ISOLATED TRACHEOESOPHAGEAL FISTULA – A RARE CONGENITAL MALFORMATION

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Abstract

Introduction: Isolated congenital tracheoesophageal fistula is a very rare malformation with an incidence of 1 in 50,000-80,000 births. In most of the cases this anomaly is detected in the first year of life because of the suggestive symptoms, but sometimes the diagnosis is delayed even until adulthood.

Aim: The purpose of this paper is to analyse clinical, imagistic and therapeutic sights of this malformation.

Material and Methods: We report three cases of isolated congenital tracheoesophageal fistula, diagnosed and successfully treated in our department between 2005 and 2012.

Results: The age at time of diagnosis was 3 months, 18 months, and 3 years. The lesion was revealed by contrast radiography and endoscopy in all cases. Treatment consisted in section and ligation of the fistula. In one case the intervention was performed by right cervicotomy and in the other two by right thoracotomy. The postoperative evolution was uneventful in all cases.

Conclusion: Despite its rarity, isolated tracheoesophageal fistula should be taken into account in the presence of recurrent chest infection, associated with choking and cyanosis on feeding. The surgical strategy should be adapted to the location of the lesion.

Key words: isolated congenital tracheoesophageal fistula, diagnosis, H-tracheoesophageal fistula

Introduction

Isolated tracheoesophageal fistula represents a rare type (4-5\%) [1] of tracheoesophageal anomaly. It is frequently associated (30\%) with other malformations, including VACTERL syndrome, CHARGE syndrome, Goldenhar’s syndrome, esophageal stenosis, syndactyly [2, 3], and exceptionally with duplication cyst with or without esophageal atresia [4].

The characteristic signs and symptoms are described as the triad of Helmsworth and Pryles, which consists of coughing and aspiration during feeding, recurrent pneumopathy, and abdominal distension [2]. The early diagnosis of this disorder is difficult and some cases may remain undiagnosed until late in infancy or childhood [5]. In addition, false-negative results of all diagnostic tools are not uncommon [6]. The first surgical repair of such a defect was reported by Imperatori in 1939 [7]. Different surgical approaches have been described for this anomaly.

Case reports

Case 1

A 3 years old girl presented with persistent coughing, associated with repeated episodes of respiratory tract infections, considered related to a gastro-esophageal reflux. Her feeding problems were especially with liquids. The clinical aspect of the child was normal. A tube esophagogram was highly suggestive for the diagnosis of isolated tracheo-esophageal fistula (Figure 1).

The child was operated on through a right thoracotomy (Figure 2). Division of the tracheo-esophageal fistula was performed without any complications and the symptoms disappeared completely.
Case 2
A 18 months old male was admitted on for repeated episodes of choking and cyanosis related with feeding. The clinical features were unremarkable. Having a high index of suspicion regarding a H-type TEF, an esophagogram was performed, revealing the fistula positioned at the cervical base level (Figure 3).

The fistula was surgically divided through a right cervicotomy (figure 4). No postoperative complications were noted.

Case 3
A 3 months old was admitted in our department for severe coughing and choking at feeding. The clinical examination revealed cyanosis associated with these symptoms. The diagnosis was made by contrast esophageal X-ray and confirmed by an esophagoscopy.

We intervened by a right a thoracotomy in order to interrupt the isolated TEF. No complications in the postoperative evolution were noted.

Discussions
Isolated tracheoesophageal fistula (H-TOF) is characterized by a triad of classical signs and symptoms that are usually present from birth, but these are also nonspecific and sometimes intermittent [8].

Esophagogram is usually a reliable method for diagnosis. It is the least invasive technique for visualizing the H-type tracheoesophageal fistula, though often difficult, requiring multiple attempts before the defect is confirmed [5]. Tracheoscopy is another valuable tool, permitting the direct visualization of the fistula.

Treatment of H-TOF is surgical and should be performed as soon as possible after diagnosis [8]. For proximally located fistula the preferable approach is cervicotomy and in cases of distal fistula thoracotomy is usually performed [6]. As an alternative to thoracotomy, the thoracoscopic approach was proposed [9].

In all of our cases the diagnosis was based on esophagogram.

The surgical approach was adapted to the fistula location and it was curative.

References

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