High-grade pulmonary mucoepidermoid carcinoma supplied by the pulmonary artery and vein in a 6-year-old child: a case description and literature analysis

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Submitted Mar 15, 2022. Accepted for publication Jul 28, 2022. Published online Sep 07, 2022.
doi: 10.21037/qims-22-244
View this article at: https://dx.doi.org/10.21037/qims-22-244

Introduction

Primary pulmonary mucoepidermoid carcinoma (PMEC) originates from ductal epithelial cells in the submucosa of the trachea and bronchus and accounts for fewer than 1% of all primary lung malignant tumors (1). PMEC is also a primary lung cancer in children but extremely rare. Generally, PMEC in children is classified as low grade, and the high-grade subset is uncommon (2). However, in the relevant literature (3,4), the blood supply of PMEC was not extensively investigated. We here report a case of a 6-year-old child with high-grade PMEC supplied by the pulmonary artery and vein, mimicking pulmonary arteriovenous fistula and pulmonary sequestration.

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and the Helsinki Declaration (as revised in 2013). Informed consent was obtained from the patient's parents to publish this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

A 6-year-old girl developed a cough without an obvious cause, accompanied by left lower chest pain 3 days prior. The patient had a severe fever twice during the disease duration with a peak of 39.3 ℃. Her temperature could be reduced to normal after taking ibuprofen. One year earlier, she had been diagnosed with pneumonia in the left lower lung. After she presented to Children's Hospital of Chongqing Medical University, the laboratory tests showed an elevated white blood cell count of 15.9×10^9/L (normal value: 4.3–11.3×10^9/L), an increased percentage of neutrophil of 88% (normal value: 31–70%), a decreased percentage of lymphocytes of 7% (normal value: 23–59%), and a significantly increased C-reactive protein amount of 211.93 mg/L (normal value: <8 mg/L).

Chest non–contrast-enhanced computed tomography (CT) showed an irregular mass located in the dorsal and posterior basal segments of the lower lobe of the left lung 3.2 cm × 3.1 cm × 3.4 cm in size. The mass had an uneven density, and some calcification was located primarily in the center; a scattered cystic transparent shadow was observed (*Figure 1*). Slightly enlarged mediastinal lymph nodes and a few pleural effusions on the left were also observed. The contrast-enhanced CT showed uneven and obvious lesion enhancement (*Figure 2A*), with reduced enhancement areas, and the maximum CT value was 174 HU. The left inferior pulmonary artery and pulmonary vein branches entered the lesion (*Figure 2A,2B*). At the level of the fifth thoracic vertebra, 2 bronchial arteries about 1.2 mm in width on the right side and 1.1 mm in width on the left side were sent from the thoracic aorta into both lungs, respectively (*Figure 2C*). The distal part of the left bronchial artery is closely related to the branch of the left pulmonary artery.

The patient underwent thoracoscopic-assisted left lower lobe resection and left upper lobe partial resection. During

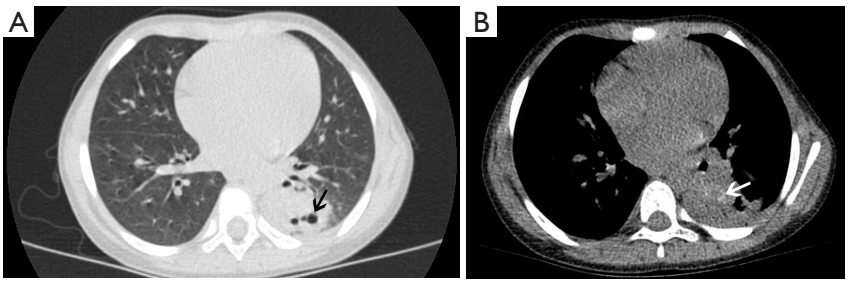

Figure 1 Non–contrast-enhanced computed tomography images. (A) A mass was visible in the dorsal segment of the lower lobe of the left lung, adjacent to the left lower lobe bronchus, with an irregular margin and cystic lucency (black arrow). (B) Calcification (white arrow) was apparent in the center of the lesion.

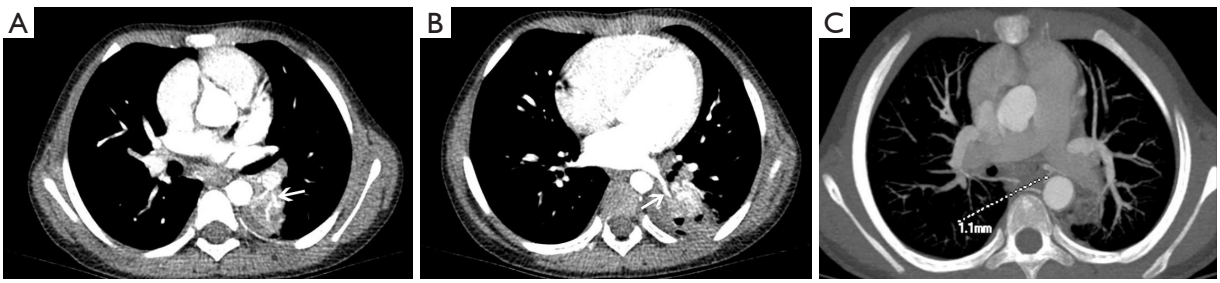

Figure 2 Contrast-enhanced computed tomography images. (A) The lesion was obviously enhanced unevenly, with branches of the left inferior pulmonary artery (arrow) entering the lesion. (B) Branches of the left pulmonary vein (arrow) also entered the lesion. (C) CTA-MIP showed 2 bronchial arteries at the level of the fifth thoracic vertebra about 1.2 mm in width on the right side and 1.1 mm in width on the left side, which were sent from the thoracic aorta into both lungs, respectively. CTA-MIP, computed tomography angiography-maximum intensity projection.

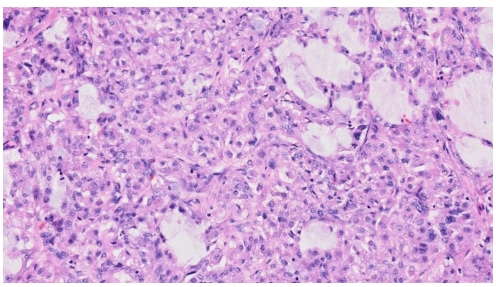

Figure 3 The pathologic view of the surgically resected specimen (hematoxylin and eosin staining, 400×). The tumor cells were arranged in an alveolar shape. Some tumor cells were epidermoid, and mucous cells were scattered.

the operation, the lesion was found to be in the lower lobe of the left lung, mainly in the dorsal segment, with a size of 4.0 cm × 3.5 cm × 3.0 cm, an unclear boundary, and no capsule. Pathological examination revealed an indistinct boundary between the lesion and surrounding tissues. Multiple cystic cavities with diameters of 0.5–1.5 cm, containing colloidal substances, were observed in the internal section. Microscopically, the tumor cells were arranged in an alveolar shape, and some tumor cells were epidermoid. We observed mucinous cells that were infiltrated and grew in nests, as well as focal calcification. The mitotic pattern was about 4–5 per high-power field (*Figure 3*). Immunohistochemistry results were as follows: cytokeratin (CK) +, P63+, Ki-67+ (5–10%), and periodic acid–Schiff (PAS)+.

Discussion

PMEC is rare and can develop in both adults and children. Although primary lung cancer rarely occurs in children, PMEC is the most common primary lung cancer in children, with its pathogenesis and causation still being unclear (5). The composition of PMEC is complex and mainly consists of different proportions of epidermoid cells,

mucinous cells, and intermediate cells that lack histological evidence of keratinization or squamous cell carcinoma *in situ* (5). PMEC can be divided into categories of low-grade and high-grade according to histological manifestations and cellular atypia. It has been reported that PMEC in children is mainly low grade, and tumor nuclear division is often invisible, while high-grade PMEC usually contains fewer mucous cells, and cystic components and tumor nuclear division are often not present simultaneously (6).

The predisposition site of PMEC is variously reported in the literature (3,4,7). PMEC in adults and children may be located in the lobe or segmental bronchus (3). Wang *et al.* (4) found that the pathological grades of PMEC had different predisposition sites, with high-grade PMEC tending to be peripheral. A previous report (7) showed that PMEC mostly appears as a regular round mass, sometimes with shallow lobulation, distributed in and outside the trachea or bronchial lumen or growing along the bronchial wall. When PMEC blocks the trachea or bronchus, atelectasis may occur, and it is easily misdiagnosed as pneumonia when secondary infection occurs. PMEC is usually of low to moderate density. Occasionally, a cystic shadow is observed, the pathological basis of which is related to mucus and cystic components of PMEC (5). Compared with other types of lung cancer, punctate calcification is prone to occur in PMEC, which is associated with the accumulation of mucus deposits secreted by mucous cells (4).

One study (4) demonstrated that PMEC mostly shows moderate to obvious enhancement after contrast is used, and high-grade PMEC mostly shows obvious uneven enhancement, which may be related to the dense distribution of blood vessels in the region of mucus cells (8). PMEC in children is mainly supplied by branches of the bronchial artery (9). The lesion in this case was mainly supplied by the branches of the left inferior pulmonary artery and pulmonary vein. However, the distal end of the left bronchial artery is closely related to the branch of the left pulmonary artery, and the left bronchial artery may also be involved in blood supply. In this case, it was mainly differentiated from 2 nonneoplastic diseases: pulmonary arteriovenous fistula and pulmonary sequestration. For pulmonary arteriovenous fistula, hemoptysis is the main symptom. Non–contrast-enhanced CT of the corresponding lesion area can show atelectasis or increased density. CT angiography (CTA) or digital subtraction angiography can show thickened and tortuous bronchial arteries and abnormal traffic vessels, which is helpful for diagnosis. Pulmonary sequestration is usually located in the paraspinal sulcus, including the intralobar and extralobar types. Contrast-enhanced CT can show the supplying arteries and draining veins from abnormal systemic circulation.

In conclusion, PMEC is rare in children. It is mainly distributed in and outside the trachea or bronchial lumen, and calcification can be observed in most lesions. After enhancement, most lesions are obviously unevenly enhanced, and CTA is helpful in clarifying the blood supply of the lesion. The biological behavior of PMEC is relatively good, and its clinical manifestations are nonspecific, commonly leading to its misdiagnosis as pneumonia. When children present with recurrent respiratory tract infection symptoms, we should be aware of the possibility of PMEC. Fiberoptic bronchoscopy is helpful for timely diagnosis.

Acknowledgments

Funding: The work was supported by the Medical Scientific Research Project of Chongqing Municipal Health Commission (No. 2022WSJK026), the Scientific and Technological Research Program of Chongqing Municipal Education Commission (No. KJQN202000440), and the Basic Research and Frontier Exploration Project (Yuzhong District, Chongqing, China; No. 20200155).

Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-244/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Informed consent was obtained from the patient's parents to publish this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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Cite this article as: Wang H, Chen X, He L. High-grade pulmonary mucoepidermoid carcinoma supplied by the pulmonary artery and vein in a 6-year-old child: a case description and literature analysis. Quant Imaging Med Surg 2023;13(1):546-549. doi: 10.21037/qims-22-244

