TYPE III CONGENITAL PARAESOPHAGEAL HIATAL HERNIA – A RARITY IN PEDIATRIC SURGERY. A CLINICAL CASE AND LITERATURE REVIEW

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Abstract
A case of type III giant paraesophageal hernia in a child of 1 year and 6 months is presented. The child was operated through superior median laparotomy and the total reduction of the stomach in the abdominal cavity, the mobilization of the hiatal defect and hernia sac with its removal were performed. The posterior cruroraphy was done with strengthening of the zone by application of the equine pericardial acellular graft fixed by interrupted sutures. The anchoring of the stomach fornix to the left hemidiaphragmatic dome (gastropexy) and 180º partial anterior fundoplication were performed. The postoperative evolution was without complications and the patient was discharged in satisfactory condition on the 7 th day postoperatively. Conclusion: Using of canine pericardial acellular grafts could be a suitable option for the hiatal defect repairing in paraesophageal hiatal hernias in children, but this technique needs an adequate follow-up regimen.

Keywords: Hiatal hernia, equine pericardial acellular grafts

Introduction

Hiatal hernia (HH) is a variant of diaphragmatic hernia, characterized by transdiaphragmatic protrusion of the abdominal organs into the posterior mediastinum through the esophageal hiatus of the diaphragm [1, 2]. Pediatric HH occurs as a result of existence of the congenital diaphragmatic defect and many cases are asymptomatic [3].

Depending on the location of the gastroesophageal junction regarding diaphragm there are 4 types of HH [4, 5, 6]. Type I (85-95%) is an axial (sliding) hernia characterized by migration of the gastric cardia into the chest cavity, lack of the Hiss angle between the stomach and the esophagus and the development of the gastroesophageal reflux disease [7, 8]. Sliding HH is a result of widening of the muscular hiatal channel and the circumferential laxity of the phrenoesophageal membrane [9].

Types II, III and IV are paraesophageal hernias, constituting 5-15% from the totality of HH, which clinical significance is determined by the potential of mechanical complications [10, 11], although they are associated with gastroesophageal reflux too [12]. Type II is a paraesophageal hernia (rolling HH) characterized by normal position of gastroesophageal junction which is fixed to the preaortic fascia and median precardiate ligament, and the hernia sac contains the gastric fornix [4, 9, 13]. Type III (mixed HH) is a combination of type I and II hernias in which more than 50% of stomach is located in the mediastinum. In type IV the stomach is protruding in the mediastinum together with other abdominal organs. Along with the intrathoracic herniation of the stomach and gastroesophageal junction [14], the protrusion of duodenum, colon, omentum, spleen and pancreas could occur [15, 16, 17, 18, 19].

Paraesophageal hernia is a frequent diagnosis in adults [20], however in children it could be a complication after gastroesophageal or antireflux surgery or could be of congenital origin. [21, 22].

Congenital paraesophageal hernia is a rare nosological entity in children with obscure etiology and constitutes 3.5-5% from the all HH [6, 23, 24, 25]. The majority of cases occur sporadically, although familial cases of paraesophageal hernia are described [26, 27]. The term “giant paraesophageal hernia” is used in cases when more than 30% of stomach migrates into the chest cavity [28, 29].

We present a case of type III paraesophageal hernia in a child of 1 year and 2 months which was incidentally discovered.

Case report
The child I.M. 1 year and 6 months old was referred to the outpatient clinic of PMSI Mother and Child Institute with a suspicion of pulmonary tumor and pneumonia. At the admission the child complained loss of appetite, frequent regurgitations, periodic postprandial agitation. The onset of that signs was 3-4 months earlier. The plain chest X-ray in the outpatient clinic revealed a right sided cavitated mass (Fig. 1A). On upper gastrointestinal series the location of the gastroesophageal junction and a portion of stomach in the thoracic cavity was established (“sandglass sign”) (Fig.1B) and the child was admitted in the “Natalia Gheorghiu” National Scientific and Practical Center of Pediatric Surgery.

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Fig. 1. Simple chest X-ray (A) – right sided intrathoracic cavitary mass. Chest X-ray with upper gastrointestinal contrasting (B) – “sandglass” appearance of the stomach.

Fig. 2. Patient I.M. Computed tomography (the explanation is in the text).

Fig. 3. Schematic presentation of paraesophageal hernia detected in patient I.M with partially intrathoracic located twisted stomach (A); B - the scheme of the curoraphy, C
At the admission clinical examination revealed a satisfactory general condition without deficit of body weight, the child was afebrile. From anamnestic data the child was born from the 2nd pregnancy without peculiarities. The child was born through normal vaginal delivery with a body weight of 3259 kg and a height of 48 cm. There were no problems in the perinatal period. Afterwards the child development was normal, but he had repeated respiratory infections from the first months of life.

The CBC (Complete Blood Count) reveals low erythrocytes (3,6x10⁶ /L), hemoglobin –116 g/l, hematocrit – 34,0%, leucocytes – 11,5x10⁹/L. The hydroelectrolyte balance, hepatic and renal function and coagulation tests were within normal range.

The chest and abdominal simple CT scan as well as CT scan with upper gastrointestinal tract contrasting, performed for differential diagnosis, confirmed the location in the posterior mediastinum on the right side of the stomach fundus and partially of the stomach body, the diagnosis of type III paraesophageal hernia being established (Fig.2).

After preoperative preparation the abdominal cavity was opened by the superior median laparotomy. On surgical exploration the gastroesophageal junction was discovered in the chest cavity. The partially twisted stomach (the greater curvature was anteriorly located) (Fig. 3A) was also positioned in the thoracic cavity through the esophageal hiatus, which was markedly dilated. The large hernia sac was located in the chest cavity with some adhesions to the parietal pleura and pericardium.

After total reduction of the stomach into the abdominal cavity the gastrohepatic ligament was divided and the mobilization of the hiatal defect and hernia sac were performed. The hernia sac was sectioned and excised excepted a small portion which was firmly fixed to the pericardium. After identification and fixation of the gastroesophageal junction the slightly dilated distal abdominal segment of the esophagus was mobilized, and the posterior cruroraphy was performed. To avoid the excessive tightening of diaphragmatic cruses and the mechanical stenosis of the esophagus in the hiatus the Collins maneuver was performed. The cruroraphy zone was enforced with equine pericardium acelular biologic graft (Bioteck Heart) fixed by interrupted sutures (Fig. 3B). Then the gastropexy by fixation of the fornix to the left hemidiaphragmatic dome and 180º partial anterior fundoplication was performed.

The postoperative evolution was favorable. After hydroelectrolyte resuscitation the patient recovered without any complications and was discharged on the 7th day postoperatively in satisfactory condition. The control X-ray on the 7th day postoperatively revealed normal pulmonary areas without any opacities, the stomach was located below diaphragm and the esophagogastroduodenal passage was good. At the Th8 level there was a small residual space in the chest cavity, where the herniated stomach has been, but not containing contrast material (Fig.4).

**Discussions**

Although the first reports about congenital and posttraumatic diaphragmatic hernia occurred in XVI-XVIII centuries, the first description of the HH as a clinical entity was published by Henry Ingersoll Bowditch in 1853, the first operation for that type of diaphragmatic defect was performed in 1919 by Angelor Soresi [5, 29, 30]. It’s worth mentioning that the first description of the HH was probably done by Bright in 1836, who observed at the necropsy of a 19 years old girl a portion of the stomach localized in the chest cavity, the cardia being located at the.

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Paraesophageal hernias in children have several components. The defect is localized at the level of esophageal hiatus, covered by peritoneal sac, which extends to the right anterior side of the esophagus, as well as to the posterior mediastinum[23,33]. During migration the stomach tends to rotate around its axis (organoaxially), that could cause a partial or complete gastric obstruction between the above located esophagus and below situated duodenum [22].

In some cases HH could be associated with other congenital malformations such as diaphragmatic hernias [34], pulmonary sequestration [35], pulmonary agenesis [36], gastrochisis [37], gastrointestinal malformations such as short esophagus and microgastria [38, 39].

Usually the paraesophageal hernias in children are characterized by asymptomatic evolution, symptomatic cases manifest itself by recurrent respiratory infections, obscure gastrointestinal symptoms and anemia [6, 24]. Regurgitations and intermittent vomiting are the most frequent symptoms in children with HH [14]. In some cases the evolution of the paraesophageal HH could be aggravated by the development of severe complications including intrathoracic gastric volvulus [26], strangulation [40], incarceration and ulceration [4, 11]. The anemia in some cases could [41] be a manifestation of Cameron ulcer, which presents as linear lesions or erosions localized on the gastric mucosa folds at the diaphragmatic level. These lesions are determined by mechanical trauma during respiratory diaphragmatic contractions in combination with acid and ischemic injuries [42].

Prenatal diagnosis of HH established by ultrasound exam and MRI is of paramount importance and results in an early diagnosis of paraesophageal hernias in newborns which makes possible the surgical correction before the onset of complications [23].

In cases of paraesophageal hernias the imaging evaluation should be started with chest X-ray, which indicates the presence of abdominal organs in the thoracic cavity (usually gas bubbles). The contrast studies are performed to confirm the diagnosis and attest the full with contrast material stomach localized in the posterior mediastinum, often with an organoaxial volvulus [10, 22, 24]. The computed tomography is used to establish the definitive diagnosis, to assess the extent of the hernia content and to reveal affected lung complications [24].

The differential diagnosis of paraesophageal HH should be performed with pulmonary abscesses, congenital pulmonary cysts, hydatid cysts, pericardial cysts, esophageal duplications and epiphrenic diverticulum [22].

The paraesophageal hernia in children is an absolute indication for surgery, even in cases of incidental discovering or in the neonatal period, because of the high risk of potential fatal complications [6, 10]. There are some controversial issues regarding surgical approach in mixed paraesophageal HH [13]. Usually the abdominal approach through a superior median incision or right subcostal incision are preferred in children. These incisions allow an adecvated exposition of the subdiaphragmatic space. In some cases the thoracotomy could be used [22]. Lately more and more surgeons preferred the laparoscopic technique [43], even in the complicated forms of the disease [10].

Despite multiple controversies, the surgical treatment include the following elements: reduction of the hernia content into the abdominal cavity, hernia sac excision, mobilization of the distal esophagus to provide adequate length, closure of the hiatal defect, antireflux procedure and exploration for associated anomalies [24, 44].

Enforced hiatoplasty with synthetic or biologic protein grafts in paraesophageal hernias is an attractive option, with the aim to provide an additional resistance support for repaired esophageal hiatus, as well as safety of the reconstruction zone, decreasing the recurrence risk [45, 46, 47, 48]. In this context in the literature there are a lot of biologic materials used for this purpose in the adult surgery [49], including human acellular dermal matrix [50, 51], porcine small bowel submucosa [52]. However this problem is discussed rarely in pediatric surgery [53].

It is considered that antireflux procedure is a key element in the surgical treatment of HH, including paraesophageal hernias, because of the fact that anatomic and physiologic mechanisms of prevention of the gastroesophageal reflux are disturbed in this pathology [22, 54]. In children the Nissen 360º complete fundoplication is the gold standard, being the most frequent antireflux procedure used in the surgical treatment of gastroesophageal reflux [43]. As an alternative the partial antireflux procedures are proposed, including Toupet 270º posterior fundoplication, and anterior fundoplications on180º,120º and 90º. Techniques of partial anterior fundoplication differ from each other in terms of anchoring of the gastric fornix to the right diaphragmatic crus [55]. The most used are Thal [56, 57], Boix-Ochoa [58] and Watson [59] techniques of partial anterior fundoplication. In paraesophageal HH the advantage of partial fundoplication is the reduction of the risk of postoperative dysphagia, as well as anchoring of the gastric fornix to the right crus provides a support and stability of the hiatal reconstruction [60].

The recurrence rate of the congenital paraesophageal hernias is around 1,1%, the mortality rate varies between 0 and 20% and strongly depends on the associated comorbidities [60].

Conclusions

Type III paraesophageal hiatal hernia is a rare diagnosis, that could be identified incidentally because of the asymptomatic evolution, and a careful differential diagnosis, including thoracic cysts is needed. The partial torsion of the stomach that could be found in this mixed form of hiatal hernias has a high risk of severe complications development, which is a strong reason for
planned surgery if the diagnosis is confirmed. Using of equine pericardial acellular grafts could be a suitable option for the hiatal defect repairing in paraesophageal hiatal hernias in children, but this technique needs an adequate follow-up regimen.

References


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