Cystic adenomatoid malformation of the lung: A diagnostic dilemma

Anup Mohta, Ravi P. Kanojia, Sapna Bathla[1], Nita Khurana[2]

ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) of the lung is an uncommon anomaly that arises from excessive disorganised proliferation of tubular bronchial structures excluding the alveoli. These are believed to represent focal pulmonary dysplasia because skeletal muscle may be identified from within the cyst wall. This report describes a case of an infant operated for presumed diagnosis of congenital diaphragmatic hernia. Diaphragm was found to be normal and further investigations revealed cystic mass in the lower lobe of the left lung. Thoracotomy was done to resect the lesion that revealed a type II CCAM on histopathological examination. The case stresses the need for better clinical examination and advanced radiological investigations in doubtful cases.

Key words: Adenomatoid malformation, congenital diaphragmatic hernia, infant

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INTRODUCTION

Congenital cystic adenomatoid malformations of the lung (CCAM) are uncommon congenital cystic lung lesions that arise from excessive disorganised proliferation of tubular bronchial structures excluding the alveoli.[1] Preoperative diagnosis of this condition can be difficult at times, as it can be confused with other surgical causes of respiratory distress in the infants.[2] This case report describes a case of congenital adenomatoid malformation erroneously diagnosed as congenital diaphragmatic hernia.

CASE REPORT

A two-month-old boy presented with respiratory distress of two months duration. He was born at full term after normal vaginal delivery out of nonconsanguineous marriage. There was no antenatal examination of the mother for any congenital anomalies; and there was no fever. Clinical examination revealed signs of respiratory distress and chest examination revealed decreased air entry on the left side. Plain X-ray chest [Figure 1] was interpreted as that of left congenital diaphragmatic hernia. The patient was referred to a surgeon who operated with a presumptive diagnosis of congenital diaphragmatic hernia (CDH) by abdominal route. At surgery, the diaphragm and abdominal structures were found to be normal, and the incision was closed. In the postoperative period, the child continued to have respiratory distress. The child was referred for further investigations and management. Computed tomography of the chest [Figure 2] suggested a diagnosis of cystic disease of the left lower lobe of the lung. Left lower lobectomy was performed through left thoracotomy. The child improved clinically in the postoperative period. Signs of respiratory distress disappeared and the air entry in the left chest improved. Postoperative chest X-ray improved without any abnormality. Histopathological examination of the excised specimen suggested Type II congenital cystic adenomatoid malformation.

DISCUSSION

Respiratory distress in the infants due to surgical causes is uncommon. Surgical conditions leading to respiratory

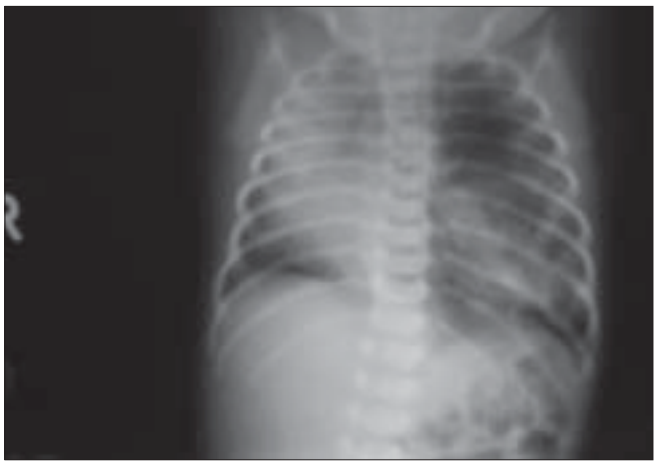


Figure 1: Plain X-ray chest diagnosed as congenital diaphragmatic hernia

Departments of Paediatric Surgery and [1]Anaesthesiology, Chacha Nehru Bal Chikitsalaya and Maulana Azad Medical College, Delhi, [2]Pathology, Maulana Azad Medical College, New Delhi, India

Address for correspondence:
Dr. Anup Mohta,
28-B, Pocket-C, S.F. S. flats, Mayur Vihar Phase-III,
Delhi - 110 096, India. E-mail: mohtaanup@hotmail.com

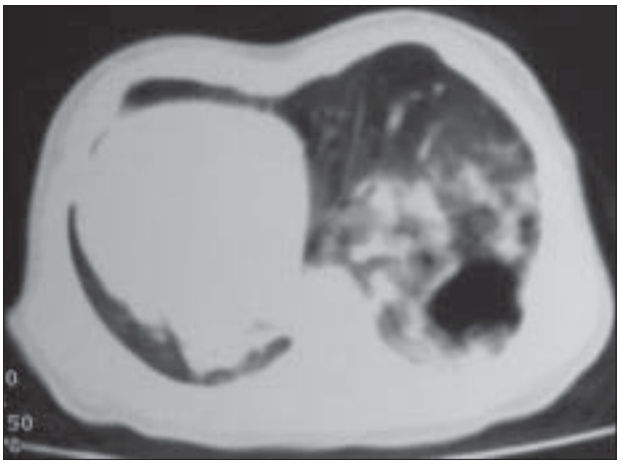


Figure 2: Computed tomography showing cystic lesion in the lung

distress include congenial lobar emphysema (CLE), congenital cystic adenomatoid malformation (CCAM), and pulmonary sequestration.[1,3] Diaphragmatic defects such as CDH or eventration and bronchogenic cysts; and acquired lung pathology such as empyema or lung abscess can also present with distress.

Usually CDH can be diagnosed on the basis of clinical examination and chest X-ray. The abdominal gas pattern in the abdomen should suggest the clinician, whether the patient is dealing with a case of CDH or cystic lung disease. There are reports which suggest that there may be difficulty in differentiating CCAM from CLE, or CDH may be diagnosed as CCAM.[2,4] Sometimes, CCAM may coexist with CDH or sequestration.[2] In doubtful cases, a high resolution CT[5] or MRI helps in confirming the diagnosis, demonstrating unsuspected findings, with better depiction of anatomic extent of thoracic anomalies, thus allowing better planning for surgery. It is necessary to confirm the diagnosis before surgery to plan approach.[6,7]

CCAM is the second most common congenital lung lesion in children. It is more common in males and is the result of an embryologic insult before the 50th day of gestation causing maldevelopment of the terminal bronchiolar structures. Stocker[8] described different types of CCAM on histopathology. Although, it is said to sometimes disappear before birth, in-utero regression of CCAM on antenatal ultrasound may not represent genuine resolution and need fetal MRI for prenatal diagnosis. [9] Postnatal computed tomography of thorax should be considered in all newborns with antenatally diagnosed congenital cystic adenomatoid malformation; and if confirmed, early operation before first hospital discharge is advised to prevent progressive lung hypoplasia.[10,11]

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