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Case Report

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H-type tracheoesophageal fistula with incomplete double aortic arch in an infant

Abstract

H-type tracheoesophageal fistula and incomplete double aortic arch may result in recurrent respiratory symptoms due to communication between trachea and esophagus in former and external compression of airway in later. Though both these malformations are not very rare and are commonly diagnosed during infancy, the combination of these two is never reported in literature. The double aortic arch has important clinical and therapeutic implications especially during surgical correction of H-type tracheoesophageal fistula. Therefore, clinicians and surgeons should be aware of these vascular malformations when dealing with a case of H-type tracheoesophageal fistula. We present an 8-month-female who presented with recurrent respiratory symptoms and diagnosed to have H-type tracheoesophageal fistula and incomplete double aortic arch with atresia of the distal left arch and aortic diverticulum. H-type tracheoesophageal fistula should be suspected in infants with recurrent respiratory symptoms and radiological investigations should be focused to diagnose it and associated vascular malformations.

Keywords: H-type tracheoesophageal fistula, double aortic arch, esophagogram

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Abbreviations: TEF, H-type tracheoesophageal fistula; EA, esophageal atresia

Introduction

H-type tracheoesophageal fistula (TEF) is an uncommon malformation, accounts for 4-5% of all congenital tracheoesophageal malformations.[1,2] Incomplete double aortic arch is a rare form of complete vascular ring, encircling both the trachea and esophagus resulting in respiratory and esophageal symptoms like stridor, dyspnea, cough, and recurrent respiratory infections.[3] The association of H-type TEF and incomplete double aortic arch has never been reported in literature. This prompted us to report this rare but interesting association which has important clinical and therapeutic implications in preoperative diagnosis and planning of H-type TEF repair.

Case report

An 8-month-female presented with short history of fever, cough and rapid breathing. There was history of recurrent episodes of cough, fever, and respiratory distress since 1 month of age, which used to respond to oral antibiotics and bronchodilators. One month back, she was treated a case of severe pneumonia with intravenous antibiotics at a local hospital. Her weight was 7 kgs (10th centile), length 67 cms (25th centile) and head circumference 44 cms (50th centile). Examination revealed tachypnea, nasal flaring, subcostal retractions, and crepitations over right infraclavicular area. Investigations revealed hemoglobin 8.8 gm/dl, total leukocyte count 14400/cumm (60% polymorphs, 34% lymphocytes); platelet count 314000/cumm; and normal liver and renal functions. Chest X-ray revealed right upper lobe infiltrates Figure 1a. In view of recurrent respiratory symptoms, further investigations were done. Echocardiography was normal. Contrast esophagogram was suggestive of H-type tracheoesophageal fistula in mid-esophageal region and a narrowed segment of esophagus at same level due to external compression from posterior aspect Figures 1b & 1c. CT angiography revealed incomplete double aortic arch with atresia of left aortic arch distal to origin of left subclavian artery and aortic diverticulum Figures 2a & 2b. She was treated with and intravenous antibiotics for 10 days and planned to undergo surgery involving cardiothoracic-vascular and pediatric surgery teams but parents refused at present and will decide surgery in follow up.

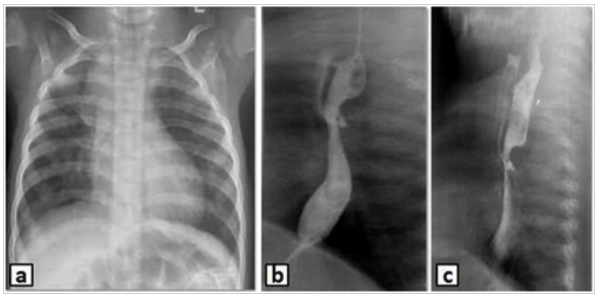

Figure 1 (a) Chest X-ray showing bilateral hyperinflation and infiltrates in right upper zone. (b and c) Contrast esophagogram showing H-type TEF in mid-esophageal region. A narrowed segment of esophagus was also seen at this level, 2 cms in length, due to external compression from posterior aspect of esophagus.

Discussion

In congenital H-type TEF, the fistulous connection between esophagus and trachea is commonly located in the lower cervical region.[2] Pressure changes between esophagus and trachea can cause entry of air into the esophagus, or entry of esophageal content into the trachea manifesting as cyanosis, cough and choking with feeding, recurrent chest infections and persistent gastrointestinal distension.[4] In our case, the diagnosis was promptly confirmed with contrast esophagogram. The primitive foregut and the paired aortic arches begin to develop at the beginning of the fourth week of embryogenesis. With the development of embryo, primitive paired vessels are formed and lost in sequential programmed fashion and any disturbance in the

involution of paired aortic arches can result in congenital vascular anomalies of aortic arch. The esophagus and trachea are derived from the primitive foregut bud and formation of foregut includes separation of the esophagus from the trachea; and incomplete formation of this septum results in TEF.[5] The time of embryogenesis of aortic arch, esophagus and trachea is overlapping, so any disturbance at this stage explains the associated anomalies of these structures.

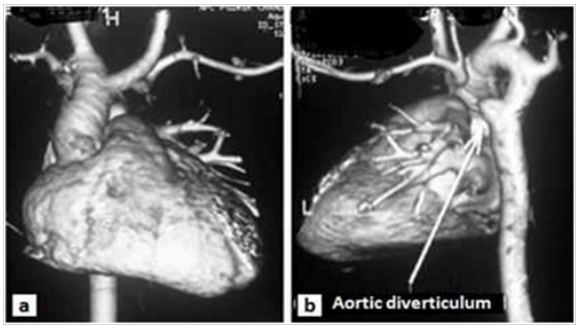

Figure 2 CT angiography (a) anterior view; (b) posterior view: Showing incomplete double aortic arch with atresia of left aortic arch distal to origin of left subclavian artery. Bilateral common carotid arteries, subclavian arteries and right vertebral artery had separate origin from corresponding aortic arch whereas left vertebral artery arises from left subclavian artery. The descending aorta is placed in midline and shows a short (4X6 mm) and round diverticulum approximately 7 mm cranial to carina. Two linear vascular channels seem to arise from the diverticulum; one channel is blind ended and other shows branches with one of the branch communicating with left pulmonary artery at 7 mm from its origin (ductus) (not shown in figure).

During the development of aortic arch, complete involution of the portion of the right arch between the right subclavian artery and the descending aorta occurs and this leads to formation of normal left aortic arch. But, sometimes the involution occurs in the same region in the left arch, and this results in a right aortic arch with mirror-image branching. Most of the times, the involution occurs between the left ductus and the descending aorta and this results in right aortic arch with mirror-image branching without a complete vascular ring (type 1 right aortic arch). Uncommonly, the involution occurs between the left subclavian artery and the left ductus and this results in a right aortic arch with mirror-image branching with a left ductus connecting a diverticulum of the descending aorta to the left pulmonary artery thus completing a vascular ring (type 2 right aortic arch).[3,6] In index patient the defect is of type 2. Esophageal atresia (EA) with TEF is commonly associated with cardiovascular anomalies and mortality is higher with these anomalies, especially those undiagnosed prior to surgery. Various studies showed that 2.5-13.1% of infants with EA/TEF have right aortic arch.[5,7–9] Thus, the patients with EA/TEF should be screened for cardiovascular anomalies prior to surgical correction. The presence of right aortic arch will determine the side of thoracotomy while surgical correction of EA/TEF and the atretic fibrous cord must be transected to relieve the symptoms associated with this complete vascular ring.

Conclusions

H-type TEF is an important treatable cause of recurrent/persistent respiratory symptoms in infants and children. The association of H-type TEF and incomplete double aortic arch, a rare vascular anomaly, is not hitherto reported in literature. The preoperative diagnosis of these co-existing defects is important for correct preoperative planning and repair of H-type TEF.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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