Clinical Case

Congenital esophageal stenosis diagnosed at 10 months of age

Ricardo Jorge Hernández-Herrera¹, María Guadalupe Salas-Rubio¹, Felipe Javier Rodríguez-Herrera¹, and Juan Ramón Cepeda-García²

Abstract

Background. Congenital esophageal stenosis (CES) occurs in 1/25-50,000 live births. It is a rare diagnosed and must be differentiated from gastroesophageal reflux disease (GERD).

Case report. We present the case of a 10-month-old female with vomiting and regurgitations since the first day of life. The patient was treated as having GERD. She was administered different infant formulas including elemental formulas. On three occasions she had imminent bronchial aspiration. The patient’s weight was normal because she received a diluted diet and elemental formula until 10 months of age. Esophagogram and endoscopy confirmed CES diagnosis. Surgery was done after the second endoscopic dilatation failure. End-to-end anastomosis was done at 11 months. Histopathological findings reported fibromuscular type of CES.

Conclusion. CES is a rare entity with a frequently delayed diagnosis. Patients are often treated according to diagnosis of GERD until intolerance to semisolid diet appears and CES is suspected. Esophagogram and esophageal endoscopy followed by balloon dilatation prior to end-to-end anastomosis is the treatment of choice.

Key words: congenital esophageal stenosis, delayed diagnosis.

Introduction

Congenital esophageal stenosis (CES) is an extremely rare disease and presents with postprandial vomiting beginning in the first hours of life.¹ Frequent vomiting or regurgitation are key symptoms to suspect an abnormality of the esophagus. These usually appear almost immediately after food intake and are confused with gastro-esophageal reflux disease (GERD), a pathology that affects 2-7% of Western populations.¹,² Some signs that should guide the clinician to an early diagnosis are difficulty in food ingestion and dysphagia, especially when the diet changes from breastfeeding or commercialized milk to solid foods,³ along with repeated signs of choking during food ingestion and during the following minutes. When esophageal abnormalities present a blockage such as in H-type esophageal atresia (2-5%), they may be associated with esophageal stenosis.⁴ There are three types of CES: fibromuscular, membranous and tracheobronchial remnant. After histologically confirming the variety, therapeutic behavior is then defined.⁵ It is considered that the first two types respond well to balloon dilatation.

CES should not be ruled out by first intention in a patient who initiates with vomiting during the neonatal period. It must be differentiated from poor feeding technique, reflux disease, esophagitis, intolerance to milk protein or achalasia in

¹Pediatra; ²Cirujano Pediatra, Servicio Médico del Municipio de San Pedro Garza García, Hospital OCA, Monterrey, Nuevo León, México

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older children. In CES, intolerance to solid or semi-solid foods occurs at the beginning of weaning. In GERD, this intolerance is more frequent with a more insidious onset and then decreases during the first year of life. It is present in 87% of patients at 2 months of age, in 70% at 4 months of age, in 45% at 6 months of age, in 22% at 8 months of age and in 7% at 12 months of age. Intolerance to milk protein (≤1%) is even less frequent. Although esophageal stenosis is itself associated with GERD, it is related to poor weight gain.

Clinical case report

A female infant was seen for the first time at 10 months of age due to the presence of recurrent postprandial vomiting since birth. The infant was treated for gastroesophageal reflux and milk intolerance. She was the product of a third, full-term gestation culminating in a cesarean delivery due to placenta previa. The product was a single, live female infant with a birthweight of 3 kg. She did not display choking behavior and had an early discharge with the mother. There is no history of inherited significant or pathological problems associated with her condition. She was breastfed for 1 month, and from the first day of life has suffered from regurgitation and immediate postprandial vomiting controlled in part by slow feedings.

At 15 days of life she presented with imminent bronchoaspiration with cyanosis, flaccidity and apnea. She was admitted into the hospital for 24 h and was discharged with a diagnosis of GERD and was prescribed cisapride and antireflux formula. During her first 2 months of life she continued with immediate postprandial vomiting: observed from a few seconds to >5 min after milk ingestion. Although there was some improvement, this was only partial because she presented three near-drowning episodes within the first 4 months of life. For this reason, ranitidine was added along with antireflux measures, slow feedings and a change to soy and elemental formulas (Nutramigen). At 5 months of age, her diet was based on milk and liquefied food. At 10 months she showed intolerance to liquids and experienced near-drownings with semi-solid foods. She presented ten episodes of vomiting in 24 h prior to being admitted. For this reason, an esophagogram was performed. The study showed an area with an esophageal obstruction at the junction between the third middle and distal portion with luminal narrowing of <2 mm and with a rat-tail image resembling that seen in hypertrophic pyloric stenosis (Figure 1). An endoscopy was performed to confirm the findings of CSE and a foreign body was extracted out of the proximal esophagus identified as a portion of an apple (Figure 2). Esophageal balloon dilatations were started and during the second attempt, bleeding developed. The patient remained fasting and surgery was scheduled. A central catheter was installed and vancomycin were initiated to counter the risk of mediastinitis. Esophageal resection was performed at the site of the stenosis, and there

Figure 1. Esophagogram where the site of the stenosis at the junction of the middle third with the distal third level is observed. Also observed are food remains: apple (arrow).
was good postoperative evolution. Histopathological report was fibromuscular type CES (Figure 3). The patient was discharged from the hospital 7 days postoperatively and tolerated oral feeding. There were no complications from surgical wound infections or infections at the site of the chest tube.

Discussion

Congenital anomalies of the upper gastrointestinal tract include the esophagus, stomach and duodenum. In the majority of these, obstructive symptoms with reflux are presented. At times they manifest during infancy or childhood, whereas other cases are delayed until adulthood. Barium, tomography and endoscopy studies are very helpful. Esophageal abnormalities (up to 25%) are associated with other defects such as imperforate anus, pyloric stenosis, duodenal atresia, and annular pancreas and, less frequently, genitourinary abnormalities, cardiac and vertebral anomalies, VACTERL association (vertebral anomalies, imperforate anus, congenital heart disease, tracheal, esophageal, renal abnormalities and abnormalities of the extremities). CES is present in 1/50,000 of all live births. Intolerance to solid or semi-solid foods is one of the signs that should cause suspicion for CES. It presents when weaning is begun at 5 to 6 months of age, although there are reported cases of CES from childhood to adulthood. It is clear that at the time that treatment outcomes are not satisfactory, CES should be suspected. Most patients should initially be treated with esophageal dilatation, but if this is ineffective we should consider thoracotomy.
and resection of the stenosis site. Although the fibromuscular type of CES may respond to dilation, severe grades of stenosis require surgical excision. Among a large study of CES cases reported, in 36 patients usefulness of balloon dilation was reported as the only treatment. However, in other studies reported, there was a need for surgery in up to 78% of cases. Among complications that may occur with balloon dilation is esophageal perforation and, depending on the rapidity with which it is handled, mediastinitis and sepsis can be avoided.

In our patient, the presence of vomiting from her first day of life along with imminent near-drowning three times and the presence of perilesional healthy mucosa supported a congenital problem. Lack of response to the frequent changes of formulas and intolerance to solid or semi-solid foods supported the suspicion of CES. Because the patient’s feedings were administered slowly, she achieved a height and weight acceptable for her development. Because GERD is a common disease in infants and most patients are given only dietary restrictions, medications and change to an antireflux milk formula, although rare, CES should be considered in the comprehensive study of reflux using esophagogram or endoscopy. These studies help in establishing a comprehensive assessment in addition to excluding the presence of esophagitis associated with GERD. New options have emerged in the diagnosis and treatment, such as the use of endoscopic ultrasound, to define the type of stenosis, scheduled dilatations or surgery, and surgical treatment by thoracoscopy, which eliminates thoracotomy.

We can conclude the following. This infant, first seen at 10 months of age, was treated for GERD from birth, appeared to almost drown several times and received various treatments. Lack of response to medical treatment, poor tolerance to semi-solid foods and persistent postprandial vomiting all led to suspicion of CES. Diagnostic confirmation was performed with imaging studies such as esophagogram and endoscopy. When there is a report of vomiting in infants, GERD is the most likely diagnosis; however, symptomatology may help to differentiate GERD from other esophageal diseases that should not be ruled out by first intention.

Correspondence to: Dr. Ricardo J. Hernández Herrera Pediatra, Servicio Médico del Municipio de San Pedro Garza García Hospital OCA Monterrey, Nuevo León, México E-mail: richdzher@hotmail.com

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