Case report:

Congenital cystic adenoid malformation of lung

Dr Suhas Kumbhar

Professor, Department of Pediatrics, Bharati Vidyapeeth Medical College, Sangli, India

Corresponding author: Dr Suhas Kumbhar

Abstract

Congenital cystic lesions of the lung in children is uncommon but potentially life threatening and warrants urgent diagnostic work up. Congenital Cystic Adenoid malformation is one of the major congenital cystic lesions of the lung. This causes recurrent lung infections in children. Recently we could successfully diagnose and treat such rare case in our institute.

A 5 year old male child presented with history of one episode of hemoptysis and fever, cough, breathlessness since last one month. Parents gave history of recurrent LRTI since 6 months of age and theirs is a 2nd degree consanguineous marriage. For these complaints he was consulted and treated on OPD basis several times. Child also received indoor treatment for six times with temporary relief. On examination child was febrile; tachypnea, pallor and grade 2 clubbing were present. Child has moderate under nutrition. On respiratory system examination there was dull note on percussion and air entry reduced with crepitation in left lower zone.

Child was investigated X-ray chest showed large cavity 4 by 4 cm in left lower lobe with air fluid level. In spite of IV antibiotics for 14 days X-ray chest showed persistent cavity in left lower lobe. So CT scan was done which showed multiple abscesses with one large cavity 4cm in left lower lobe. With history and investigations chronic suppurative lung disease was diagnosed and referred to pediatric surgeon. The child underwent left lung lower lobectomy and the specimen was sent for histopathological analysis. Section showed large cysts with surrounded small cysts. The findings were consistent with TYPE 1 CCAM of lung. Surgery was uneventful. Child required 2 bags of PCV and discharged on 16th post-operative day. At present the child is coming for regular follow up and growing well and doesn’t require any hospital admission.

So in cases of recurrent LRTI with detail history taking and careful examination we can suspect congenital lesions like CCAM of the lung which can be confirmed by radio imaging techniques. With prompt surgical intervention definitely we can expect good outcome.

Key words – Congenital, malformation, cystic lesion, Lung disease

Introduction

Congenital cystic lesions of the lung in children is uncommon but potentially life threatening and warrants urgent diagnostic work up. Congenital Cystic Adenoid Malformation (CCAM), pulmonary sequestration, congenital lobar emphysema and bronchogenic pulmonary cysts are the major congenital cystic lesions of the lung. Of this CCAM is the commonest cause of recurrent lung infections in children. We diagnosed and successfully treated such rare case of CCAM of lung in 5 year old child in our institute.

Case Presentation

A 5 year old male child residing in a nearby village born of 2nd degree consanguineous marriage was admitted in this hospital with chief complaints of one episode of hemoptysis and fever, cough, breathlessness since last one month. Parents gave history of recurrent LRTI since 6 months of his age. For these complaints he was treated on OPD basis several times by general practitioners. He received indoor treatment in private nursing homes six times, every time he was investigated and treated for 5-10 days with injectable antibiotics.
with temporary improvements in symptoms. Child also received 6 months of antituberculous drug but no relief although child didn’t have close contact with known tuberculosis case.

In view of persistent fever and breathlessness child was referred to this institute for diagnosis and treatment. There was no History of FBA, CHD, chest pain, jaundice other congenital malformation. Birth history was normal, immunized up to date and the child have mild motor delay. On examination child was febrile and has tachypnea, pallor and grade 2 clubbing. Child also has moderate undernutrition. On Respiratory system examination, there was dull note on percussion and air entry reduced with crepitation in left lower zone.

Child was investigated thoroughly. CBC showed Total Leucocyte Count 24200 with polymorphs 81%, Hb 9 gms, ESR was persistently high 120 and 90. HIV and HBSAG were negative. Blood culture sterile and induced sputum for AFB and GRAM staining was negative. X-ray chest showed large cavity 4 by 4 cm in left lower lobe with air fluid level. In spite of IV antibiotics for 14 days X-ray chest showed persistent cavity in left lower lobe. So CT scan was done which showed multiple abscesses with one large cavity 4 cm in left lower lobe.

With the help of history, clinical examination and investigation findings child was diagnosed as a case of chronic suppurative lung disease. Child was referred to pediatric surgeon for consultation. In view of chronic lung abscess with multiple small cavities confining to left lower lobe. The child underwent left lung lower lobectomy and the specimen was sent for histopathological analysis. Section showed large cysts with surrounded small cysts. It was ulcerated and showed granulation tissue and multinucleated giant cells. Smaller cysts were lined by columnar epithelium with numerous foamy histocytes. At places compressed lung tissue was visible. There was diffuse infiltration by mononucleated cells and polymorphs in interstitial tissue.

These histopathological findings were suggestive of TYPE 1 CCAM of lung and it was confirmed.
Post operative lung specimen.

Surgery was uneventful. Child required 2 bags of PCV and discharged on 16th post operative day. At present the child is coming for regular follow up and growing well and episodes of LRTI significantly reduced and doesn’t require any hospital admission.

**Discussion:**

CCAM of lung is a rare but commonest surgical condition which present with repeated LRTI in infancy and childhood. CCAM consists of hamartomatous or dysplastic lung tissue mixed with normal lung and generally confined to one lobe. Its incidence is 1 to 4/100000 live births. CCAM was first described by Chin and Tang in 1949 and referred to group of several pathological entities characterized by architecturally abnormal lung tissue with or without cyst formation. 

CCAM probably results from a cessation of bronchial maturation and concomitant overgrowth of mesenchymal elements which produce the adenomatous appearance of the anomaly in the early stage of development. CCAM usually presents in infancy with respiratory distress, repeated LRTI, pneumothorax or persistent pulmonary infection. Smaller lesions may be asymptomatic upto mid childhood and may present with recurrent LRTI. Histologically, cartilage is absent suggestive of bronchiolar maldevelopment.

**TYPE 1 :-** Macrocystic (50%) single or several large cysts lined with pseudostratified epithelium and mucous secreting cells. Prognosis is good with long term survival but they are prone for mucin secreting bronchi- alveolar carcinoma.

**TYPE 2 :-** (40%) LESION is microcystic and consists of multiple small cysts with histology similar to type 1 lesion. Type 2 is associated with other congenital anomalies and carries a poor prognosis.

**TYPE 3 :-** Lesion is solid with bronchiole like structure lined by cuboidal ciliated epithelium and separated by areas of nonciliated cuboidal epithelium. This lesion is fatal and carries poorest prognosis.

Antenatal diagnosis of CCAM can be done by USG. Postnatally it can be confirmed by CT SCAN. Pulmonary sequestration, congenital lobar emphysema, bronchogenic cysts and lung abscess are the close Differential diagnosis for CCAM. Treatment is surgical excision for symptomatic patient and mortality is less than 10 %. Resection of all CCAM is recommended in view of direct complications, such as recurrent infections and...
pneumothorax and for malignant potential in latter life. (6)

Prognosis depends upon the size of the lesion rather than type of lesion, larger the size worse the prognosis. Among the children with severe lung compromise and contralateral lung not hypoplastic, early surgical removal offers good outcome. (6)

Conclusion
In cases of recurrent LRTI with detail history taking and careful examination we can suspect congenital lesions like CCAM of the lung which can be confirmed by radio imaging techniques. With prompt surgical intervention definitely we can expect good outcome.

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References: