Lung Cysts Following Pulmonary Artery Operations: Diagnostic and Therapeutic Implications

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ABSTRACT

We describe the case of a child suffering from congenital cyanotic heart disease — double outlet right ventricle (DORV) with transposition of great vessels (TOGV). She underwent a left Blalock-Tausig (BT) shunt at one month of age followed by a Glenn procedure with left pulmonary artery augmentation at six months. Following the second procedure she developed extensive cyst formation in the upper lobe of the left lung and pneumothorax. She was managed by intercostal drainage of the pneumothorax. The cysts were observed and on a CT scan X-rays taken at one month and six months no cysts were seen. This case illustrates the occurrence of pneumatoceles after pulmonary artery manipulation, their proclivity for causing pneumothoraces and involution on follow-up. Cysts noted in such a setting should be monitored carefully and followed up to resolution.

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Key words: Lung cysts; Children; Post- infarction; After cardiac operations, Pulmonary artery cystadenomatoid Malformations (CAMs), pulmonary sequestration, bronchogenic cysts and congenital lobar over inflation are the usual causes of cystic lung lesions in childhood. These conditions may present early in life, often with cystic air filled lesions in the periphery of the lung, and, usually require surgical treatment.

Acquired condition may also cause cyst formation in the lungs. Cystic fibrosis, infections like staphylococcal pneumonia, pulmonary contusion and hydrocarbon poisoning are all cited in this context.

This report describes the unique case of a child who underwent extensive, unilateral cystic changes in a lung following operation for a complex, congenital, cyanotic cardiac anomaly.

REPORT OF CASE

H.A.A. was the product of a cesarean section done at term, indicated for meconium staining. Soon after birth she was noted to have cyanosis. A chest X-ray showed oligemia of both the lung fields with no otherparenchymal normal lung lesions. The ECG showed normal sinus rhythm with left axis deviation.

An echocardiogram was done which revealed a secundum atrial septal defect (ASD) of 3 mm, large inlet ventricular septal defect (VSD), double outlet right ventricle (DORV), transposition of great vessels, mild hypoplasia of the right ventricle, atresia of the pulmonary valve and hypoplasia of the pulmonary arteries. The right pulmonary artery could not be made out. The aortic arch was right sided with a small patent ductus arteriosus (PDA). She was started on prostaglandin E1 infusion and referred to another centre for management of the cardiac anomaly. Here she underwent a left modified Blalock Tausig shunt (BT shunt), using a 4 mm goretex graft and ligation of the PDA. At 6 months of age the BT shunt was taken down and a bi-directional Glenn shunt and left pulmonary artery (LPA) augmentation were done. Except for a superficial wound infection after the second operation, recovery from the two operations was uneventful. At discharge, the parents were informed of the appearance of cysts in her left lung. These cysts had not been seen on a chest X-ray taken after the first operation. At home she continued to progress well, until 2 months after the second operation, she presented to our hospital with sudden onset of respiratory distress and cyanosis. On examination she had decreased air entry on the left side. The oxygen saturation was 50-60%. A chest X-ray revealed a left pneumo-thorax which was treated by insertion of intercostal- drainage (ICD) tube. A post ICD tube insertion chest X-ray and chest CT scan (Fig.1 and 2) showed extensive cystic changes in the left upper lobe. The general condition and oxygen saturation of the
congenital lobar emphysema and bronchogenic cysts usually have a lining epithelium of respiratory origin. Acquired causes like necrotizing pneumonia, hydrocarbon ingestion (e.g., kerosene, furniture polish and lighter fluid) and trauma can also lead to cyst formation. The mechanism for the development of the cysts from these acquired causes has as its basis lung injury at the alveolar level leading to destruction of the lining epithelium of the alveoli. This leads to a breakdown of the dividing walls with formation of larger cavities which are supplied by air channels in which injury has caused edema and thereby a check-valve mechanism. These cavities may thus overinflate and appear as air filled cysts on chest X-rays.

Subpleural cysts resulting from pulmonary artery occlusion have been described by Stocker et al. in an autopsy study of three children. In their series, occlusion of the right pulmonary artery in a newborn infant produced peripheral (subpleural) infarction of the lung and cyst formation. Another child had undergone an operation for correction of an ASD and PDA. The authors suggest that pulmonary arterial occlusion may have been responsible for cyst formation in all three cases and may produce cysts in other infants that lead to idiopathic spontaneous pneumothorax in older children and young adults.

We suggest that LPA occlusion during the augmentation procedure in our case led to necrosis and destruction of alveoli. These alveoli supplied by distal air passages which were injured in the process and were edematous, therefore acted like check valves. This mechanism, as in the case of pneumonias and poisoning leading to cysts is difficult to prove by direct methods. Chronology of events, lack of other precipitating causes, and the site of appearance of the cysts all point to a cause and effect relationship between the operation and the cyst formation. In this case the cysts were seen only after the second operation, there

DISCUSSION

In childhood, lung cysts can result from both congenital and acquired conditions. Congenital causes like CAMs,