

Cystic Adenomatoid Malformation of the Lungs in Adults

To the Editor: Cystic adenomatoid malformation of the lungs is a congenital defect characterized by the proliferation of structures that resemble bronchioles and that eventually lead to the formation of cysts of different sizes. In 85% of patients, respiratory distress during the neonatal period or recurrent infections enable diagnosis in the first 2 years of life.¹ Diagnosis in adults and the involvement of both lungs are extremely uncommon. We present the case of a patient who required surgical treatment after being diagnosed with bilateral cystic adenomatoid malformation of the lungs.

The patient was a 21-year-old man with a history of asymptomatic congenital renal cysts and mild respiratory infections with constant relapses. In July 2003, a cystic structure was observed at the base of the right lung during an imaging study for a respiratory infection. Computed tomography (CT) of the chest revealed blisters in both lungs and a large cyst in the right lower lobe. The remaining examinations (complete blood count, biochemistry, and respiratory function tests) were normal. Once the case had been assessed, the presence of lung and renal cysts and the repeated respiratory infections led to diagnosis of cystic adenomatoid malformation. Given that the only manifestations were mild infections but that involvement was bilateral, outpatient follow-up was scheduled. In December 2005, he presented with massive hemoptysis that required admission to hospital. The findings of fiberoptic bronchoscopy were normal, with no traces of bloody material or endobronchial lesions, but a cyst in the right lower lobe and changes in the chest wall were evident in the chest CT scan, suggesting that the cyst was the cause of bleeding. After assessing the case, we decided to resect the cyst. The large cystic lesion visible by CT scan of the lower right lobe could be discerned through a right posterolateral thoracotomy. No anomalous arteries feeding the lesion were detected. A right lower lobectomy was done without any complications. The histologic and anatomic examination of the excised piece revealed a large cyst (3 × 5 cm) occupying 20% of the resected lobe (Figure). The final pathology report indicated a type I cystic adenomatoid

malformation of the lungs according to the Stocker classification.

To date, the patient has had no further episodes of hemoptysis or recurrent infection and is leading a normal life, similar to the one he was leading before the intervention.

Adult-onset cystic adenomatoid malformation is extremely uncommon but when it is detected, it is usually as a radiographic finding. The few patients who present with clinical symptoms usually have recurrent infections, hemoptysis, and recurrent spontaneous pneumothorax.² Malignant transformation has been reported.³ The diagnostic method of choice is high-resolution CT, which helps clarify the diagnosis by allowing excellent visualization of the cysts to determine their distribution, size, site, and wall thickness.⁴ Presentation patterns may vary; possibilities include multiple cysts, a dominant cyst among multiple cysts, and even a solid and homogeneous mass.⁵ The natural history of asymptomatic cystic adenomatoid malformation is unknown and there is no consensus about treatment. Treatment of symptomatic cystic adenomatoid malformation, both in adults and in children, is surgery. Lobectomy is the preferred type of resection, although a more extensive resection might be required when the lesion affects more than a single lobe.⁵ As indicated by Congregado et al,⁶ the initial approach may be by video-assisted thoracoscopy, as this allows complete resection of the lesions while avoiding the drawbacks of thoracotomy. In our patient, we did not use video-assisted thoracoscopy but rather direct posterolateral thoracotomy to assess the extent the parenchyma to be resected by palpation and meticulous inspection.

In conclusion, diagnosis of cystic adenomatoid malformation is based on high-resolution CT and the treatment of choice in symptomatic cases is surgery (by video-assisted thoracoscopy or thoracotomy) in view of the risk of recurrent infections and, in particular, the risk of malignant transformation.

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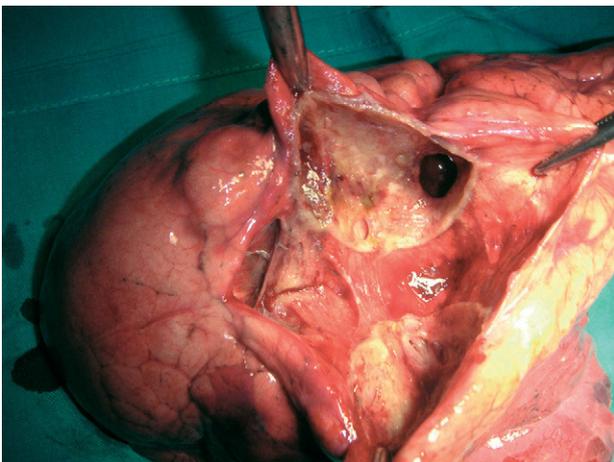


Figure. Resected piece from right lower lobectomy with a cystic adenomatoid malformation inside the lobe.