CASE REPORT

Hybrid Lesion of Congenital Cystic Adenomatoid Malformation and Bronchopulmonary Sequestration

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ABSTRACT

Hybrid congenital cystic lesions of lung comprising of both congenital cystic adenomatoid malformation (CCAM) and broncho-pulmonary sequestration (BPS) are very rare congenital malformations of the lung. We present a case of a newborn who presented to us with severe respiratory distress and later on found to have a hybrid lesion of the lung. It is a very rare occurrence and no such case has been reported from Pakistan so far.

Key Words: *Congenital cystic adenomatoid malformation (CCAM). Broncho-pulmonary sequestration (BPS). Hybrid lung lesions.*

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) and broncho-pulmonary sequestration (BPS) are rare congenital cystic malformations of lung. Occurrence of these lesions together in hybrid form is even rarer.[1] These lesions usually present in neonatal period with respiratory distress and may be associated with hydrops.[2] They are more commonly found in males.[3]

If undiagnosed in neonatal period, they can lead to repeated chest infections.[4] CCAM is rarely associated with other congenital anomalies; however, BPS is frequently associated with other congenital anomalies such as congenital heart disease, congenital diaphragmatic hernia, lung hypoplasia, and cystic lesions of kidney and liver.[3]

Here, we present a case of a neonate with congenital cystic hybrid lesions of the lung.

CASE REPORT

A baby boy, born through spontaneous vertex delivery at home, was brought to Neonatal Intensive Care Unit (NICU) on his first postnatal day with complaints of breathing difficulty since birth, which aggravated after trial of feed. According to parents, baby was unable to tolerate oral feeds and was constantly grunting since birth. Baby passed meconium after 6 hours of birth. There was no history of vomiting, lethargy or fits.

The baby was third child of a non-consanguineous marriage. Birth history was unremarkable. Mother took routine vitamin supplements and also underwent anomaly scan at 20 weeks, which was unremarkable. There was no history of pregnancy-induced hypertension, diabetes mellitus or prolonged rupture of membranes.

On examination, baby had severe respiratory distress with nasal flaring, subcostal recessions, and tachypnea with respiratory rate >70 breaths /min. He was cyanosed and oxygen saturation on room air was up to 80%. He was afebrile with a heart rate of 165 bpm. There was no edema, jaundice or pallor. His blood sugar at the time of presentation was 119 mg/dl.

His chest was bilaterally clear with no crackles. However, there was decreased air entry on left side. Apex beat was shifted towards midline. First and second heart sounds were audible with a loud P2. No added heart sound was heard. Rest of the systemic examination was unremarkable.

Baby was immediately placed on Continuous Positive Airway Pressure (CPAP) and started on first line IV antibiotics and IV fluids, according to unit protocol. His septic screen was normal.

Chest X-ray (CXR) showed a large mass occupying most of the left lung field with a well demarcated dome shaped upper border and a mediastinal shift towards right side (Figure 1). 2D-Echocardiography showed evidence of moderate pulmonary hypertension with shunting through small patent foramen ovale.

The clinical features and investigations mimicked presentation of congenital diaphragmatic hernia.

Paediatric surgery review was sought. Since CT scan of chest could not be performed due to non-availability of portable mechanical ventilator; baby's portable ultrasonography abdomen and chest was done. However, ultrasonography chest did not provide any leading clue. Surgeons proceeded for thoracotomy with probable plan of diaphragmatic hernia repair. Per-operatively, diaphragmatic rent was noted and repaired accordingly. Postoperatively, baby was placed on mechanical ventilation.

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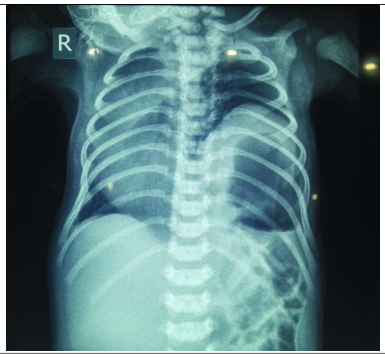

Figure 1: A large mass accupying left lung.

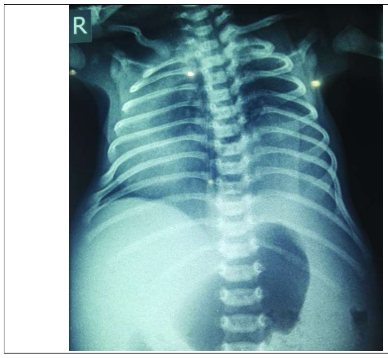

Figure 2: Mediastinal shift persisting in lower zone.

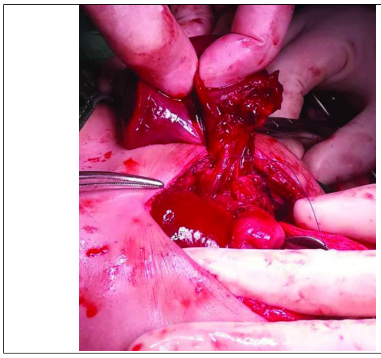

Figure 3: Extra-pulmonary tissue on base of left lung.

Baby still required high pressures for maintaining saturation. Postoperative CXR was also not encouraging. Mediastinal shift as seen in earlier x-ray was persisting with a consolidation in lower zone of left lung (Figure 2). Three differentials were kept in mind: Re-eventration; Sequestration; and Consolidation. On 11th day of life, baby underwent diagnostic laparoscopy and exploratory laparotomy. Peroperatively, extra-pulmonary tissue on base of left lung was found without any connection with terminal bronchioles. Its arterial blood supply was derived from abdominal aorta and venous drainage by an aberrant pulmonary vein (Figure 3). This lung tissue was excised and sent for histopathology. Based on the blood supply, a provisional diagnosis of extralobar BPS was made.

Postoperatively, baby was again placed on mechanical ventilation on Pressure-Controlled Ventilation (PCV) mode. However, this time the postoperative CXR showed a marked improvement with right lung collapse. Clinically baby was also comfortable. Gradually, the pressures were tapered and baby was weaned off from mechanical ventilation on 3rd postoperative day. Chest physiotherapy and nebulisation were also started.

Baby's sepsis markers deteriorated initially, so he was shifted to second line antibiotics. Small volume oro-gastric feed was also started on third postoperative day which, when tolerated well, was gradually increased in amounts.

On 15th postoperative day, he was weaned off from oxygen support with no apparent respiratory distress.

Histopathology report, however, showed findings consistent with CCAM of left lung tissue, with no evidence of malignancy.

His ultrasonography abdomen and pelvis was done to rule out other associated anomalies and was found to be unremarkable.

On the basis of histopathological findings being consistent with CCAM and the aberrant blood supply from aorta which favored sequestration, a final diagnosis of hybrid lesion of BPS and CCAM was made.

DISCUSSION

During fetal lung development, certain insults may lead to formation of abnormal cystic lung masses.

CCAM is a rare cystic congenital malformation of lung. Its incidence is of the order of 1 in 25,000 to 35,000 live births. CCAM is a hamartomatous change in the tertiary bronchioles which leads to formation of cysts in the fetal lung. It contains tissues from various pulmonary origins and is more often unilateral.[3,4]

BPS is also a cystic lung lesion which occurs in general population with an incidence of 0.15 - 1.7%. It is defined as a mass of lung parenchyma that lacks a normal connection to the tracheobronchial tree, thus a non-functioning lung tissue which possesses an anomalous systemic arterial blood supply.[5] It occurs more often on left side.

More rarely, a few cases of congenital cystic lesions have been reported as being hybrid of both the CCAM and BPS. In our case report, we present a similar rare congenital cystic lung lesion in a neonate found on left lung base as an extra-lobar sequestration with systemic arterial blood supply but having a histological appearance of CCAM. No local data on such cases has been reported previously up to date.

CCAM receives arterial supply from pulmonary vasculature, whereas BPS receives arterial supply from aorta. CCAM tissue has connections with the tracheobronchial tree, whereas pulmonary sequestration does not connect with the bronchioles.[6]

The incidence of hybrid cases of CCAM and BPS is rare, with only 36 such cases reported in English literature so far.[7]

A study conducted in Taiwan showed an incidence of 19% of mixed type hybrid lesions from 1995 to 2008. They highlighted the fact that the increasing proportion of such lesions may suggest a similar developmental origin.[1]

Both the lesions have malignant potential. CCAM accounts for 25% of all congenital malformations and 95% of all congenital lung lesions.[8]

To conclude, very few hybrid cases of CCAM and BPS have been reported. One should keep a high index of suspicion in order to diagnose such lesions in any neonate who presents with severe respiratory distress in neonatal life or recurrent chest infections outside neonatal period.

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