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MULTICYSTIC DISEASE OF LUNG IN CHILD AND ADOLESCENTS: A RARE DISEASE

Medical Science

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ABSTRACT

A number of diseases produce focal or multiple thin-walled or thick-walled air- or fluid-containing cysts or cavitary lung lesions in both infants and children. In infants and children, there is a spectrum of focal or multifocal cystic and cavitary lung lesions including congenital lobar emphysema, congenital cystic adenomatoid malformation, pleuropulmonary blastoma, bronchogenic cyst, pulmonary sequestration, Langerhans cell histiocytosis, airway diseases, infectious diseases (bacterial infection, fungal infection, etc.), hydatid cysts, destroid lung, and traumatic pseudocyst.

For the evaluation of cystic or cavitary lung lesion in infants and children, imaging plays an important role in accurate early diagnosis and optimal patient management. Therefore, a practical imaging approach based on the most sensitive and least invasive imaging modality in an efficient and cost-effective manner is paramount. We reviewed the conventional radiographs and computed tomography findings of the most common cystic and cavitary lung lesions in infants and children.

Clinical presentation:

A 14 year teenager from jamua, giridih presented to us with h/o repeated episode of fever, cough cold & breathing difficulty since the age of 1 yr. This type of recurrent attack of cough & cold, fast breathing affected the child at an interval of 3-4 months for which he was treated by local doctor every time with oral/ injectable medicines. The symptoms intensity gradually increases till 5 yrs of age then it gradually decreased in severity. He was referred to this hospital for diagnostic evaluation at 12 years of age.

There was no h/o blood in sputum, chest pain, swelling of abdomen & lower limbs, palpitation, decreased urine output, reddish urine, pain abdomen, bulky fatty stool, jaundice, abdominal mass, wt. loss, or Polyuria.

The child is a product of non consanguineous marriage, term normal vaginal delivery at home with uneventful postnatal period without any h/o jaundice, delayed passage of stool, abdominal distension. Out of 3 other sibling, elder sister suffers from similar type of complaints but she is has not being investigated.

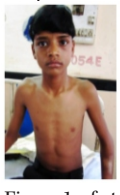

Figure 1-of pt

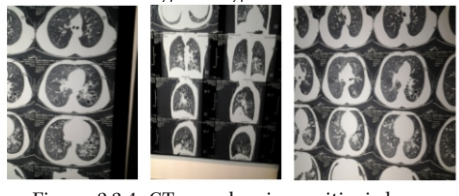

Figure -2,3,4 - CT scan showing cavities in lungs

Clinical examination:

Child is fully conscious, alert, co-operative with ht-157cm&wt-30 kg, 1st degree clubbing with burn scar on left forearm and right thumb onycomycosis. Chest ant-post elongated with trachea central and apex beat left 5th ICS on MCL. Vocal fremitus &Vocal resonance both are B/L normal. Right side of chest is distinct with presence of diffuse rhonchi and crackles left side only with crackles. Chest expansion is 4.5cm (insp-70.5cm&exp-66cm). On abdominal examination Rectus muscle bifurcation noted above umbilicus on leg rising test and soft non tender liver is palpable of about 1.5cm below costal margin in MCL. CNS findings are WNL and genitalia shows sparse pubic hair of thanner stage grd 2.

Investigations:

Routine haemogram shows no abnormality, ECG& echocardiography are also not distinctive.USG whole abdomen shows multiple mesenteric LN with largest 1.2cm and MT with 5 TU is negative. HRCT chest shows cystic and cylindrical dilatations involving left lower lobe, right middle lobe and apical region of right lung field.

Discussion:

With the above mentioned clinical profile of the patient and investigations following could be the probable diagnoses-

1. Congenital cystic adenomatoid malformation (CCAM).
2. Mutant variant of Cystic Fibrosis (CFTR class-IV/V).
3. Cystic Bronchiectasis.
4. Simple pulmonary cysts.
5. Bronchogenic cysts.
6. Bronchopulmonary sequestration.
7. Post Staphylococcal or tubercular lung condition.
8. Rarely Primary Lymphangiectasia limited to lung.

CCAM usually remained confined to one lobe of lung but in our case there is involvement of both the right & left lung with 3 separate lobes of lung . Although among all 3 types of CCAM, only type-1 appears to be a possibility as other 2 variants are usually associated with congenital anomalies. Type-2 & type-3 has poor prognosis & fatal outcome that denies their existence.

Mutant variant of (CFTR) Cystic Fibrosis may also be a possibility. But as there is absence of any symptom and sign related to Gastrointestinal or Genitourinary system since birth. The symptoms of patient has improved with conservative management. This indicates to CFTR class IV/V. The confirmation of this condition may made by the CFTR gene mutation test or Sweat Chloride test.

Cystic Bronchiectasis is the 3rd possibility of our patient's presentation which is to some extent supported by the HRCT findings. But the symptoms and sign and long standing sequel is absent in the patient.

Bronchopulmonary sequestration is another provisional diagnosis as it is usually associated with 40-50% cases of other congenital anomalies.

Bronchogenic Cyst, though more commonly right sided and near midline structure, but peripheral intrapulmonary cysts are not infrequent. This is does not fits in present context.

Post Staphylococcal lung pathology or post Tubercular lung pathology, in rare instances may behave with these types of clinical symptoms. But again any history or other related findings are absent.

Of the rare diagnosis, Primary Lymphangiectasia limited to lung only, lacks its existence with the absence of any evidence of pleural effusion or chylothorax throughout the disease process in our patient.

So finally our provisional diagnosis may be Cystic adenomatoid Malformation or Mutant variant of CFTR protein or Cystic Bronchiectasis.

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