Case report

Congenital pulmonary airway malformation in the asymptomatic adult: A rare presentation

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A B S T R A C T

Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental abnormality of the lower respiratory tract, primarily diagnosed in the neonatal period. The most concerning sequelae for patients with CPAM are recurrent respiratory infections and malignancy. Rarely discovered in asymptomatic adults, CPAM presents challenging questions for management. We describe such a case and discuss the risks and benefits of resection.

1. Introduction

Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental abnormality of the lower respiratory tract with an estimated incidence of 1:25,000–1:35,000 births [1]. CPAMs are commonly diagnosed in the prenatal or neonatal period. The majority of neonates with CPAMs are asymptomatic however; others will develop respiratory distress and require urgent surgical resection [2]. Adults with CPAMs can develop recurrent pulmonary infection, malignancy, and pneumothorax but some are asymptomatic. Given the paucity of data on asymptomatic adults, the optimal management of CPAM in the adult patient population is unknown and can make management decisions difficult. We describe a CPAM case in an asymptomatic adult patient that was managed with surgical resection.

2. Case presentation

A previously healthy 22-year-old female presented to the emergency department with several hours of nausea, vomiting, and abdominal pain. The patient was afebrile and denied any cardiopulmonary symptoms. Physical examination was only remarkable for tachycardia. She denied any current or history of cardiopulmonary disease. She also denied history of recurrent pulmonary infection or pneumothorax as a child or an adult. A computed tomography (CT) of the abdomen and pelvis showed an incidental left lower lobe (LLL) consolidation. A chest CT with contrast was obtained in the emergency department with several hours of nausea, vomiting, and abdominal pain. The patient was referred to a cardiothoracic surgeon and subsequently underwent a sublobar LLL resection by video-assisted thoracoscopic surgery (VATS). There were no immediate surgical complications. Surgical pathology revealed a dominant cyst with multiple smaller cystic spaces lined by respiratory-type epithelium along with many cysts surrounded by smooth muscle. Air-fluid levels on chest CT were concerning for active or prior infection however; the histopathology findings were consistent with CPAM Type 1 without evidence of acute infection or inflammation. Tissue cultures were sterile.

3. Discussion

Although the overall incidence of CPAM is very low, CPAMs represent 25% of all congenital lung malformations [3]. Most patients remain asymptomatic at birth however; complications such as recurrent pulmonary infection, malignancy, and pneumothorax can occur in adults [4]. The primary differential diagnoses are bronchopulmonary sequestration (BPS), congenital lobar emphysema, and bronchogenic cyst. All patients should undergo a chest CT with contrast to evaluate for a feeding vessel, which can arise from the descending aorta. BPS and CPAM have been observed together in the same patients [5]. The mean age of adult presentation of Type 1 CPAMs is 36, accounting for 60–70% of cases [3]. Type 1 CPAMs are characterized by a single or multiple cysts more than 2 cm in diameter lined by pseudostratified ciliated columnar epithelium and walls containing smooth muscle and elastic tissue. CPAMs are classified from type 0 to type 4 malformations, which reflects the progression of developmental abnormalities from the large airways to the bronchioles and alveoli [6].

Recurrent pulmonary infection is the most common complication of CPAM however; the dreaded complication is malignant transformation. Type 1 CPAMs have been associated with the development of adenocarcinoma in situ (AIS), formerly known as bronchoalveolar carcinoma, in adults. The incidence of malignant transformation from Type 1
CPAM to AIS is unknown and some authors postulate the overall risk is low [6]. Pleuropulmonary blastomas (PPB) can complicate Type IV CPAMs but have been mostly described in infants and young children [7].

Our adult patient presented with radiographic findings consistent with CPAM but without any pulmonary symptoms. Previous studies have suggested that most pulmonary infections associated with CPAMs occur in the first three years of life [8]. We suspected our patient's risk for developing recurrent pulmonary infection was low however; the patient's risk for malignant transformation was less clear. The literature suggests an association of CPAM type 1 and AIS but overall this relationship is poorly established.

After discussion of risks and benefits, our patient opted to undergo surgical resection given her low surgical risk and concern for future CPAM complications. The patient had a successful sublobar resection with an uncomplicated postoperative course. Our case exemplifies several important points. First, a transparent discussion with the patient regarding risks, benefits and the lack of high-quality data on this topic is important. Second, a VATS sublobar resection can be successful and well-tolerated depending on the size and location of the CPAM. Lastly, CPAM should be considered as a differential diagnosis in adults presenting with a multicystic pulmonary mass, with or without air-fluid levels.

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