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Bilateral congenital cystic adenomatiod malformation of the lungs : a case report

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Summary
A six weeks old infant presented with scalp ulcer and fever, and on examination was found to have resonant percussion notes bilaterally. The initial chest radiograph revealed multiple lucencies which were initially thought to be due to diaphragmatic hernia, but the dilemma was resolved by Computerised tomography which revealed the lucencies to be multiple cysts characteristics of Congenital Cystic Adenomatiod Malformation (CCAM) type II.

Keywords: *Congenital cystic adenomatiod malformation, radiological diagnoses,*

Résumé
Un bébé de six semaines avait l'ulcère du crâne et la fièvre. A l'examination clinique, il a été observe qu'il avait des notes de résonance de percussion bilatérale. Un examen de la radio revelait des multiples traces qui étaient initialement pense due a l'hernie diaphragmatique. Le delimme était résolu par la tomographie informatisée qui revelait que ces traces étaient des kystes caractéristique d'une malformation cystique adendritique congénitale de type II.

Introduction

Congenital cystic adenomatiod malformation (CCAM) is a developmental hamartomatous abnormality of the lung with adenomatiod proliferation of cysts resembling bronchioles. Ch'in and Tang first described this disease entity in 1949. CCAM represents approximately 25% of all congenital lung lesions [1]. Although the cause is not known, CCAM is believed to result from focal arrest in fetal lung development before the seventh week of gestation secondary to a variety of pulmonary insults. Most infants born with CCAM have no symptoms at birth. Many women in the developing world do not have access to routine prenatal ultrasound scan; therefore CCAM is rarely diagnosed de novo except accidentally as in this case report; as an extensive literature search had not revealed any reported case of CCAM in this environment.

Case report

A.O was a female infant admitted at six weeks of birth with scalp ulcer and fever of twelve and ten days duration respectively. The patient was well until about two weeks before presentation when the mother noticed that the baby was having low-grade fever and two days later noticed an area of scalp discolouration over the occipital region, which was discharging purulent materials with associated sloughing of the overlying skin. She had received treatment at home with antibiotics and other unknown drugs given by the mother. The baby was born at term and the pre- and post-natal history was essentially normal with no ingestion of any unusual drug during pregnancy by the mother. On physical examination, patient was found to be acutely ill, pale, anicteric, acyanosed and dehydrated. In addition she had bilateral pitting pedal oedema up to the ankle, worse on the left and a tender swelling on the right middle finger. She had a scalp ulcer, extending from the right parieto-temporal region across the midline involving the whole occipital region, with surrounding necrotic skin. The floor of the ulcer was hard, with an island of bone at the centre measuring approximately 6cm x 5cm.

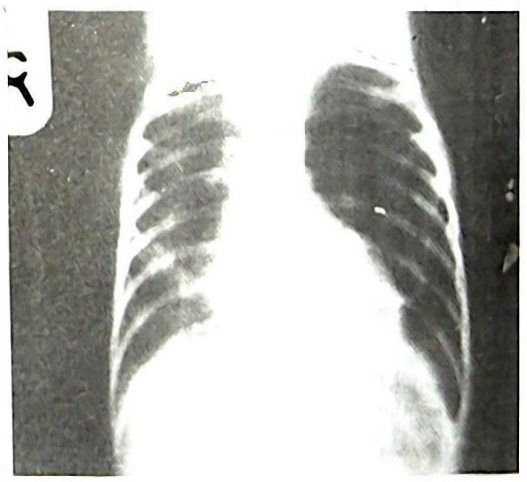

Fig. 1:

Chest percussion was resonant bilaterally with vesicular breath sounds; the pulse was irregular at 164/minute. There was no cardiac murmur. The antero-posterior chest radiograph showed bilateral areas of large cystic lesions, devoid of lung markings with surrounding septated walls one of which showed a fluid level on the left upper zone. The lateral film showed these large cystic lesions to be mostly posterior, suggesting a diaphragmatic hernia (Fig. 1). However a limited barium meal

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study revealed no abnormality of the gastrointestinal system. Computerised tomography of the chest showed multiple well-defined cystic lesions of varying sizes in both lung fields, with the largest measuring 4.2 x 3.5 cm (Fig. 2). Based on the overall radiographic appearances a diagnosis of congenital cystic adenomatiod malformation type II was made.

The patient however died before any surgical resection could be performed.

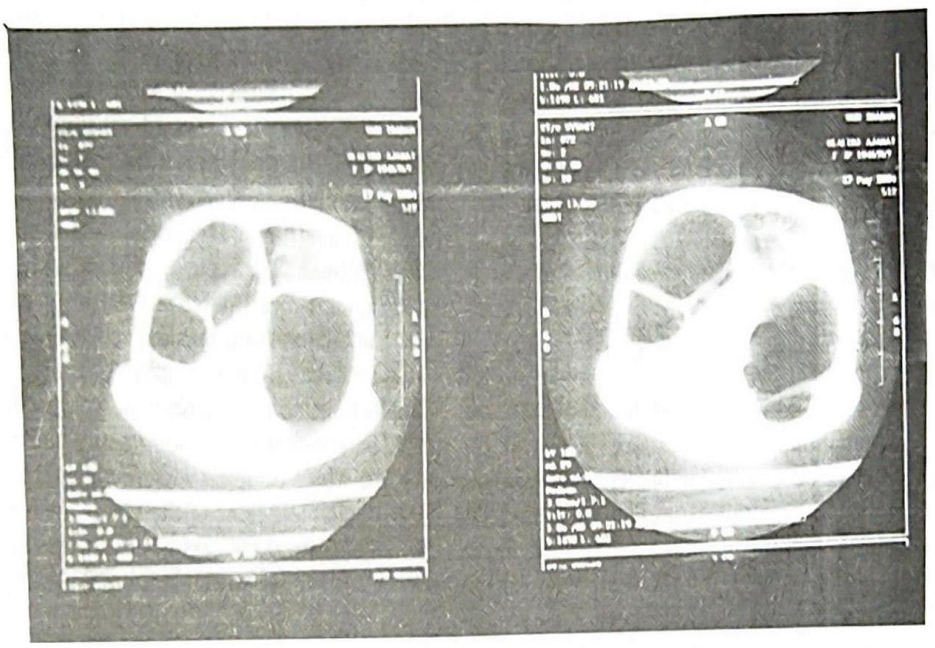

Fig. 2.:

Discussion

Congenital cystic adenomatiod malformation (CCAM) is a developmental hamartomatous abnormality of the lung with adenomatiod proliferation of cysts resembling bronchioles, and accounts for approximately 25% of all congenital lungs [1]. Stocker classified congenital cystic adenomatiod malformation (CCAM) into three types based on clinical, gross and microscopic criteria. Type I lesion is composed of single or multiple large cysts (more than 2 cm. in diameter), frequently producing mediastinal herniation. The cysts are lined by ciliated psuedostratified columnar epithelium. The walls of the cysts contain prominent smooth muscle and elastic tissue. Mucus producing cells are present in approximately one-third of the cases, and cartilage in the wall is rarely seen. Relatively normal alveoli may be seen between the cysts. The type II lesion is composed of multiple small cysts (less than 1 cm. in diameter) lined by ciliated cuboidal to columnar epithelium. Structures resembling respiratory bronchioles and distended alveoli are present between the epithelium-lined cysts. Mucous cells and cartilage are not present. Striated muscle fibres may be seen rarely. The type II lesion is associated with a high frequency of other congenital anomalies, and the prognosis is poor [2]. The type III lesion is a large, bulky non-cystic lesion producing mediastinal shift. Bronchiole-like structures are lined by ciliated cuboidal epithelium and separated by masses of alveolus-sized structures lined by non-ciliated cuboidal epithelium [3, 4].

Majority of patients present as respiratory emergencies in the newborn although the disease may remain undetected for some years. Diagnosis is usually made prenatally using ultrasound, and postnatally using chest radiograph and computerised tomography (CT). On chest radiograph, the malformation is usually composed of air filled cysts of varying sizes which may contain fluid levels, while on CT, air filled cysts of varying sizes which may contain fluid are seen [5- 8], and these were demonstrated in our patient Figs 1 and 2. In the study done by Habbard *et al*, [9] MRI is useful as adjunct to prenatal diagnosis of CCAM thus allowing for choice of treatment and parental counselling. Khosa J.K, *et al* in their study also concluded that CT scanning is mandatory for postnatal evaluation because chest radiograph could be normal [10]. The major diagnostic differential is diaphragmatic hernia, which can produce a similar multicystic appearance on plain chest x-ray, however in CCAM there is normal bowel gas pattern [5].

Due to an increasing trend in the detection of asymptomatic antenatally diagnosed CCAM, consideration is given to early surgical excision to prevent complications and recurrence [9]. Prognosis is based on findings of prenatal ultrasound and it has been found that microcystic lesion, bilateral lung involvement and hydrops were each highly correlated with poor prognosis.

while neither polyhydramnios nor mediastinal shift was significantly associated with bad outcome [4].

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