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Images in Clinical Medicine

Congenital Cystic Adenomatoid Malformation in a Two-Month-Old Infant

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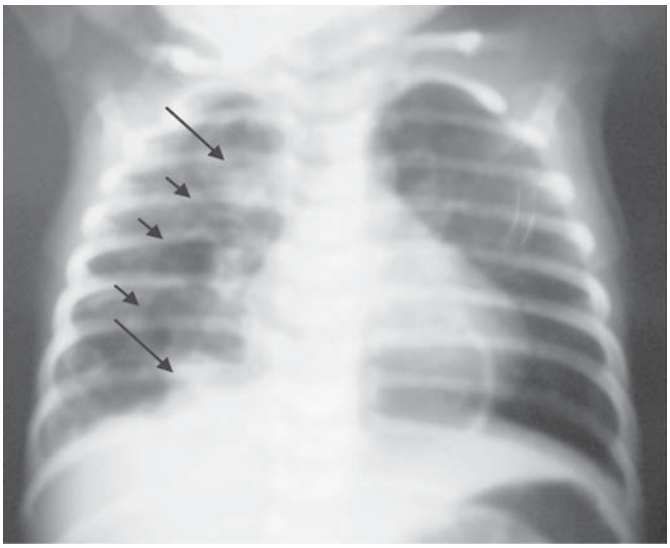

Fig 1. X-ray chest showing multiple cysts (short arrows) of varying size in right hemithorax and homogeneous opacity in the right upper and lower zones (long arrows)

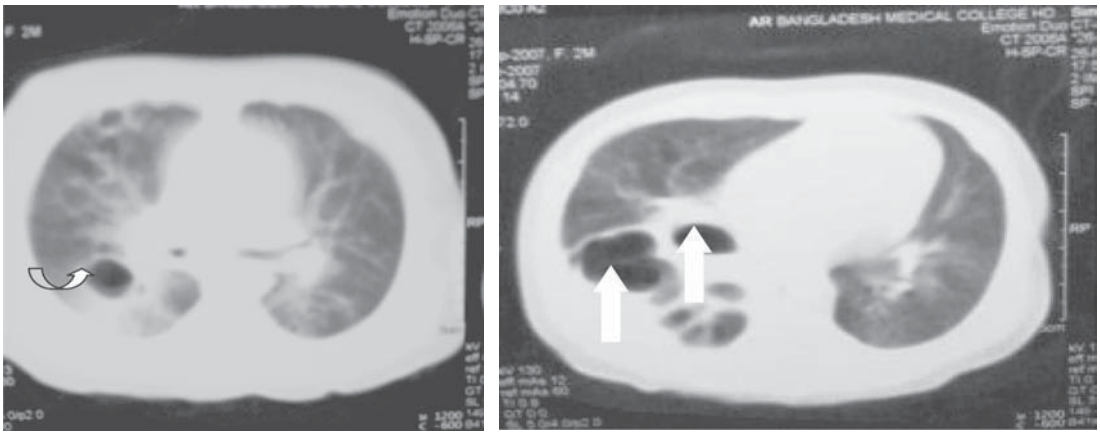

Fig 2. CT scan of chest showing multiple cystic lesions in right lung (bold arrow) with pericystic consolidation (curved arrow)

A 2-month-old infant was admitted with cough for 15 days and high grade continued fever and respiratory distress for 12 days. The baby was delivered normally at term at home by a traditional birth assistant (TBA) without any complications. She was exclusively breastfed. On admission, the baby was irritable, febrile, cyanosed, dyspneic, mildly pale and BCG mark was present. Anthropometry showed occipitofrontal circumference (OFC) 38 cm which was within normal limit, weight for age

4.3 kg [–1 SD], and height for age 55 cm [–1 SD]. There was chest indrawing, respiratory rate was 68/min and heart rate was 152 beats/min and breath sound was vesicular with coarse crepitations in both lung fields. Laboratory investigation revealed hemoglobin 9.5 gm/dL, white cell count 12,900/µL with neutrophils 75%, peripheral blood film showed anisochromic anisocytic red blood cells, mature white blood cells and few toxic changes in the lymphocytes. ESR was 35 mm in 1st hour and blood culture showed no growth. Chest radiography revealed homogeneous opacity in the right middle and lower zones with multiple cysts of various sizes in the right hemithorax (Fig 1). The diagnosis of congenital cystic adenomatous malformation (CCAM) with pneumonia was entertained. O_2 inhalation and broad spectrum antibiotics were started immediately after admission. The condition of the baby was improving with reduction of body temperature and disappearance of cyanosis. But the baby was still dyspneic with mild chest indrawing and rapid breathing on third day of admission. CT scan of the chest showed multiple cystic lesions in right lung with pericystic consolidation (Fig 2). The patient recovered uneventfully with conservative treatment on 10th day. The baby was discharged and advised to come for follow-up every month. The child was found symptom-free in monthly follow-ups till her last visit after one year and her chest radiography was normal.

CCAM was first described as a distinct entity by Ch'in and Tang in 1949.[1] CCAM is a developmental hamartomatous abnormality of the lung, with adenomatoid proliferation of cysts resembling bronchioles. It represents approximately 25% of all congenital lung lesions. In 1977, Stocker grossly classified CCAM into 3 types based mostly on cyst size.[2] Type 1 lesions are most common (50%) and composed of single or multiple large cysts, usually 2–10 cm in diameter and chance of other congenital malformations is very rare. Prognosis is excellent. Type 2 lesions are composed of smaller uniform thin walled cysts less than 1 cm in diameter, about 60% are associated with other congenital anomalies that may affect prognosis, specifically renal agenesis. Type 3 appears solid with microscopical cysts; chance of congenital anomalies is only 9%. Fetal hydrops and maternal polyhydramnios are common with this type. Patients usually present with respiratory distress, cough, fever, failure to thrive, recurrent chest infection, and dyspnea. Pulmonary hypoplasia, polyhydramnios, recurrent chest infection and pneumothorax may also be present.[3] Laboratory studies are generally not helpful in the diagnosis of CCAM. Chest radiography is essential in the workup of the child with suspected CCAM.[4] The usual appearance is of a mass containing air-filled cysts. CT scan of the thorax provides a safe and rapid means of defining the extent of CCAM in all age groups. The typical appearance is of multilocular cystic lesions with thin walls surrounded by normal lung parenchyma. The presence of superimposed infection with the lesion may complicate the appearance.[5]

The routine use of prenatal ultrasonography has led to frequent prenatal diagnosis and has provided better understanding of the natural history of CCAM. Improvement in surgical techniques along with newer imaging modalities have altered the surgical approach to this lesion.

Ishrat Jahan
Associate Professor
Department of Pediatrics
Enam Medical College & Hospital, Savar, Dhaka
Email: ishratdr@gmail.com

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