CLINICAL IMAGES

Congenital Cystic Adenomatoid Malformation

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Congenital cystic adenomatoid malformation (CCAM) is a rare abnormality of lung development. Its perinatal incidence is 1:25000 to 1:35000 (1). It presents as a cystic area within lung that is because of abnormal embryogenesis due to adenomatous overgrowth of terminal bronchiole with reduction in alveolar growth.Large cysts are associated with hydrops fetalis in 40 percent of cases. Polyhydramnios has also been associated with CCAM as elevated intrathoracic pressure leads to compression of esophagus and inability to swallow (2). Large cysts in addition compress IVC leading to decreased venous return , cardiac output and effusions.

Because of compromised pulmonary growth from cystic mass effect it result in pulmonary hypoplasia leading to post natal respiratory distress, mediastinal shift,spontaneous pneumothorax , pleural effusion . There is risk of malignant transformation in later years (3). Mortality rate is 9 to 49 percent. Bilateral lesions, CCAM with polyhydramnios have poorer out come .

Present case is 32 years old primigravida who conceived after eight years of marriage reported to gynecologist with five months of amenorrhea. Per abdominal examination revaled 24 weeks uterus and foetal heart sounds were normal, ultrasonographic examination was advised. Ultrasound examination revealed single live fetus of 21 weeks gestation, there was a large uniloculated cystic lesion in right side of thorax anterior to dorsal spine . The cystic lesion measured 33x28mm (*Fig 1 & 2*) Amniotic fluid was increased . Heart rate was 142 beats per minute and regular . Nucheal thickness was 5 mm .No ascites or pleural effusion was observed.

Mainstay of treatment is surgical intervention including foetal surgery like thoracoamniotic shunt , lobectomy and thoracocentasis . Differential diagnosis include intrapulmonary bronchogenic cyst which develop from abnormal budding of ventral foregut (called as foregut duplication cyst). It occurs later between 26 weeks to 40 weeks. However, the intrapulmonary site is rare (15%) as compared with mediastinal site (85%).

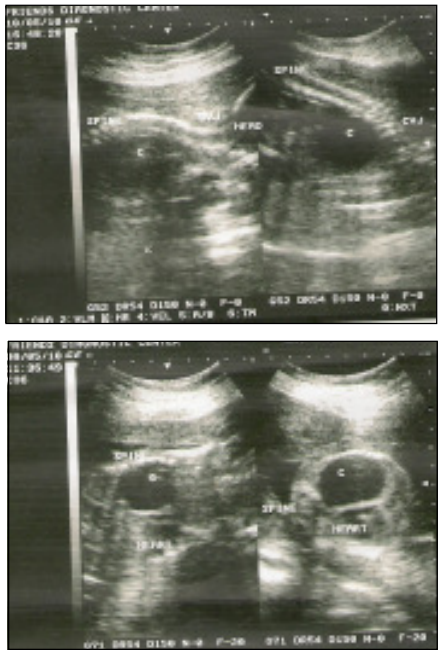

Fig.1& 2 Showing Large Uniloculated Cystic Lesion in Right Side of Thorax Anterior to Dorsal Spine (Lateral & Transverse Section)

Pulmonary sequestration another condition with may occur as cyst though commonly present as echogenic mass in lower lobes (98%) , arising from non functional primitive tissue with systemic blood supply from aorta rather than from pulmonary circulation and not communicated with tracheo-bronchial tree.

References

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