

Video-assisted thoracoscopic lobectomy for congenital cystic adenomatoid malformations

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ABSTRACT

Aim The aim was to show rare cases of congenital cystic adenomatoid malformation (CCAM) and the manner of its surgical treatment with video-assisted thoracoscopic surgery (VATS).

Methods Two male and one female child, 7, 4 and 3 years of age were treated for symptoms of cough and high temperature in district hospitals. In all three children laboratory blood tests and chest radiography were done. Auscultatory findings showed the presence of pneumonia. Children were treated with appropriate doses of antibiotics. After the rehabilitation of inflammation, they were sent to the University Clinical Center Sarajevo, where video-assisted thoracoscopic lobectomy (VATS) was indicated after computerized tomography (CT).

Results Chest CT scan pointed to the CCAM and pulmonary sequestration (PS) changes to the lungs. This has required surgery lobectomy of an affected part of the lungs. In two children with PS, the aberrant systemic artery came from the most proximal part of aorta abdominals, the third patient did not have an anomalous artery.

Conclusion VATS lobectomy is an alternative to the traditional thoracotomy for the treatment of CCAM and PS, however, it should be investigated in the future for its safety and effectiveness.

Key words: lung malformation, thoracoscopic surgery, pulmonary sequestrations, cystic adenomatoid

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INTRODUCTION

The most common malformations of the lower respiratory tract are congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS) (1). Congenital adenomatoid lung malformation represents developmental pathologically changed lung tissue that normally communicates with the tracheobronchial tree and typically has proper circulation (2). Intralobar sequestration is a congenital malformation in which embryonic lung tissue is supplied by systemic arteries (2). Thoracoscopic lobectomy for congenital cystic adenomatoid airway malformation has been indicated from the neonatal period to adolescence (3). Surgical treatment is the only method that provides certain success and consists of removing parts of the lung tissue with cystic formations (3).

The reported incidence of CCAM ranges from 1/11,000 to 1/35,000 live births, and BPS is even more rare with no published population incidence (1).

The aim was to describe rare cases of congenital pulmonary airway malformation (CCAM) and the manner of their surgical treatment with video-assisted thoracoscopic lobectomy (VATS).

PATIENTS AND METHODS

Two male and one female child, 7, 4 and 3 years of age were treated for symptoms of cough and high temperature in district hospitals during 2015. In all three children laboratory blood tests, chest radiography, as well as auscultatory findings showed the presence of pneumonia. Children underwent long treatment with appropriate doses of antibiotics. After the rehabilitation of inflammation they were sent to the University Clinical Centre (UCC) Sarajevo for further treatment. Chest computerized tomography (CT) (Siemens Somatom, Munich, Germany) was done indicating the CCAM and pulmonary sequestration (PS) changes to lungs. CCAM has been classified histologically by Stocker et al. in 1977 based on cyst size and by Adzick et al. based on cyst appearance (micro or macrocytic) on antenatal ultrasound (1,4).

Surgery lobectomy of the affected part of a lung was indicated in all three patients and performed using video assisted thoracoscopic lobectomy

(VATS) with thoracoscopic video camera with deflectable tip (Olympus, Tokyo, Japan).

RESULTS

The CCAM right lower lobe in one case, pulmonary sequestration with CCAM (mixed lesions) in two cases (in one case right lower lobe, in the other case left lower lobe) was found. In both patients with pulmonary sequestration the aberrant systemic artery came from the most proximal part of aorta abdominalis. All three patients had extensive pulmonary infections and strongly enlarged hilar lymph nodes.

Thoracoscopic assisted lower lobe lobectomy was performed in all three children.

Surgical report

The operation was done under a general anesthesia with one sided-lung ventilation and collapse of the lung with the disease.

Patients were positioned in the stable lateral decubitus position, with their arm above their head and a roll under the axilla (caudal and parallel to the arm) to protect neurovascular structures crossing the axilla. The pelvis was supported posteriorly, and a strip of tape was applied for immobilization of the patient. The hemithorax was disinfected and draped. The surgeon was working from the front side of the patient. Three small cuts on the chest about 1 cm long were performed in the 5th intercostal space (ICS) mid-axillary line for 5 mm 30° telescope to look inside the chest. The other two incisions were made for the working instruments, one in the 4th ICS front axillary line (5mm port) and one in the 8th ICS front axillary line (10 mm port).

In cases with pulmonary sequestration, firstly the division of the aberrant artery was done. The artery was easily found in the lower pulmonary ligament which was divided and the artery freed over the length of 2-3 cm from adhesions and fibrotic sheath. In one case the artery could be clipped using Weck Hem-o-lok polymer (Teleflex, Wayne, Pennsylvania, USA) locking ligation system and in the other case the artery was more than 11 mm thick and vascular stapler Endo-GIA (Medtronic, Dublin, Ireland) 30-2.5 mm was used. The pulmonary ligament was further divided with hook cautery until the exposure of the inferior pulmonary vein, which was completely exposed.

Table 1. Summarized overview of three children with congenital cystic adenomatoid malformation (CCAM)

	Case No 1	Case No 2	Case No 3
Sex	Male	Male	Female
Ages (years)	4	7	3
Diagnosis at admission	Infiltration of lung	Bilateral bronchopneumonia	Infiltration of lung
Symptoms	Cough, high temperature, abdominal pain, hepatosplenomegaly	Cough, high temperature, vomiting	Cough, high temperature epiphora
Auscultation	Impaired respiratory murmur from late aid inspiration cracks	Late inspiratory crackles in left basal lung lobe	Late inspiratory crackles
Chest X-ray	Consolidation in the left lower lung lobe	Consolidation in the left lower lung lobe	Right hilar basal supradiaphragmatic, oval, relatively clearly limited inhomogeneous shading, diameter of 68x45 mm
Chest CT scan	Changes in density, a lot of cystic areas with the largest diameter of 24 mm, consolidations of lung parenchyma with small atelectasis	Changes in density, a lot of cystic areas with the largest diameter of 23 mm	On the right side and along the right main bronchus multi-cystic mass zones filled with air and liquid collections
Location	Left basal lobe	Left basal lobe	Right basal lobe
Infection	+	+	+
Supplying vessels	4.5 mm diameter artery derived from coeliac trunk (proximal part of a. abdominalis)	Proximal part of a. abdominalis	No supplying blood vessel
Associated anomalies	Cryptorchidism, Dolichocolon	Reversal of organs, total (situs inversus totalis)	No anomalies
Pulmonary sequestration type	Intralobar	Intralobar	Intralobar
Histopathology	CCAM type 2, with superimposed chronic inflammatory changes of lymphoid nodes and hyperplasia of lymphoid follicles	CCAM type 2, with superimposed chronic inflammatory changes of lymphoid nodes and hyperplasia of lymphoid follicles	CCAM type 2, with superimposed chronic inflammatory changes of lymphoid nodes (to 12 mm) and hyperplasia of lymphoid follicles
Final diagnosis	CCAM type 2	Bilateral bronchopneumonia CCAM type 2 Pulmonary sequestration	Bronchopneumonia CCAM type 2 Pulmonary sequestration

The dissection continued with the hook cautery opening the oblique interlobar fissure (major fissure) up to the mediastinal pleura. The branches of the lower pulmonary artery were localized, exposed and completely visualized. Due to extremely post-inflammatory situs and many adhesions in the major fissure this dissection was extremely difficult but successful. The segmental arterial branches to the lower lobe were identified and all of them one by one from medial to lateral divided after positioning of Weck Hem-o-lok clips through a 5 mm port.

To allow for further resection of the lower lobe, the major fissure was completely divided using a LigaSure (Medtronic, Dublin, Ireland) 5 mm blunt tip 37 cm sealer and divider.

The pulmonary vein was dissected but the adhesions between the vein and the bronchus with strongly hypertrophied lymphatic tissue and big lymph nodes were in all three cases so strong and difficult to dissect that in all cases at this stage a mini-thoracotomy was performed.

A 6 cm skin incision was done along the port in the 5th ICS and the soft tissues divided with

the cautery. The chest was entered in the 5th ICS along with the upper edge of the 6th rib. The final dissection between the lower lobe bronchus and the pulmonary vein was performed partially bluntly partially with the aid of Liga Sure. The lower lobe bronchus was finally always divided using the stapler two times, once for the segment 6 and once for the main part of the lower lobe bronchus before branching to other segments.

Finally, the inferior pulmonary vein was divided using the vascular stapler.

The mediastinal pleura was closed with Vicryl 4/0 stitches (Somerville, New Jersey, USA). With a warm saline solution in the thoracic cavity, the remaining lung was re-inflated to check for air leakage. At the end of the procedure, one thoracic drain was left behind, to evacuate residual air and blood. A 16-French chest tube (Medtronic, Dublin, Ireland) was placed anteriorly in middle position. The drain came out through the skin incision for the port in the 8th ICS and it was fixed by suture. The tube was connected to a chest drainage unit consisting of a collecting chamber and an underwater seal.

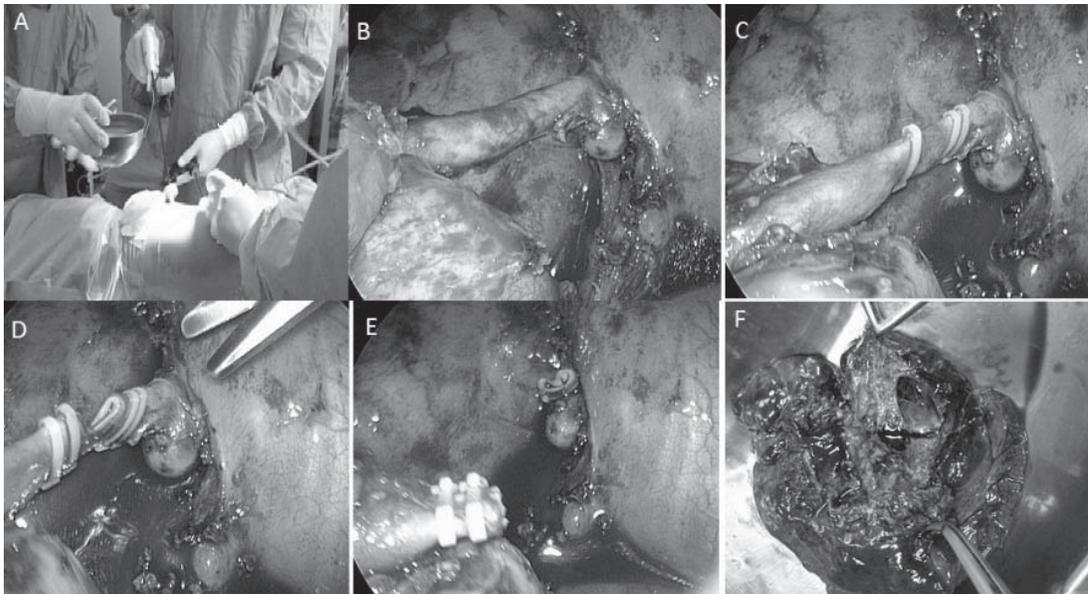


Figure 1. A) The operating table with thoracoscope, B) Division of aberrant artery, C) Ligature with Weck Hem-o-lok polymer locking ligation system, D) Cutting aberrant artery, E) Sectioned aberrant artery, F) Removed part of cystic altered lung tissue (Selimović A, 2015)

After securing hemostasis, four resorbable sutures Vicryl 2/0 were placed around the ribs to approximate the mini-thoracotomy. The muscle, subcutaneous tissue and skin were closed with resorbable sutures Vicryl. Dressing was applied.

DISCUSSION

Congenital pulmonary diseases such as CCAM and PS are the most common lung diseases in the pediatric population that require surgical treatment consisting of resection of the affected part of the lung (1). Patients with these diseases experience recurrent upper respiratory tract infection and pneumonia more frequently than healthy children (1,2).

The children described in this presentation had CCAM type 2, which is usually associated with other developmental abnormalities, and intralobar pulmonary sequestration, which is associated with other anomalies in 17% of cases (1). Type 2 CCAM is present in 10 to 35% of patients, cha-

racterized by many small cysts with diameter 1 to 10 mm, lined with cylindrical or cubic epithelium with prominent smooth or striated muscle (1). Type 2 of CCAM in a majority of cases (60%) is associated with other findings, including cardiac anomalies (truncus arteriosus, tetralogy of Fallot), renal agenesis/dysgenesis, gastrointestinal atresia, and skeletal anomalies) (1,5).

The CCAM is usually detected in the neonatal period or early childhood (2). In developing countries, a large number of children with congenital lung lesions are initially seen and treated in district hospitals before being referred to tertiary level clinical centers (6). At least half of the patients diagnosed with CCAM antenatally are asymptomatic at birth (5). Because of the risk of infection and of malignant transformation, most authors recommend resection of all antenatally diagnosed CCAMs, although the surgery can often be deferred until several months after birth (5). All removed tissue should be examined histologically (6).

Our patients had had repeated slowly regressive pneumonia scanned by radiographs and computed tomography and displayed as incorrect tumor like changes in the posterior basal parts lungs. This experience obliges to consider pulmonary sequestration as a real differential-diagnostic possibility in patients with localized repeated bronchopneumonias.

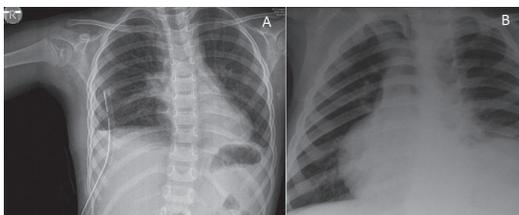


Figure 2. A) Missing right basal lung lobe and chest drain, B) Reversal of organs, total; missing left basal lung lobe (Selimović A, 2015)

These patients should be referred to a tertiary care center, where they can be diagnosed and treated early, so as to prevent the morbidity and mortality associated with late resection of the lesion (6).

Prognosis of CCAM depends on the size of the lesion, the degree to which the remaining lung is underdeveloped, and the presence of other associated congenital anomalies (7). Preoperative imaging studies are essential for assessing not only the congenital lung disease but also the anatomy of vessels, bronchi and fissures (7).

The VATS lobectomy in children is technically more challenging than in adults because maintaining successful single-lung ventilation in children is often difficult due to relatively small thoracic cavities and the narrow intercostal spaces in children (8).

Thoracoscopic lobectomy for congenital pulmonary airway malformation has been indicated from the neonatal period to adolescence (8). However, it is difficult to approach the pulmonary artery for lobectomy in congenital lung malformations with incomplete or absent interlobar fissure (8).

The treatment consists of identifying and division of the aberrant feeding vessel and usually the lobectomy of the affected lobe. However, some authors describe VATS sublobar and wedge resections when possible (9,10). The aberrant systemic artery can be freed and dissected safely with VATS approach despite the frequently occurring inflammatory changes (11).

After the division of the anomalous artery, the lower pulmonary vein must be dissected but care should be made to ensure that a superior pulmonary vein exists, and there is no common pulmo-

nary vein (12). Most of the authors describe the VATS approach with three to four incisions (12). The operation may be successfully carried out making only two incisions (12).

There are few reports of VATS approach for pulmonary sequestration. Kestenholz et al. reported on an analysis of 14 thoroscopically treated pulmonary sequestrations in a single institution with good postoperative results (13).

Compared with the conventional posterolateral thoracotomy approach, the most important step during resection of PS via VATS is the identification of the aberrant artery. The aberrant artery may be thickened or fragile because of recurrent infections. If an unanticipated injury to the aberrant artery occurs bleeding cannot be effectively managed because of high blood pressure (14).

Children with chronic pneumonia and failure of single-lung ventilation have independent risk factors for conversion to thoracotomy (7). A minithoracotomy combined with video assistance performed predominantly via direct visualization was a secure, integrated, minimally invasive approach to the large fused fissure and the severe adhesion (3).

In conclusion, VATS lobectomy is an alternative to the traditional thoracotomy for the treatment of CCAM and PS, but VATS lobectomy should be investigated in the future for its safety and effectiveness.

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TRANSPARENCY DECLARATION

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Video-asistirana torakoskopska lobektomija kod kongenitalne cistične adenomatoidne malformacije

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SAŽETAK

Cilj Prikazati rijetki slučaj urođenih malformacija disajnih puteva (CCAM) i način njihovog hirurškog liječenja s video-asistiranom torakoskopskom hirurgijom (VATS).

Metode Dva muška djeteta i jedno žensko dijete, u dobi od 7, 4 i 3 godine, liječeni su zbog simptoma kašlja i povišene temperature u regionalnoj bolnici. Kod sve troje djece urađena su laboratorijska ispitivanja krvi i radiografija grudnog koša. Auskultatorni nalazi su pokazali prisutnost upale pluća. Djeca su liječena odgovarajućim dozama antibiotika. Nakon izliječenja upale, transportovani su do Kliničkog centra Univerziteta u Sarajevu, gdje je nakon kompjuterizirane tomografije (CT) urađena video-asistirana torakoskopska lobektomija (VATS).

Rezultati CT grudnog koša je pokazao CCAM i plućnu sekvestraciju (PS), te je bila potrebna lobektomija da se odstrani dio zahvaćenog plućnog tkiva. Kod dvoje djece s PS-om aberantna krvna žila poticala je od proksimalnog dijela trbušne aorte, dok treće dijete nije imalo PS.

Zaključak VATS lobektomija je alternativna metoda za tradicionalnu lobektomiju za liječenje CCAM-a i PS-a, ali će njena sigurnost i učinkovitost u budućnosti biti predmet istraživanja.

Ključne riječi: plućne malformacije, torakoskopska hirurgija, plućna sekvestracija, cistično adenomatoidna