

Video-Assisted Thoracoscopic Surgery in 3 Cases of Adult Cystic Adenomatoid Malformation

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Congenital cystic adenomatoid malformation involving the lung is a rare hamartomatous condition that is usually diagnosed in the neonatal period. The presentation of this malformation in older patients is exceptional and usually manifests in a series of recurrent lung infections affecting a single lobe or segment. The treatment of choice is complete surgical exeresis.

This report of 3 cases of late presentation focuses on the surgical approach used and the unusual manifestation of recurrent spontaneous pneumothoraces in 1 patient. The patients were females aged 15, 16, and 25 years with histories of various respiratory diseases (extrinsic asthma, recurrent pneumonias, and pneumothoraces). The patients were referred to us for surgery with suspected diagnoses that were different from the final diagnoses in all cases. All underwent diagnostic video-assisted thoracoscopy to explore the affected hemothorax, and definitive treatment was possible during the procedure for 2 patients (a lobectomy and an atypical segmentectomy) by video-assisted surgery. The third patient underwent lobectomy by lateral thoracotomy after exploratory video-assisted thoracoscopy. Short- and long-term outcomes were excellent for all 3 patients.

Key words: *Congenital cystic adenomatoid malformation. Video-assisted thoracoscopic surgery.*

Tres casos de malformación adenomatoidea quística en el adulto tratados por cirugía videotoroscópica

La malformación adenomatoidea quística congénita del pulmón es una entidad hamartomatosa de carácter congénito y poco frecuente que suele diagnosticarse en el período neonatal. Su presencia en etapas más adultas es excepcional y se manifiesta, generalmente, como un cuadro de infecciones pulmonares de repetición que afectan a un solo lóbulo o segmento. El tratamiento de elección es la exéresis quirúrgica.

Se presentan 3 casos en los que se destacan la edad tardía de presentación, la rara forma de manifestarse uno de ellos (neumotórax espontáneo recidivante) y el abordaje quirúrgico utilizado para su resolución. Se trataba de 3 mujeres de 15, 16 y 25 años, con antecedentes de enfermedad pulmonar diversa (asma extrínseca, infecciones pulmonares de repetición y neumotórax recidivantes), que tras ser estudiadas en los servicios correspondientes (en ninguno se sospechó el diagnóstico final) nos fueron remitidas para tratamiento quirúrgico. A las 3 pacientes se les practicó un abordaje videotoroscópico para el diagnóstico y la exploración del hemitórax correspondiente (videotoroscopia exploradora) que permitió, además, el tratamiento definitivo en 2 de ellas (una lobectomía y una segmentectomía atípica) mediante cirugía videoasistida; a la tercera se le realizó una lobectomía por toracotomía lateral tras la videotoroscopia exploradora. En todas ellas se obtuvieron excelentes resultados tanto inmediatos como a distancia.

Palabras clave: *Malformación adenomatoidea quística congénita. Cirugía videotoroscópica.*

Introduction

Congenital cystic adenomatoid malformation of the lung is a rare cause of respiratory distress in neonates and is often associated with anasarca, ascites, and/or

polyhydramnios. The diagnosis of this malformation is unusual in older individuals and the usual clinical presentation includes recurrent respiratory infections and cavitation on chest radiographs, with images reminiscent of tuberculosis¹⁻⁴; rarely has spontaneous pneumothorax been described as the presenting event.^{5,6} This congenital malformation has also been associated with bronchioloalveolar carcinoma.^{2,7} Generally a single lobe or segment is involved, unlike the presentation in neonates, in whom the entire lung is usually affected. Treatment is surgical resection of the diseased lobe or segment, and full resolution is usually

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Manuscript received June 17, 2003. Accepted for publication October 14, 2003.

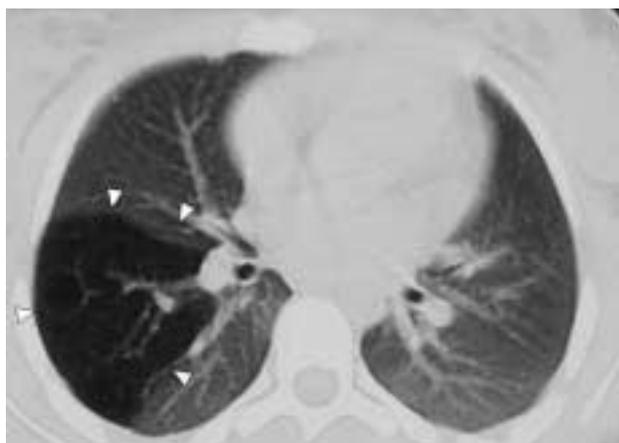


Figure 1. Computed tomography image showing an area consistent with diagnosis of lobar emphysema (arrows).



Figure 2. Tissue cross-section (hematoxylin and eosin, $\times 10$) pertaining to Case 2 showing cysts lined by columnar-type epithelium. An inflammatory infiltrate can be seen in the stroma, a finding typical of adult forms.

achieved.^{3,8,9}

Case Reports

Once surgery had been shown to be necessary, all 3 patients went through the usual preoperative protocol in our department: preanesthetic assessment, lung function studies, and stress testing if there was a question about the patient's tolerance for the proposed resection. None of the 3 cases we report required later follow-up spirometry, given that the values for forced vital capacity and forced expiratory volume in the first second (FEV_1) exceeded the established minimum ($FEV_1 > 2000$ mL in all cases) even for pneumonectomy. Surgery commenced according to our standard protocol for all 3 patients: contralateral decubitus position, selective intubation, and video-assisted thoracoscopic exploration of the pleural cavity using 3 entry incisions. This protocol allows the surgeon to plan the resection and choose an approach; it also facilitates biopsy sampling and intraoperative tissue study if indicated. Next, the required exeresis took place. Patient characteristics are shown in Table 1.

Case 1

A 15-year-old female diagnosed with rhinoconjunctivitis and extrinsic bronchial asthma had lesions consistent with congenital lobar emphysema in the right upper lobe (Figure 1) visible on radiographs, computed tomography (CT) scans and lung perfusion scans. She was referred to the surgeon for evaluation. Following satisfactory preoperative testing, with normal lung function tests, surgical excision of the lesion was scheduled. During video-assisted thoracoscopy, a cystic malformation was observed in the right upper lobe,

differentiated from the rest of the lobe by an intersegmental scissure. A 4 cm utility incision was made along the anterior line in the sixth right intercostal space, and the supernumerary lobe/segment was removed by video-assisted thoracoscopic surgery with simultaneous individual stapling, as described by Lewis,¹⁰ using an endostapler. The patient did not require intensive care after recovery and was discharged on the third postoperative day. The histologic diagnosis was type II congenital cystic adenomatoid malformation. Two years after surgery she was asymptomatic and radiographs and spirometry were normal.

Case 2

The patient was a 25-year-old woman whose relevant medical history started with fever diagnosed clinically as pneumonia at the age of 15 years. Since then she had experienced various respiratory infections that had been increasing in frequency and severity until at age 22 years she was hospitalized with a mass in the right upper lobe visible by radiography. After resolution of symptoms, a CT scan revealed a paramediastinal polycystic lesion in the right upper lobe. Tuberculosis was ruled out and she was referred for surgery.

Right upper lobe inflammation and adhesions at the apex and superior vena cava were observed during thoracoscopic exploration. The adhesions were cut and removal of the right upper lobe followed. A 4 cm utility incision was made in the sixth intercostal space and lobar exeresis was performed along with removal of the hypertrophic hilar and mediastinal nodes, under video guidance. The patient did not require intensive care upon recovery but discharge was delayed until the seventh postoperative day because of apical accumulation of air that resolved with chest physiotherapy. The firm histologic diagnosis was type II congenital cystic

TABLE 1
Clinical and Surgical Data for 3 Cases of Cystic Adenomatoid Malformation*

Case	Sex	Age	Side	Clinical/Radiologic Presentation	Resection	Surgical Approach	Histopathologic Diagnosis
1	F	15	Right	Congenital lobar emphysema	Segmentectomy	ExVAT/VATS	CCAM type II
2	F	25	Right	Recurrent infections	Lobectomy	ExVT/VATS	CCAM type II
3	F	16	Left	Recurrent pneumothorax	Lobectomy	ExVAT/lateral thoracotomy	CCAM type II

*F indicates female; ExVAT, exploratory video-assisted thoracoscopy; VATS, video-assisted thoracoscopic surgery; CCAM, congenital cystic adenomatoid malformation.

adenomatoid malformation (Figure 2). Examination of the excised mediastinal nodes indicated the presence of chronic reactive lymphadenitis.

Case 3

The third patient was a 16-year-old female with a history of 3 left spontaneous pneumothoraces at ages 14, 15, and 16 years. The first 2 episodes were treated with rest but after the third—involving a collapse of approximately 40%—she was referred to surgery. A pleural drain was inserted. A CT scan revealed part of the left upper lobe to contain thin-walled cysts of varying sizes within an area of reduced vascularization. During exploratory video-assisted thoracoscopy apical adhesions were cut away from the upper lobe. Intraoperative examination of the biopsy revealed cystic lesions in the pulmonary parenchyma consistent with a diagnosis of cystic adenomatoid malformation. A utility incision in the fifth intercostal space was made to attempt video-assisted lobectomy, but a finding of significant hilar fibrosis prevented dissection and suture of the vein of the upper lobe. Conversion to a lateral thoracotomy in the same intercostal space allowed the lobe to be removed. The patient remained in the intensive care unit for 24 hours and was discharged to home on the third postoperative day. The definitive histologic diagnosis was also type II cystic adenomatoid malformation.

Discussion

Cystic adenomatoid malformation is a rare, congenital hamartomatous pulmonary lesion described in 1949 by Ch'in and Tang.¹¹ Most reported cases present in neonates. Cases are infrequent in childhood and exceptional in adults.⁴ Most cases are unilateral, involving the entire lobe in neonates and a lobe or segment in adults. The literature suggests no difference in the prevalence in males and females, but curiously, all three of our patients were females. Nor has a predilection for one side or another been described: in our patients 2 malformations were on the right and the third was on the left.

During embryonic development, cystic lesions arise distal to a zone where bronchial segmentation is lacking. The lesions consist of an area of tumor development in immature lung tissue that may be accompanied, in varying degrees of severity, by cystic abnormalities in the form of spaces lined by a pseudostratified respiratory epithelium with diffuse mucus-secreting cellular elements resembling interconnected bronchiolar formations. The adult form has an abundant inflammatory component never seen in neonates, in addition to the pseudostratified respiratory epithelium and mucin-secreting cells. Classification into 3 types is based on cyst size.^{12,13} In type I, accounting for 65% of cases, there are few cysts present but they can reach 10 cm in diameter. In type II, accounting for 25% of cases, numerous small, uniformly shaped cysts will be found with diameters ranging from 0.5 to 2 cm, and other malformations are often present. Type III, accounting for 10% of cases, involves an essentially solid mass formed of very small

cysts less than 0.5 cm in diameter and other malformations are rarely present. Our patients had type II malformations, although none had associated lesions.

Although two of our patients had lobar cysts and one had segmental involvement, a pattern typical for adults, only one developed with the typical clinical picture of recurrent respiratory infections. Radiographic images of congenital lobar emphysema taken during tests ordered in relation to bronchial asthma led to surgical evaluation in the first case we have described. Finally, the uncommon pattern of presentation with recurrent spontaneous pneumothorax made diagnosis of the third case difficult, although similar cases have been described.^{5,6,14} All preoperative lung function studies indicated that tolerance of the planned resection would be optimal, given FEV₁ values exceeding 2000 mL, or greater than 60% of the predicted FEV₁. Video-assisted thoracoscopy was used for surgical exploration according to standard practice at our hospital¹⁵ and according to Rothenberg¹⁶ and Yim.¹⁷ Approaching the tissue to be excised through 3 points of entry allowed us to ascertain the feasibility of the planned resection, to biopsy the site intraoperatively in the third case, and to perform the resection by video-assisted surgery in the other 2 cases. In those 2 cases, a muscle-sparing minithoracotomy 4 to 5 cm was made along the anterior line at the fifth intercostal space, without use of a rib separator. A thoracotomy was necessary to complete resection in the third case, given that the severe hilar fibrosis made endoscopic vascular dissection risky. The third patient was the one who required 24-hour postoperative observation in the intensive care unit.

These cases are noteworthy for their unusual form of presentation, only one of which followed the typical pattern, and for the surgical approach taken. Video-assisted thoracoscopy led to correct diagnosis in these cases and in two of them we were able to treat the lesions surgically, using the same procedure as has been described in the literature.⁸⁻²⁰

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