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A CASE REPORT ON RIGHT LUNG APLASIA: A RARE CONGENITAL ANOMALY

Pathology

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ABSTRACT

Pulmonary agenesis is a difficult to diagnose rare congenital condition representing failure of development of the primitive lung bud. It can have a varied age and clinical presentation. Here we present a case of a 6 month old female who was admitted with complaints of severe respiratory distress and cough. She was initially suspected as a case of right lung collapse, but the patient was eventually diagnosed to have right lung aplasia with vertebral segmentation anomaly of T8 vertebrae following a contrast enhanced tomography of the chest.

KEYWORDS

right lung agenesis, aplasia, CT scan of chest, vertebral segmentation anomaly

INTRODUCTION

Pulmonary agenesis is a rare congenital anomaly and is defined as complete absence of bronchus, parenchyma and blood vessels [1]. It differs from pulmonary aplasia by the absence of carina or bronchial stump that is seen in pulmonary aplasia. Pulmonary agenesis can be unilateral or bilateral. Bilateral pulmonary agenesis is incompatible with life [2]. The exact incidence of pulmonary agenesis is not known though estimated to vary from 0.0034% to 0.0097% [3]. Having a varied age of presentation from neonatal period to adulthood; a case of pulmonary aplasia can either be aymptomatic and diagnosed incidentally or can present with recurrent chest infections [4].

CASE REPORT

A 6 month old girl presented to our institute with the chief complaints of cough, cold and fever and fast breathing for 2-3 days. The patient was the first issue of a non consanguineous marriage. She was an institutional delivery born vaginally. The mother had no significant antenatal complaints. The first six months of the patient's life were uneventful. There was no significant family history. On clinical examination the patient had tachypnoea with signs of increased work of breathing. The chest was bilaterally symmetric with no apparent chest deformity. On percussion the note was dull in all the three lung zones on the right. On auscultation the right sided air entry was grossly reduced .The heart sounds were also better appreciated on right. The blood investigations suggested leucocytosis with neutrophilia and increased ESR. The chest X-ray (Figure 1) revealed an opaque right hemithorax with crowding of ribs and mediastinal shift to right. On the basis of clinical findings and chest skiagram a provisional diagnosis of right lung collapse was made. Ultrasonography of chest revealed absence of lung tissue on right. For confirmation of diagnosis a CECT chest (Figure 2) was obtained which showed right lung aplasia with short blind ended right main bronchus and absent right pulmonary artery .The mediastinum was shifted to right with cardiac apex pointing towards right side. The left lung was shown to be hyperinflated. Vertebral segmentation anamoly with T8 hemivertebrae was also reported. Echocardiography revealed dextrocardia with no other associated anomalies. The work up for tuberculosis did not show any evidence for the same. No other congenital anomaly was detected on USG abdomen. The patient improved on antibiotics, chest physiotherapy and supportive therapy and was subsequently discharged after 20 days of hospital stay. She was readmitted after one month with similar complaints. This time also she improved after two weeks of stay. In the second admission, parents were counseled about nebulization and oxygen support to be given at home which they have been following well.

DISCUSSION

Pulmonary agenesis/ aplasia is an extremely rare congenital anomaly being first accidentally discovered in 1673 by DePozze during an autopsy of an adult female[5].In India this clinical entity was first reported by Mohammed in the year 1923[6]. Pulmonary aplasia has been reported to occur due to disruption of normal lung development in the 4th and 5th week of gestation in the embryonic phase when normally the heart shifts to left and the bronchial analogue divides equally in two lung buds. An aberration in this process can lead to dextrocardia with failure to develop the right lung [7]. The etiological factors leading to this condition though not exactly known have been implicated to be autosomal recessive inheritance [2], vitamin A deficiency, intrauterine infections and environmental factors [3]. The incidence of unilateral lung agenesis is more on left though the right sided pulmonary aplasia has been associated with more congenital anomalies and therefore worse prognosis [2,9]. This can also be attributed to the fact that right sided pulmonary aplasia causes more mediastinal shifting and thus kinking and compression of trachea.

The age of presentation varies from neonatal cases to first presentation in old age. The clinical spectrum in patients with unilateral agenesis can vary from few symptoms and nonspecific findings to recurrent pneumonia. About 50% of these cases succumb to death before 5 years of life [9]. Pathologically pulmonary aplasia was classified by Schneider & Schwalbe which was later modified by Boyden. Its classification is as follows [10]:

Type 1 (agenesis) – Complete absence of the lung and bronchus and no vascular supply to the affected side

Type 2 (aplasia) – Rudimentary bronchus with the complete absence of pulmonary parenchyma

Type 3 (hypoplasia) – Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

The patient in concern is classified as type 2 (pulmonary aplasia).

About 50% of cases of pulmonary aplasia are associated with cardiac and non cardiac anomalies like genitourinary, oesophageal and diaphragmatic anomalies. The most common congenital anomaly associated with lung hypoplasia is hemivertebrae[11]. In a case series by Rajshekhar et al hemivertebrae was found in three out of five patients [12]. In the absence of major congenital anomalies unilateral pulmonary aplasia is often compatible with life. Though respiratory distress and recurrent respiratory infections can occur owing to pooling of secretions in blind bronchial pouch, tracheal compression by aorta in right lung aplasia and posterior curvature of trachea [7] are also the major factors contributing to the recurrence of pneumonia and non resolution of pneumonia.

No treatment is required for asymptomatic patients. Symptomatic

patients are managed conservatively on antibiotics and chest physiotherapy for drainage of secretions. Rarely surgical ligation of the blind bronchial stump may be required when accumulation of secretions leads to recurrent chest infections and non resolving pneumonia [13].

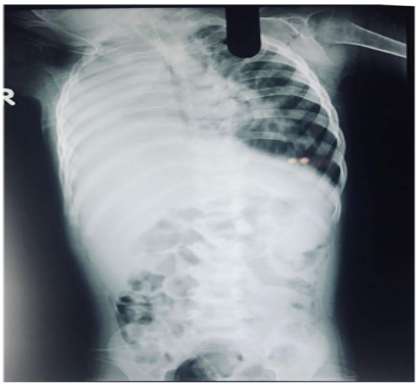

Figure 1: chest X-ray showing mediastenal shift to right with right opaque hemithorax and hyperinflated left hemithorax.

Figure2 CECT chest showing right lung aplasia with a short blind ended right main bronchus and absent right pulmonary artery and hyperinflated left lung herniatingalong anterior and posterior junctional lines.

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