CASE REPORT

Esophageal atresia and hypertrofic pyloric stenosis: An association to consider

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Abstract

Esophageal atresia (EA) with distal tracheosophageal fistula (TEF) develops hypertrophic pyloric stenosis (HPS) during the late postoperative period with a described prevalence of 1%-10%.The aim of this retrospective study is to show our experience in this association and to report two specific cases. Of 61 patients with EA treated in our institution, 2 developed HPS. Both cases underwent a Ramstedt extramucosal pyloromyotomy. Currently, patients are asymptomatic.

Our frequency (3.3%) is similar to other publications, about 1%-10%. It is important to be aware of this association in a patient with a history of EA and recurrent vomiting in the late postoperative period. The etiology of this association should be investigated.

Keywords

Esophageal atresia, Hypertrofic pyloric stenosis, Gastroesophageal reflux

1 Introduction

The European surveillance of congenital anomalies (EUROCAT) informs EA with TEF with a prevalence of 2.43 cases per 10,000 births [1, 2]. Its etiology is unknown, although multifactorial causes have been described and it is established that this anomaly results from a disturbance in the organogenesis. The incidence of associated congenital malformations ranges from between 40%-57% of patients, the most frequent are congenital heart diseases [2] followed by gastrointestinal anomalies such as intestinal atresia or colorectal malformations [2, 3]. After repair of (EA), symptomatic GER is frequent (close to one half), furthermore, fundoplication becomes necessary in more than 40% of this patients [6].

Moreover, infantile HPS is a common paediatric condition with an incidence of 1 in 400 live births [3, 5]. This pathology is 30 times more frequent in patients with EA [1]. This association has been reported in 1 to 10% of previous series of patients with EA [2].

2 Case reports

2.1 Case 1

A female newborn of 36 gestation weeks with an esophageal atresia, type III of Ladd´s classification. A descompressive gastrostomy was performed because the operation was rejected due to an acute cardiac failure during anesthetic induction.

After that, a feeding jejunostomy was performed on the fourth day of life. The patient underwent reconstructive surgery with primary anastomosis on the seventh day of life. The postoperative passed with feeding by jejunostomy and without complications.

The oral feeding was started after 15 days but the newborn began with not projective and postprandial vomits. GER was suspected and the patient continued being by jejunostomy with an incomplete tolerance by mouth.

Due to the lack of improvement of the condition after two more weeks, a contrast study by mouth was realized (see Figure 1). GER was refused and typical features of HPS were found. An ultrasound confirmed the diagnosis (see Figure 2).

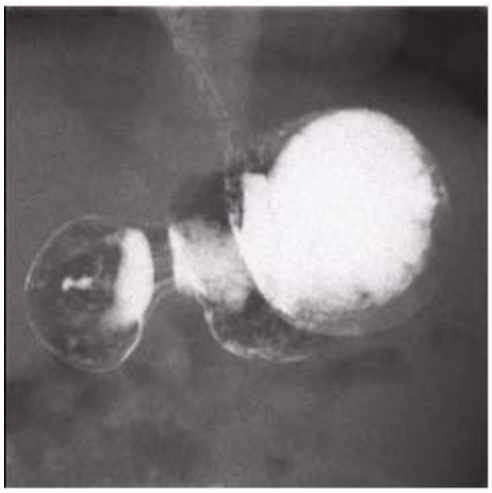

Figure 1. Typical features of HPS on an upper gastrointestinal study: Large gastric residue with increased peristalsis and delayed emptying.

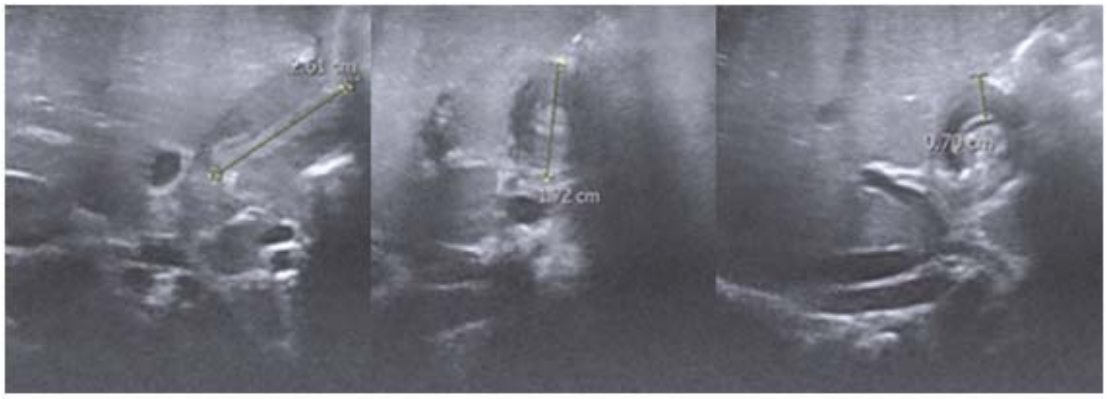

Figure 2. Measurement of pyloric canal, pyloric diameter and muscle layer with ultrasound.

A Ramstedt extramucosal pyloromyotomy was performed, the liquid feeding was started 12 hours after surgery and the jejunostomy was removed after 5 weeks.

2.2 Case 2

A female newborn of 39 gestation weeks with type III esophageal atresia and with any associated malformation. A termino-terminal esophageal anastomosis and fistula closure was performed on the second day of life without complications.

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After 3 weeks, a contrast study demonstrated a stenosis greater than 50% of the esophageal lumen, and then a dilation program was commenced.

After the first dilation oral feeding was started. The liquid feeding was continued at home for one week until the patient developed a cough and regurgitation after meals. Because of the recent diagnosis of EA with HPS, an abdominal ultrasound was performed and HPS, with 5 millimeters of muscular thickness, was confirmed.

A Ramsted extramucosal pyloromyotomy was performed on the 42nd day of life. After that, the patient tolerated the oral feeding completely and was discharged 48 hours later.

Both patients are currently asymptomatic whit a complete diet (see Figure 3).

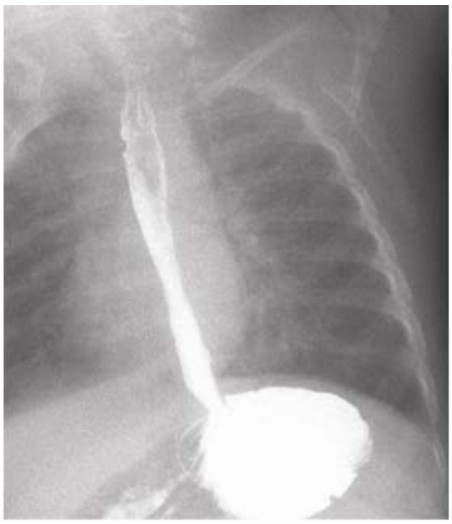

Figure 3. Normal upper gastrointestinal study of the second patient in the actuality.

3 Discussion

EA is the most common malformation of the oesophagus. These 2 cases fit into type III of Ladd´s classification (1944), that is the most common variation (86%) [1, 2].

Patients with EA have a short esophagus and dismotility, that is these patients require a follow-up for possible GER. Between 50 to 95 % of patients with repaired atresia have GER symptoms such as dysphagia and airway diseases, and could need a medical, or even surgical treatment [6].

HPS is a common surgical cause of vomiting. The etiology of isolated HPS is still unknown. Mean age at time of admission of HPS is 5.4 weeks and the typical presentation is progressive, projective, postprandial and non-bilious vomiting [1, 2], the majority of cases are diagnosed with ultrasonography.

It is necessary to perform a differential diagnosis in an infant with recurrent vomiting and regurgitation after reparation of EA. In the first case reported, too much time was spent to obtain a diagnosis, but due to the recent case we were able to suspect promptly HPS in the second newborn and the ultrasonography was requested at the appropriate time.

HPS is associated with many clinical syndromes but the occurrence simultaneously of EA and HPS is reported in the literature with a prevalence of 1%-10%; it is important to suspect it [2]. In 1986, Magilner reported 2 cases of this

association and reviewed 35 reported cases. Since these reports, less than 20 cases have been reported in literature, we can realize that the delayed diagnostic is common [3, 5].

Now, we report our experience. We have done a review based on the medical histories. There have been 61 patients with EA admitted in our department between January 2004 and December 2013. Two patients developed HPS, a prevalence of 3.3%, which is 30 times higher than HPS in the normal population.

4 Conclusion

In conclusion, it is important to make a differential diagnosis, HPS included, in infants with vomiting after surgery repair of EA. A prompt recognition of any anomaly is sometimes decisive in these patients in order to avoid problems such as anastomosis failure or respiratory complications.

Actually, in our protocol, we include an ultrasonography in all patients with vomiting and inadequate tolerance after 3 weeks after EA repair. The reason for this strong association should be investigated.

Conflict of interest statement

All authors disclose any financial and personal relationships with other people or organizations that could inappropriately influence their work.

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