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

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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
special infection were normal. Besides, exami-
nations for hepatitis and human immunodefi-
ciency 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 comput-
ed 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 tu-
mor markers including carcinoembryonic anti-
gen, cytokeratin 19 fragment, squamous cell
carcinoma, neuron specific enolase, carbohy-
drate antigens (CA) such as CA125 and CA19-9
were all in normal range. The score of Eastern
Cooperative Oncology Group Performance Sta-
tus of the patient was 1. Further abdominal CT,
enhanced cranial magnetic resonance images
(MRI) and bone emission computed tomogra-
phy (ECT) excluded detectable distant metasta-
sis. Positron emission tomography was not per-
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 metastas-
Bronchial MEC are exceedingly rare, accounting for 0.1-0.2% of primary lung cancers, which often present 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.

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Disclosure of conflict of interest

None.

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References

Bronchial MEC in a very young patient


