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CONGENITAL PULMONARY AGENESIS AND DEXTROCARDIA

Paediatrics

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ABSTRACT

Pulmonary agenesis is often associated with anomalies in the cardiovascular, gastrointestinal, genitourinary, or musculoskeletal systems.1,2-4,5 Both lungs are equally affected but right lung agenesis shows a worse prognosis.3,5 Here we report a similar case of congenital pulmonary agenesis with dextrocardia, term born live born female baby who survived for 30 days.

KEYWORDS

Pulmonary Agenesis, Dextrocardia, Term Born.

INTRODUCTION:

Pulmonary agenesis is an extremely rare congenital anomaly defined as a complete absence of the lung parenchyma, bronchus, and pulmonary vessels.[1] This anomaly was first discovered accidentally by De Pozze (1673) during the autopsy of an adult woman.[2,3]

The estimated prevalence is 24–34 per 1,000,000 live births, and 1 per 10,000–15,000 autopsies with a slight preponderance of females.[4] The etiology remains unknown, but its pathogenesis may be associated with genetic factors, such as the duplication of the distal part of the upper arm of chromosome 2, viral factors, or vitamin A deficits.[5-6]

In normal development, the embryonic phase begins during the fourth week of gestation with the formation of the respiratory diverticulum from the ventral wall of the primitive foregut.[7] It is possible that pulmonary agenesis occurs due to the failure of the bronchial analogue to divide equally between the two lung buds. If this balance is not established, one side will develop normally while the other side will fail to-develop-and will lead to pulmonary agenesis/aplasia or pulmonary hypoplasia.[4]

Pulmonary agenesis is often associated with anomalies in the cardiovascular, gastrointestinal, genitourinary, or musculoskeletal systems.[1,2-4,5] Both lungs are equally affected but right lung agenesis shows a worse prognosis.[3,5]

Dextrocardia with situs solitus and normally related great arteries without intracardiac malformation may be present secondary to agenesis or hypoplasia of right lung. Most of the patients present with recurrent respiratory infections, cough, tachypnea, strider, wheezing, and cyanosis from an early age.[8]

CASE REPORT:

We report one such case of a female baby born with pulmonary aplasia Mrs. X primigravida an unbooked case from a lower socioeconomic status delivered a term baby at 38 weeks of gestation in view of Non progress of labor by lower segment cesarean section with an APGAR score of 8/10, and the baby cries immediately after birth, birth weight was 2.6 kg. Baby was admitted in NICU In view of respiratory distress with a suspicion of transient tachypnea of newborn (TTN), on examination she had decreased breath sounds on right half of chest , X-Ray chest was done it revealed homogenous opacity right side of chest left lung field was hyper inflated and cardiac silloute was also not visible clearly (fig 1). CT scan of Chest was ordered which revealed heart is identified on right side in region of lung, mediastinum is shifted to right, lung tissue on right side is not identified, and right main bronchus is not seen (fig 2). USG of abdomen revealed liver, stomach n both kidneys in normal position. Peripheral blood film shows macrocytic picture, Haemoglobin of 19.7, total leucocyte count of 28000/cu.mm. , platelet count – 209000/cu.mm

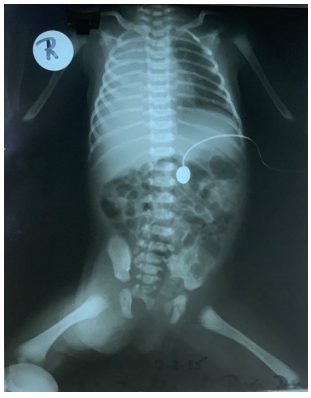

Figure 1

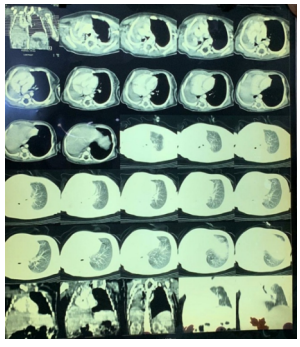

Figure 2

The baby had a hospital stay of 5 days she was maintaining saturation of 90-95% with room air, she was given BCG vaccine, she had established breast feed as well and she was discharged satisfactorily after 5 days. Baby was followed up for 15 days then we lost follow up with her but on consequent enquiry we came to know that she died on 30th day of her life.

On reviewing the literature we could find only five cases of right lung agenesis with dextrocardia.

Thus, in cases of repeated chest infections with opacification of right

hemithorax and herniation of left lung to the affected side, this rare entity must be kept in mind. It is important to investigate the coexistence of this anomaly with other entities. Asymptomatic cases do not require any treatment, if there are no additional anomalies. However this entity carries a high-risk in any surgery because of low respiratory reserve in the patient.

DISCUSSION:

Pulmonary aplasia (agenesis) is thought to result from the negative effects that occur on the 4th week of fetal life. Although its etiology is not fully understood, Vitamin A or folic acid deficiency or the use of salicylates may be responsible for it[9]. Its incidence in males and females and the occurrence of the anomaly in the right or left lung are about the same.[10]

Nearly one-third of the patients have congenital heart diseases. Although the most common one is the atrial septal defect, ventricular septal defect, patent ductus arteriosus, or aorta coarctation can also be observed[11]

In our case baby had associated dextrocardia with patent foramen ovale with minimal left to right shunt, no significant valvular or regurgitant lesion

Classification of Pulmonary Agenesis

1. Agenesis, with no lung parenchyma, bronchus, or vascular supply
2. Aplasia, absent lung tissue and pulmonary artery, rudimentary bronchus
3. Hypoplasia, rudimentary lung, and bronchial tree

Our case has complete pulmonary agenesis, i.e. type 1 variety. The presentation as TTN raises the need for keeping such a differential also in mind while dealing with such babies. The clinical presentation of the disease is quite variable. In some cases, respiratory distress syndrome is present at birth, while, in other cases, the patients remain asymptomatic until adulthood when they are incidentally diagnosed.6 More than 50% of children with lung agenesis die within 5 years of birth. However, patients with 1 lung have been known to lead healthy lives into adulthood and the oldest patient was 72 years of age when diagnosed with agenesis, as reported by Oyamada et al.[12] the reported baby however survived for 30days.

CONCLUSION:

Although pulmonary agenesis is a rare congenital anomaly, the pursuit of an accurate diagnosis is fundamental for prenatal counseling and better medical management to reduce mortality and improve prognosis.

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