhou City Science and Technology Project (No. KC16SH-102). This study was approved by the Ethics Committee of Xuzhou Central Hospital. Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Wenbin Wu, Department of Thoracic Surgery, Xuzhou Central Hospital Affiliated to Southeast University, No. 199 Jiefang South Road, Xuzhou 221009, China. Tel: +8618012018638; E-mail: wuwb2012@163.com

References

[1] Gong S, Sang C, Xu Z and Wang Y. [Clinicopathological characteristics of 130 cases of lung cancer in the youth]. Zhongguo Fei Ai Za Zhi 2014; 17: 465-468.

[2] Komiya T, Perez RP, Yamamoto S and Neupane P. Primary lung mucoepidermoid carcinoma: Analysis of prognostic factors using surveillance, epidemiology, and end results program (SEER). Clin Respir J 2015.

[3] Hsu CH, Tseng CH, Chiang CJ, Hsu KH, Tseng JS, Chen KC, Wang CL, Chen CY, Yen SH, Chiu CH, Huang MS, Yu CJ, Tsai YH, Chen JS, Tsai CM, Chou TY, Lin KC, Tsai MH, Lee WC, Ku HY, Liu TW, Yang TY and Chang GC. Characteristics of young lung cancer: analysis of Taiwan's nationwide lung cancer registry focusing on epidermal growth factor receptor mutation and smoking status. Oncotarget 2016; 7: 46628-46635.

[4] Thomas A, Chen Y, Yu T, Jakopovic M and Giaccone G. Trends and characteristics of young non-small cell lung cancer patients in the United States. Front Oncol 2015; 5: 113.

[5] Subramanian J, Morgensztern D, Goodgame B, Baggstrom MQ, Gao F, Piccirillo J and Govindan R. Distinctive characteristics of non-small cell lung cancer (NSCLC) in the young: a surveillance, epidemiology, and end results (SEER) analysis. J Thorac Oncol 2010; 5: 23-28.

[6] Thomas D, Modi Y, Dorai B and Guron G. A rare case of lung carcinoma with mucoepidermoid histopathology: a case report and review of the literature. Ann Clin Lab Sci 2015; 45: 219-221.
[7] Wang M, Ouyang S, Sun P, Li D and Huang G. Pulmonary mucoepidermoid carcinoma in Chinese population: a clinicopathological and radiological analysis. Int J Clin Exp Pathol 2015; 8: 3001-3007.
[8] Saito T, Ujiie H, Kadoyama S, Higa T, Shiono S and Teramoto A. Brain metastasis from a lung mucoepidermoid carcinoma mimicking a brain abscess. Surg Neurol Int 2015; 6: S300-303.
[9] Liu X and Adams AL. Mucoepidermoid carcinoma of the bronchus: a review. Arch Pathol Lab Med 2007; 131: 1400-1404.
[10] Nagashima O, Ohashi R, Yoshioka Y, Inagaki A, Tajima M, Koinuma Y, Iwakami S, Iwase A, Sasaki S, Tominaga S and Takahashi K. High prevalence of gene abnormalities in young patients with lung cancer. J Thorac Dis 2013; 5: 27-30.
[11] Landi S, Gemignani F, Canzian F, Gaborieau V, Barale R, Landi D, Szeszenia-Dabrowska N, Zaridze D, Lissowska J, Rudnai P, Fabianova E, Mates D, Foretova L, Janout V, Bencko V, Gioia-Patricola L, Hall J, Boffetta P, Hung RJ and Brennan P. DNA repair and cell cycle control genes and the risk of young-onset lung cancer. Cancer Res 2006; 66: 11062-11069.
[12] Polednak AP. Tobacco control indicators and lung cancer rates in young adults by state in the United States. Tob Control 2008; 17: 66-69.
[13] Roden AC, Garcia JJ, Wehrs RN, Colby TV, Khoor A, Leslie KO and Chen L. Histopathologic, immunophenotypic and cytogenetic features of pulmonary mucoepidermoid carcinoma. Mod Pathol 2014; 27: 1479-1488.
[14] Yamamoto T, Nakajima T, Suzuki H, Tagawa T, Iwata T, Mizobuchi T, Yoshida S, Nakatani Y and Yoshino I. Surgical treatment of mucoepidermoid carcinoma of the lung: 20 years' experience. Asian Cardiovasc Thorac Ann 2016; 24: 257-261.
[15] Lee GD, Kang DK, Kim HR, Jang SJ, Kim YH, Kim DK and Park SI. Surgical outcomes of pulmonary mucoepidermoid carcinoma: a review of 23 cases. Thorac Cardiovasc Surg 2014; 62: 140-146.
[16] Techavichit P, Hicks MJ, Lopez-Terrada DH, Quintanilla NM, Guillerman RP, Sarabia SF, Sayeed H, Nuchtern JG, Paulino AC, Muscal JA, Okcu MF and Chintagumpala M. Mucoepidermoid carcinoma in children: a single institutional experience. Pediatr Blood Cancer 2016; 63: 27-31.
[17] Jaramillo S, Rojas Y, Slater BJ, Baker ML, Hicks MJ, Muscal JA, Vece TJ, Wesson DE, and Nuchtern JG. Childhood and adolescent tracheobronchial mucoepidermoid carcinoma (MEC): a case-series and review of the literature. Pediatr Surg Int 2016; 32: 417-424.
[18] Yamamoto T, Nakajima T, Suzuki H, Tagawa T, Iwata T, Mizobuchi T, Yoshida S, Nakatani Y and Yoshino I. Surgical treatment of mucoepidermoid carcinoma of the lung: 20 years' experience. Asian Cardiovasc Thorac Ann 2016; 24: 257-261.