Case report: The natural history of congenital pulmonary airway malformations diagnosed in adulthood

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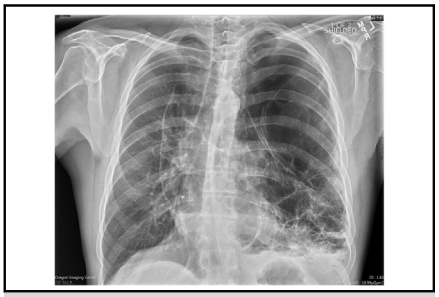

Extensive unilateral thin-walled cysts with anterior herniation.

CENTRAL MESSAGE

The natural history of prenatally diagnosed congenital pulmonary airway malformation has been described; however, none have followed patients into the fifth decade of life.

This is a 49-year-old man, with a 20-pack year smoking history, who was incidentally found to have cystic lesions of the left lung following a bicycle collision in 2011. Four years later, he developed exertional dyspnea, and radiograph of the chest showed unilateral cystic lesions with mediastinal shift (Figure 1). A nonquantitative perfusion scan demonstrated decreased perfusion and ventilation, and the patient was referred to a pulmonologist. While awaiting his appointment, his primary care physician prescribed multiple courses of prednisone for a diagnosis of emphysema exacerbation. Unfortunately, the patient was lost to follow-up. He represented in 2020 with dyspnea, and computed tomography scan of the chest showed collapse of the left upper and left lower lobes (LLL) with bullous cystic changes (Figure 2). He was seen by the pulmonologist in 2021, where the differential diagnosis widened and was referred to thoracic surgery.

Repeat computed tomography of the chest showed worsening cystic changes throughout the left lung with no definite major fissure, decreased caliber of the left main pulmonary artery, and atretic left superior pulmonary vein (PV). Pulmonary perfusion of the left and right lung was 9.5% and 90.5%, respectively, on quantitative ventilation–perfusion scan. Pulmonary function tests revealed forced expiratory volume of 46% and diffusing capacity of carbon monoxide of 67% adjusted. Given these findings, we discussed a left upper lobectomy, possible pneumonectomy, for likely congenital pulmonary airway malformation (CPAM).

We performed a left posterolateral thoracotomy, with harvest of the fifth intercostal muscle flap as is our general practice. The left main pulmonary airway and superior PV were atretic, the inferior PV appeared small, and the LLL had visible cystic changes. These findings necessitated a left pneumonectomy. Following surgery, the patient was sent to the intensive care unit per our institutional protocol, transferred to the ward on postoperative day 1, and discharged home on postoperative day 7. Final pathology revealed markedly enlarged cystic left upper lobe with poorly formed airways and vasculature, consistent with CPAM and compressive changes to the LLL. Postoperatively, our patient had near resolution of exertional dyspnea.

The institutional review board of the Oregon Health and Science University determined that the proposed activity was not research involving human subjects. Although institutional review board approval was not required, written informed consent from the patient for the publication was obtained.

COMMENT

CPAM, previously known as congenital cystic adenomatoid malformation, is a developmental anomaly of the lower respiratory tract as a result of cessation of lung development during various stages of embryogenesis. Different types of CPAMs are thought to originate at different levels of the tracheobronchial tree, leading to distinct histopathologic differentiation, clinical features, and malignant potential.[1] The most common symptoms of CPAM include recurrent

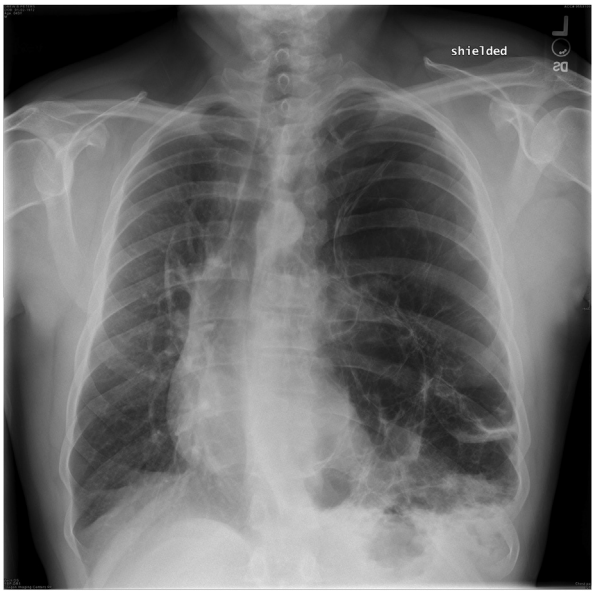

FIGURE 1. Radiograph of the chest from 2015 with thin-walled cysts and mediastinal shift.

pulmonary infection, respiratory distress, pneumothorax, and malignancy. The treatment for symptomatic CPAM is clear—surgery. However, treatment for asymptomatic CPAM remains less clear, as the long-term progression remains unknown.

Cook and colleagues[2] described the 10-year clinical course of 119 children prenatally diagnosed with CPAM. Fifty-one (43%) were managed surgically (average age of 1.6 years) and 68 (57%) conservatively. Of those managed conservatively, 52 (76%) continued to be asymptomatic for a median follow-up of 9.9 years. The conclusion of this study suggested that conservative management is a reasonable option in select cases.[2] Similarly, a retrospective Swedish study showed that 69% of patients could be managed conservatively with a median follow-up of 4 years. Prenatal regression was even demonstrated.[3] However, our case highlights that with longer follow-up, disease progression is possible.

Kotecha and colleagues[4] proposed 4 arguments to justify a surgical approach in the management of all asymptomatic CPAM—risk of malignancy, infection and pneumothorax, the potential for compensatory lung regrowth following early resection, and reduction in postoperative complications following elective surgery. Our case demonstrates an additional argument—early resection may allow for lung-sparing procedures. Stanton and colleagues[5] performed a meta-analysis of a 9-case series and calculated a greater than 2-fold increase in risk of postoperative complications in symptomatic compared with asymptomatic patients, further suggesting a role for elective surgery in asymptomatic patients. Although there were many barriers for our patient obtaining consistent care, earlier diagnosis with early resection could have entailed a lobectomy versus a pneumonectomy.

CONCLUSIONS

Although children diagnosed with CPAM may do well with conservative management, our case report demonstrates that not all CPAMs are diagnosed prenatally. Development of symptomatic disease in adulthood is possible and should be considered when patients do not respond to medical management. Our patient is recovering well from surgery, with high hopes that he will return to his active life.

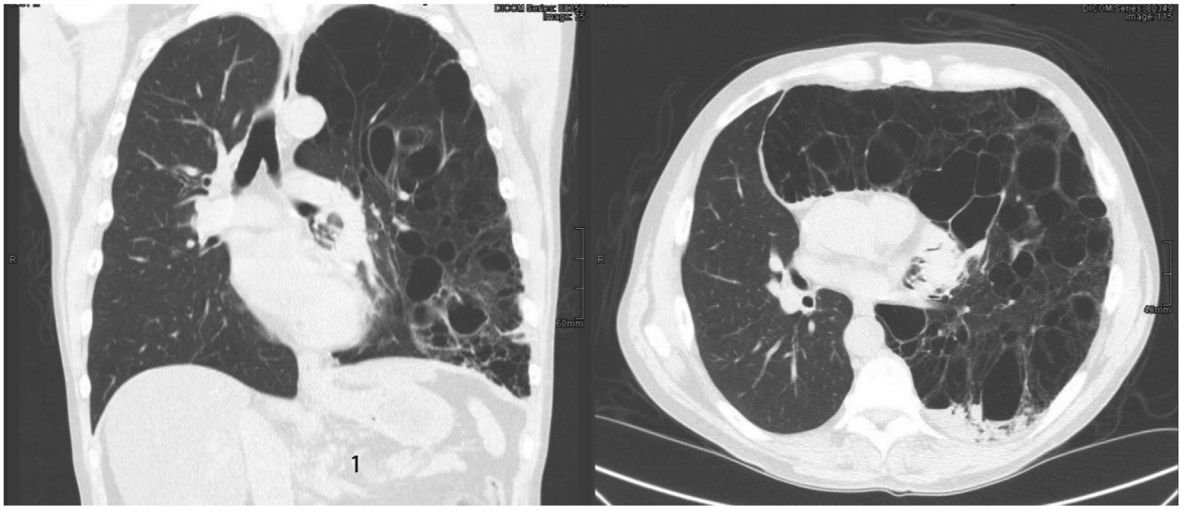

FIGURE 2. Computed tomography of the chest from 2020 with collapse of the *left upper* and *lower* lobes with bullous cystic changes and compression of the *right* lung due to mediastinal shift.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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