Rare Presentation of Congenital Cystic Adenomatoid Malformation of the Lung

Sharma A[1], Pannu MS[2], Lata N[3], Dhillon SPS[4], Singh N[5], Sareen A[6]

[1]Dr. Anubha Sharma, MBBS, MD, Senior Resident, [2]Dr. MS Pannu,MBBS, MD, DTCD, Professor and HoD Paediatrics, [3]Dr Neeraj Lata, MBBS, DCH, DNB, Senior Resident, [4]Dr. SPS Dhillon, MBBS, MD, Assistant Professor, [5]Dr. Narinder Singh, MBBS, DCH, MD, Assistant Professor, [6]Dr Ashwini Sareen, MBBS, MD, DM, Associate Professor, All from the department of Paediatrics, Government Medical College, Amritsar, India.

Address for correspondence:
Dr. Anubha Sharma
E-mail: anubha.khera@yahoo.in

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Abstract

Congenital cystic adenomatoid malformation is a rare entity presenting with respiratory difficulty in newborns and even later in infancy. Various cases have been reported but recurrent spontaneous pneumothorax in an infant with failure to thrive and pectus carinatum since birth(with uneventful antenatal history and asymptomatic course in the first seven months of life) is an extremely rare presentation of this congenital lung condition.

Key words: CCAM, CPAM, Pneumothorax

Introduction

Congenital cystic adenomatoid malformation(CCAM) is a pulmonary congenital anomaly and consists of hamartomatous or dysplastic lung tissue mixed with normal lung; it is usually confined to one lobe (incidence is 1-4/100,000)[1]. It is a rare abnormality which stems from abnormal embryogenesis[2]. The fundamental pathological feature of the lesion is adenomatoid proliferation of bronchioles that form cysts at the expense of normal alveoli. There is arrested alveolar development associated with the proliferation of terminal bronchioles in the affected lobe.

CCAM or congenital pulmonary airway malformation(CPAM) is usually discovered in neonates because of respiratory distress and may occasionally be diagnosed in infancy and older children/adults who us usually present with recurrent chest infections[3].

The pathophysiological effects may be divided into prenatal and postnatal effects. Forty percent cases may present as hydrops foetalis antenatally which is thought to be due to inferior vena cava compression and is a poor prognostic sign[4]. Polyhydramnios can also occur due to elevated intrathoracic pressure which leads to oesophageal compression and inability to swallow[5].

This entity may remain undiagnosed until it is discovered incidentally in later life, however it presents as respiratory distress in the newborn period due to pulmonary hypoplasia/mediastinal shift/ spontaneous pneumothorax /pleural effusion following hydrops. Sarcomatous and carcinomatous differentiation has been described in patients with CPAM/CCAM.[1,6]

The Case

We present a rare case report of an eight months old male infant who presented to us with cough since one month, fever off and on since one month and difficulty in breathing since the past three days. This was the first time he fell ill after birth. His weight was 5 kg, length was 70 cms.

His heart rate was 130/min, respiratory rate 60/min, SpO_2 95% in room air and the peripheral pulses were palpable at the time of admission. On examination, there was pectus carinatum (since birth) and there was decreased air entry in the right infraclavicular, mammary, inframammary and axillary areas. The cardiovascular system examination was normal and there was no hepatosplenomegaly. Hb was 8gm%, TLC: 5800/mm^3, DLC: P_{70}, L_{30}, M_0, E_0 and PBF revealed a dimorphic picture with anisopoikilocytosis. Mantoux showed no reaction, ESR 15mm/hr and gastric lavage for AFB was negative. Immediate chest X-ray showed right sided pneumothorax with mediastinal shift to the left (tension pneumothorax). Diagnosing the case to be tension pneumothorax.

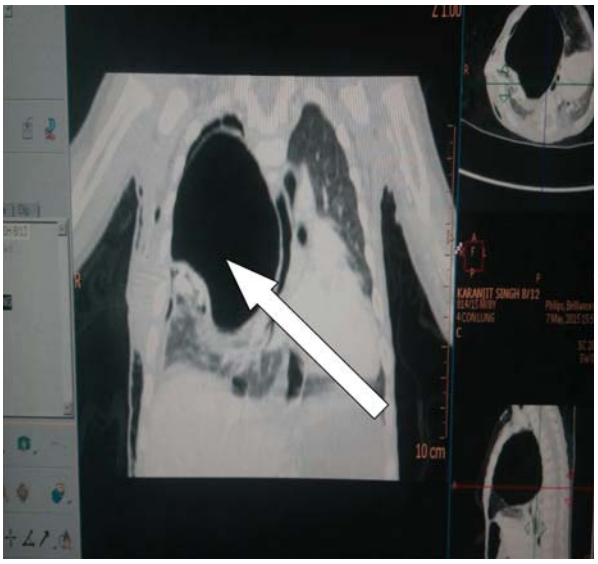

Fig 1: CT Scan coronal ct section of the chest showing CCAM/CPAM type 1, large cystic area in the right upper lobe indenting upon the horizontal fissure. Also seen are the small lung parenchymatous cystic lesions surrounding this large cyst.

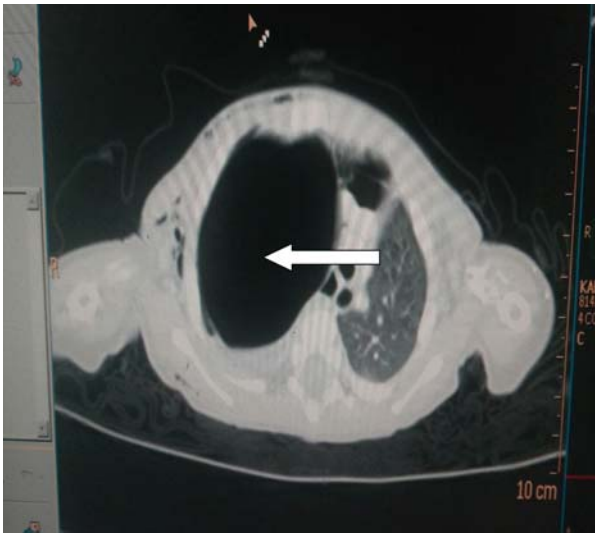

Fig 2: CT Scan, axial section of the chest showing a large cystic lesion in the right upper lobe with the evidence of air in the subcutaneous tissue suggestive of subcutaneous emphysema.

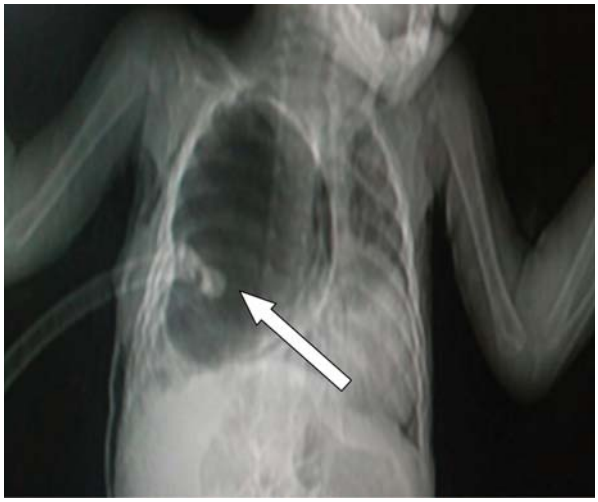

Fig 3: X Ray chest AP view at admission showing a large cystic lesion is seen in the right hemithorax displacing the mediastinum to the right side. Intercostal chest drain in situ. The condition was suspected to be tension pneumothorax, however on removal of the drain no change could be appreciated. This was later proved to be CCAM type1.

Chest tube was inserted along with intravenous antibiotics but the pneumothorax was temporarily relieved and reoccurred once the tube was accidentally pulled out. Reinsertion of intercostal tube was done but the respiratory distress persisted and the X-ray again showed tension pneumothorax. (The child also developed subcutaneous emphysema which slowly resolved on its own). So a CT Scan was ordered which showed thin walled cysts (>2cm) in the right upper and middle lobes with minimal fluid and reactionary pleural effusion suggestive of type I CCAM. The child was thus referred to higher centre for further management with intercostal chest tube in situ.

Discussion

CCAM is a congenital disorder of the lung in which an entire lobe of a lung is replaced by a non working cystic piece of lung tissue. This can be diagnosed in

utero by ultrasonography[7]. The fetus can have hydrops, pleural effusion, polyhydramnios, ascites and severe facial edema. Lesions causing hydrops have a poor prognosis but lesions appearing large in early gestation may regress in size later on and be associated with better pulmonary function later on[1]. Patients in newborn period and infancy present with respiratory distress, recurrent respiratory infections and pneumothorax. Breath sounds can be diminished with mediastinal shift to the opposite side. Differential diagnosis of lung abscess and diaphragmatic hernia should always be kept. Complications like fetal death, premature delivery, pneumonia, hemothorax and malignant potential should not be ignored. CPAM can be divided into four types on the basis of clinical and pathological features[8]. Type I is the most common (50%) with the best prognosis and consists of a single cyst or multiple large cysts (>2cm) lined by pseudostratified ciliated columnar epithelium. The cyst walls are thick and contain smooth muscle and elastic tissue. Type II (40%) has many smaller cysts <1cm. Type III forms a solid bulky mass with adenomatoid changes and lined by cuboidal epithelium (10%).It has a poor prognosis. Type 0 is incompatible with life[9].

Pulmonary resection during infancy can have better outcome along with lower chances of malignant changes[10]. The treatment is always surgical in symptomatic patients and the mortality is generally less than 10% [1]. Lobectomy is usually necessary but segmental resection is occasionally feasible[11].

Conclusion

So a possibility of congenital pulmonary airway malformation should always be kept in infants who present with tension pneumothorax which transiently gets relieved by intercostal drainage and recurs once the tube is pulled put.

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