CONGENITAL LOBAR EMPHYSEMA

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Introduction

Congenital lobar emphysema (CLE) is a rare disorder of lung development that calls for urgent diagnosis and management (1). The pathology is usually due to deficiency in cartilage of the bronchial wall (1m2m3). The usual presentation is during the 1st six months of life in most cases (1,2).Breathlessness with or without cyanosis is the common presentation. Cases of CLE are confused with other cystic malformations of the brochopulmonary tree and with pneumothorax. These cystic malformations are namely cystic adenomatoid malformation, bronchogenic cyst and pulmonary sequestration)4). The diagnosis of CLE is usually made with the help of a chest X-Ray (CXR) but it can be overlooked. The management, in most cases, is surgical excision of the affected lobe. We would like here to share with the reader our experience of successfully managing four cases.

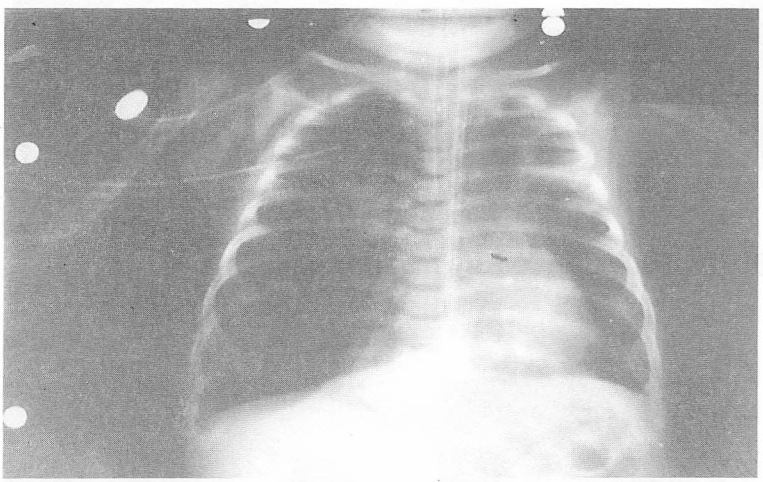
PATIENTS AND METHODS:-

The details of four patients encountered by the author over a seven years period in the children hospital of Madina Munawara were retrieved and analyzed in order to show their clinical presentation, management and outcome.

Case No.1: A six weeks boy presented with breathlessness and cough without fever. His CXR (Fig1) was initially read as pneumothorax and an intercostals tube was put to drain it. He did not improve and

a second look to the CXR revealed the features of CLE. His right lower lobe was removed surgically and he had complete recovery.

Fig No.1: Congenital lobar emphysema wrongly diagnosed as pneumothorax.



Case No.2: That was another 4 month old with a similar presentation whose CXR was typical for CLE (Fig 2). He had lobectomy for the left upper lobe.

Fig No.2: The quest for pneumothorax resulted in putting two chest tubes to drain it.

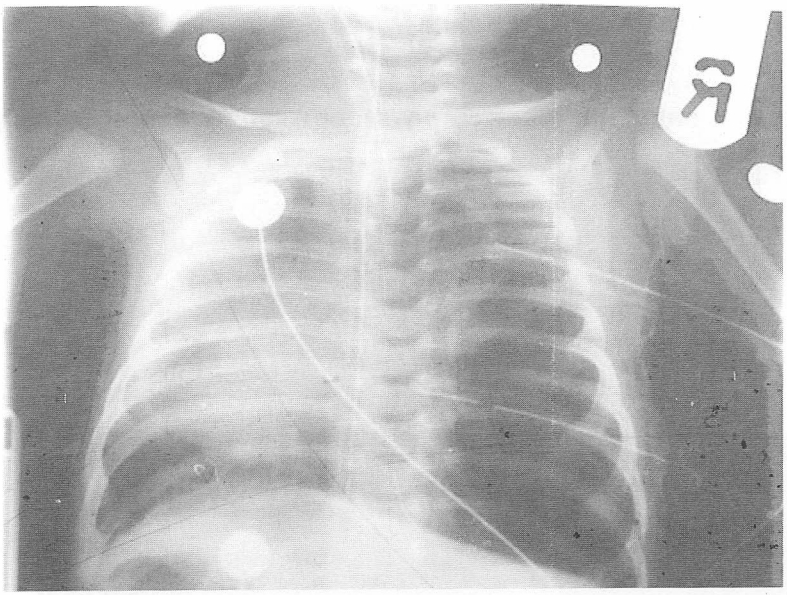
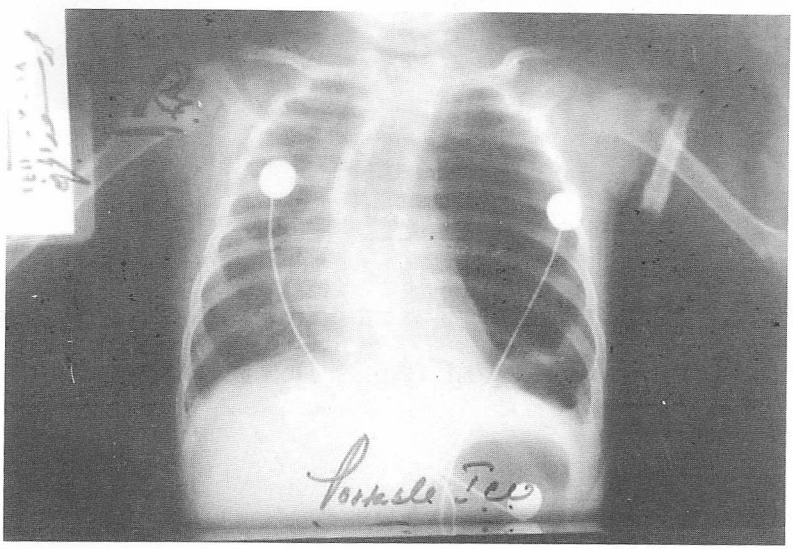
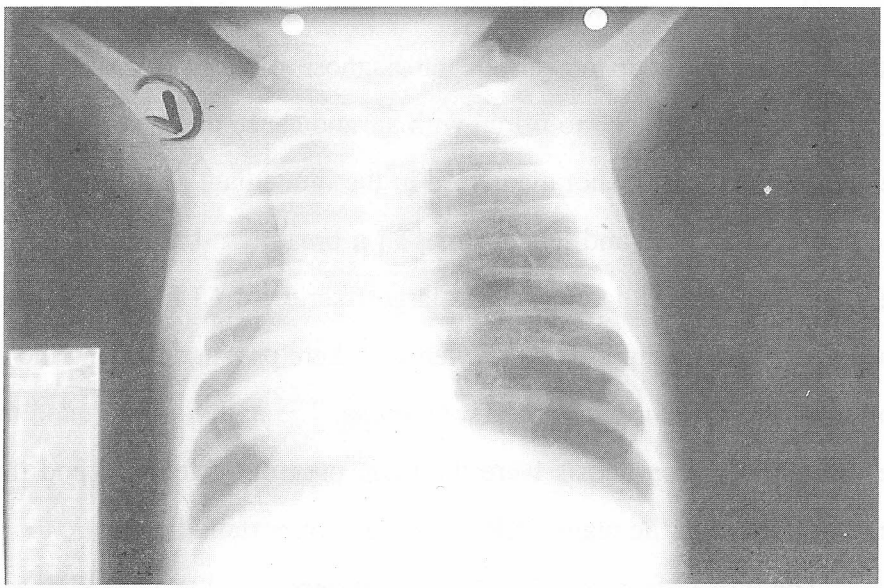


Fig No.3: Typical chest X-Ray features of CLE



Case No.3: A two weeks old girl presented with cough, breathlessness, fever and refusal of feeds. Her white blood cell count was 17000/ml with 65% polymorphs. The clinical and CXR findings favoured bronchopneumonia complicated by pneumothorax. Two chest tubes were put in an attempt to drain the presumed pneumothorax (Fig.3). Lastly CLE was diagnosed and the patient underwent lobectomy of the right lower lobe.

Fig No.4: Mild form of CLE with confusing presentation



Case No.4: That was a boy who had severe birth asphyxia and current convulsions. He was treated for recurrent chest infection since the age of two weeks. Review of his initial CXR (Fig 4) at the age of 3 months showed that he suffered from CLE of the right side from the start. Bronchospcopy and CT scan of the chest did not localize any anatomic cause for his hyperinflated upper lobe. His father did not consent for surgery. He was followed up clinically and with repeated CXR examinations. His lung condition did not deteriorate much but he died of status epileptics.

DISCUSSION:

Our experience with these cases showed that it is easy for the clinician in a busy set up to overlook the diagnosis of CLE. Because it takes time for the affected lobe to fill up with air and cause pressure symptoms the condition is usually not diagnosed in the immediate postnatal period. The absence of fever and leucocytosis in a breathless baby should alert the physician to this diagnosis. The largest series of CLE comprised 38 cases over 25 years duration. (1). Twenty of them presented during the 1st month of life and ten within the 1st 6months. The authors concluded that breathlessness and cyanosis were the main presenting features and they were satisfied with the plain CXR as the most important diagnostic tool.

The typical features, described by Stigers et al (2), were shown in the chest X-Ray of our patient No 2. These features are:

1 Increased aeration of the affected lobe with widening of the ribs interspaces.

2 Herniation to the other side of the chest

3 Mediastinal shift

4 Collapse of the adjacent lobe/s on the ipsilateral side: and

5 Flattening of the diaphragm on this side.

Other diagnostic imaging involves CT scanning and ventilation perfusion studies: but they rank second to the plain CXR. Bronchoscopy is only helpful if stenosis of the feeding bronchus is suspected (1, 2, and 3). Prenatal ultrasonography can help in diagnosis of cystic adenomatoid malformation and pulmonary sequestration but it is of limited value for CLE (5).

Treatment is usually surgical, but one can afford to follow up the mild cases albeit with the need of frequent imaging. Our decision to conserve with case No 4 was in fact partially influenced by the reluctance of his father to consent for the operation but the baby did well concerning his CLE. The long term outcome is usually good. Follow up of a series of eighteen patients over 17 years by Mikhailova and Tsareva (7) showed complete functional restoration of the lung.

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