CASE REPORT

A RARE CASE REPORT OF BRONCHIECTASIS WITH VACTERL ASSOCIATION

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ABSTRACT: VACTERL association is characterized by sporadic, nonrandom association of specific birth defects of multiple organ system. It includes vertebral (V), anal (A), cardiac (C), trachea-esophageal (TE), renal (R), radial limb (L), defects. Association is defined by presence of at least 3 of the above malformations. The management of patients with VACTERL association typically centers around surgical correction of specific congenital anomalies in an immediate post natal period, followed by long term medical management of its sequelae. We are reporting a rare case of bronchiectasis with vacterl anomalies.

KEYWORDS: VACTERL anomalies, Bronchiectasis, Lobaragenesis, Tracheal bronchus.

INTRODUCTION: VACTERL association is a mnemonically useful acronym for a condition characterized by sporadic, non-random association of specific birth defects of multiple organ system, described in early 1970.⁽¹⁾ It is typically defined by presence of at least 3 of following malformations– Vertebral, Anal, Cardiac, Tracheo-Esophageal fistula, Renal and radial Limb defects.⁽²⁾ Incidence is estimated at app 1 in 10,000 to 40,000 live births.⁽³⁾

CASE REPORT: An 11 yrs. old female child born to a consanguineously married couple, presented with the complaints of cough with expectoration and fever of 1 month duration. Cough was associated with copious amount of sputum which was increasing on lying to right side. Fever was of high grade and, intermittent, associated with chills and rigors. History of repeated attacks of respiratory infections were noted since 2 yrs. and was treated non-specifically.

Child was diagnosed to have VACTERL association at the time of birth. Out of the vacterl anomalies present in the child esophageal atresia with tracheoesophageal fistula and anovulval fistula were corrected in the neonatal period. Postoperative child responded well. One month after the surgery, esophagogram was performed which detected the presence of anastomotic stricture at T5 level for which endoscopic dilation was done. After regular endoscopic dilations child was taking feeds well.

2D echo of the child detected ASD for which infective endocarditis prophylaxis was advised. ASD was closed spontaneously on follow up.

There was no family history of congenital anomalies and tuberculosis.

At the time of presentation child was sick and tachypneic. Bilateral basal coarse crepitations were present more on left side. Chest X-ray revealed cystic changes in lower zones with cardiomegaly and hemivertebrae of T9 & T10.

Ultrasound abdomen revealed normal study.

HRCT chest findings were suggestive of left lower lobe bronchial stenosis, cystic bronchiectatic changes, significant peribronchial interstitial thickening in right middle lobe. Vertebral segmental anomalies in lower dorsal spine with fusion of posterior and lateral ends of right 5 and 6 ribs. Peripheral smear shows neutrophilic leucocytosis.

CASE REPORT

DISCUSSION: Bronchiectasis (Broncos, Airways; Ectasia, Dilatation) is a morphological term used to describe abnormal irreversibly dilated and often thick-walled bronchi. It has been classified into 3 main types- cylindrical, cystic, varicose.

Incidence is believed to be 1.06 to 1.3 per 100,000 population. Congenital bronchiectasis accounts to about 3-4% of cases.

Various causes reported for bronchiectasis are infections, congenital anomalies, inflammatory disorders, bronchial obstruction.

Respiratory congenital anomalies are known to occur in association with vacterlanomalies.

The commonly reported such respiratory anomalies are-bronchial stenosis, lobar agenesis, trachealbronchus, tracheal stenosis.⁽⁴⁾ The association of bronchiectasis with VACTERL anomalies is extremely rare. Our case represents this rare association.

Bronchiectasis is late pulmonary sequel in VACTERL association. It may be due to a combination of repeated aspirations secondary to TEF, cardiac anomaly-ASD and vertebral defects.

CONCLUSION: VACTERL association is characterized by sporadic, non-random association of specific birth defects of multiple organ system. The association of bronchiectasis with VACTERL anomalies is an extremely rare association. Bronchiectasis with VACTERL association will increase the morbidity when compared to isolated bronchiectasis cases.

Fig. 1: Chest X Ray PA View

Fig. 2: HRCT Chest

Fig. 3: HRCT Chest

Fig. 4: X- ray Hand and Forearm

CASE REPORT

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