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Original Research Paper

Paediatrics

A RARE CONGENITAL LUNG MALFORMATION – BRONCHOPULMONARY SEQUESTRATION.

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ABSTRACT Pulmonary sequestration (PS) is a rare congential malformation of the lower respiratory tract that is the lung parenchyma. It refers to region of lung parenchyma which lacks a normal connection to the tracheobronchial tree and possesses an anomalous systemic blood supply, usually from aorta or its major branches. Large pulmonary sequestrations can present in the newborn with potentially fatal respiratory distress.
We present a case report of a day old male child born to primigravida mother with extralobar type.

KEYWORDS : pulmonary sequestration, lung parenchyma, computer tomography

INTRODUCTION

Pulmonary sequestrations refers to region of lung parenchyma which lacks a normal connection to the tracheobronchial tree and possesses an anomalous systemic blood supply, usually from aorta or its major branches(1). PS is classified as " extralobar" or intralobar" position according to the absence or presence of investing pleura. Both forms derive their blood supply from the systemic circulation. It was estimated that 6.4% of all congenital pulmonary anomalies were pulmonary(2). We report a case of extralobar pulmonary sequestration in a newborn.

CASE REPORT

One day old male neonate full term normal vaginal delivery first by birth order born of non-consangnious marriage cried immediately after birth with ABGAR of (8,9,9) born to 19 years of primi mother was referred to our hospital with antenatal anomaly scan at 5th month suggestive of bilateral lung fields echogenic, there are few subcentimetric sized cysts seen in left lung largest measuring 20 mm in the lower zone suggestive of congenital cystic adenomatoid malformation or pulmonary sequestration, baby was admitted in NICU kept under observation, he had mild respiratory distress since birth, no difficulty in feeding was there. Heart rate was 120 per minute, respiratory rate 62 per minute with DOWNE's score of 1along with mild intercostal retractions, grunting nasal flaring cyanosis was not there , AF- open and at level 2.5x 2.5 centimeter(cm), Head circumference- 35 cm. Length- 53 cm, both at 50th centile. NIBP 86/65/71 mmHg , SPO_2 98% in all four limbs , capillary refilling time < 3 sec, peripheral pulses well felt. Respiratory System examination air entry equal on both sides , no adventitious sounds heard. Chest radiograph showed a left lower zone mass(figure 1). His symptoms subsided with oxygen 21% by hood for 12 hours. High resolution computer tomography (HRCT) was done suggestive of enhancing soft tissue density lesion in left lower lobe with no communication with bronchus and separate arterial supply and venous drainage suggestive of pulmonary sequestration(figure 2). For further classification Magnetic Resonance Angiogram was planned. Patient was referred to pediatric surgery department for left lower lobectomy but they asked parents for follow up as there were no symptoms at that time.

DISCUSSION

Pulmonary sequestration or accessory lung is a congenital abnormality in which a portion of the lung shows separation from the normal bronchial tree and blood supply. The estimated incidence was 0.15% to 1.7% in general population(1). Extralobar sequestrations are enclosed in their own pleura, typically receive arterial flow from small branches of the thoracic or abdominal aorta, they are associated with accessory spleen, complex heart diseases and diaphragmatic hernias(3) . Intralobar sequestration are enclosed in the pleura of the adjacent normal lobe. Their arterial supply is usually aortic branches and drainage into pulmonary veins(4). The most common presentation of pulmonary sequestration is recurrent pneumonia followed by heart failure, hemoptysis, respiratory distress. Computed tomography usually shows irregular cystic component in the lesion and there is high incidence of emphysema in the adjacent lung due to collateral air drift and air trapping. The most specific pre-operative diagnosis rests on the identification of a systemic arterial supply.

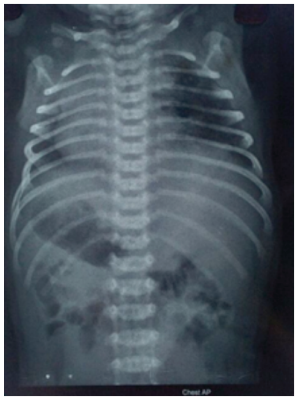

Figure 1. Chest x ray showing left lower zone consolidation.

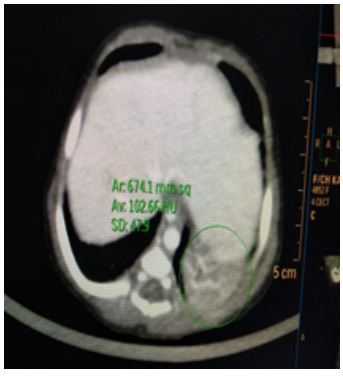

Figure 2. HRCTH

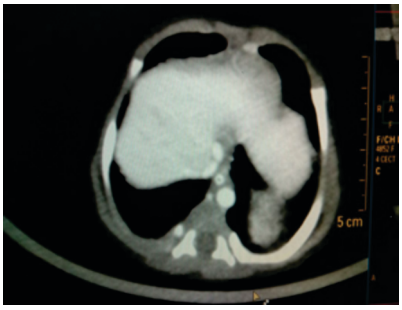

Figure 2. HRCT

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