Patch Reconstruction of Hemidiaphragm Agenesis by the Polypropylene Mesh Prosthesis

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We present a case of a middle-aged woman with right hemidiaphragm agenesis, which became evident after a blunt injury. Ultrasound, X-ray, and computed tomography confirmed the diagnosis, and the diaphragmatic congenital defect was closed by insertion of a polypropylene mesh prosthesis.

Key words: blunt injuries; congenital defects; diaphragm; hernia, diaphragmatic; implants, artificial; prosthesis implantation; wounds, nonpenetrating

The usual sites of congenital diaphragmatic hernias are esophageal hiatus, foramen of Morgagni, foramen of Bochdalek, and the dome, but also some other well-recognized sites, where various elements that make up the diaphragm fail to fuse together (1). Such hernias usually have a well-defined sac. The adhesions are not commonly found, but other congenital malformations may be present (2).

Acquired defects are either traumatic or postoperative and may occur anywhere in the diaphragm (3). Extensive adherence to surrounding structures is a common finding. The stomach, small or large bowel, omentum, or spleen commonly occupy the hernia, whereas the liver and kidney can be found less frequently.

Symptoms occur only when obstruction or strangulation supervenes. A patient may complain of vague dyspeptic symptoms, but if the hernia is large, particularly in children, respiratory embarrassment may occur. In infants, a diaphragmatic hernia is one of the causes of acute respiratory distress and demands urgent surgical repair. The lung on the affected side is often hypoplastic and surgical correction is recommended in all cases to avoid serious complications.

Case Report

Several days after a car accident, a 39-year-old woman complaining of vague discomfort, chest pain, and difficulty in breathing was admitted. As a result of a blunt trauma to the upper right abdomen and lower right chest, a herniation of the liver and colon occurred through a large right congenital defect into the thorax.

The diagnosis was based on physical signs and symptoms (bowel sounds heard during the auscultation of the chest), abnormal findings on the chest X-ray (Fig. 1), ultrasound, and computed tomography.

The initial examination by ultrasound revealed liver herniation into the chest. A barium enema proved the presence of the colon within the thoracic cavity, whereas chest computed tomography confirmed the absence of the right hemidiaphragm. On the basis of clinical findings and the presence of progressive cardio-respiratory insufficiency, a surgical treatment was indicated.

We chose laparotomy as the operative approach and found a complete agenesis of the right hemidiaphragm. The liver and the colon (ascending colon, hepatic flexure, and transverse colon) herniated into the thorax through the congenital diaphragmatic defect. There was no vestige of the hemidiaphragm except a thin, anterolaterally present muscular rim. With utmost care, the colon and the liver were removed from the thorax and placed again into the peritoneal cavity. The defect of the hemidiaphragm was repaired with a sheet of the synthetic prosthesis (Fig. 2).

As a substitute for the missing hemidiaphragm, a polypropylene (Prolene) non-absorbable mesh 20×10 cm prosthesis was used. The pros the sis was at tached to the rim of the endo-tho-
racic fascia and the muscle, using the continuous 2-0 unabsorbable polypropylene (Prolene) suture. Furthermore, a few interrupted sutures were placed to reinforce the polypropylene mesh in place. A 3-year follow-up was uneventful and no patch-related complications occurred (Fig. 3).

Discussion

A complete unilateral agenesis of the hemidiaphragm is an exceptionally rare finding in the adult hood. Usually asymptomatic and found incidentally, it does not necessarily need a surgical treatment. In our case, a 39-year-old woman had a herniation of the liver and colon into the right chest after sustaining a blunt trauma in a car accident. Intraoperatively, we found congenital diaphragmatic defect with only a thin muscular rim anterolaterally that required the patch reconstruction of the hemidiaphragm. A repair of the hemidiaphragmatic agenesis in the adulthood was not previously reported.

Cunniff et al (1) found a 6% incidence of diaphragmatic agenesis in their review of 102 live-born children with congenital diaphragmatic defects.

The diaphragm is formed between the third and eighth week of gestation, when the pleuroperitoneal membrane, septum transversum, dorsal esophageal mesentery, and portions of the wall of the coelomic cavity fuse together. The septum transversum, a thick plate of mesodermal tissue, begins to separate the thoracic and abdominal cavities of the intra-embryonic coelom during the fourth week of gestation. The esophageal dorsal mesentery forms the dorsomedial portion of the diaphragm. The pleuroperitoneal membranes extend medially and ven trally from their dorso lateral attachments to the body wall, until their free edges fuse with the septum transversum and dorsal esophageal mesentery. The division of the thoracic and abdominal portions of the coelomic cavity is thus completed around the sixth week. Myoblasts, originating from the third, fourth, and fifth cervical somites, migrate from the lateral and dorsal body wall and penetrate the pleuroperitoneal membranes, thus contributing to the muscular portion of the diaphragm (2). The embryological basis for diaphragmatic agenesis is unknown. Harrington (3) thought that unilateral absence of the diaphragm was probably the result of pleuroperitoneal membrane development failure, and therefore agenesis would be the most severe type of the posterolateral defect. However, the ultimate contribution of the pleuroperitoneal membrane to the diaphragm is small, so this theory does not explain adequately the absence of ventral diaphragmatic structures. Bingham (4) described the diaphragmatic agenesis as the anatomic entity distinct from the more usual posterolateral Bochdalek defect. Passarge et al (5) suggested that agenesis represents a developmental failure of three out of four diaphragmatic anlagen (septum transversum, eso-
phageal dorsal mesentery, and pleuroperitoneal membranes). By this schema, only the fourth anlage (myoblasts migrating in from the lateral and dorsal body walls) at attempts to develop the diaphragm, and this attempt results in a thin rim of tissue postero-laterally. Although compatible with what is an anatomically seen in many cases, this theory is unsubstantiated. The issue is difficult to resolve because of the inadequate pathologic description of the diaphragmatic defect in most cases as well as the lack of uniform terminology.

Passarge et al (5) described a family with two affected sibs whose only abnormality was the absence of the left hemidiaphragm. They reviewed the literature and found four other reports on families in which two sibs were affected, though their parents were normal. They concluded that an autosomal recessive gene might be responsible for that anomaly.

In 1988, Tzelepis et al (6) reported on the first case of complete absence of a hemidiaphragm in an adult. A computed tomography showed the absence of the left hemidiaphragm, with only the rim of the crus visible posteriorly. A barium enema and upper gastrointestinal series revealed small testes, colon, and the appendix in the left lower thorax. Reported 22-year-old man was asymptomatic and there was no specific treatment.

Diaphragmatic agenesis in an adult is extremely rare, with only four reported cases in the literature so far (6-8). Travalone and Cordova (8) reported a 39-year-old Filipino woman undergoing gynecologic surgery. Her right hemidiaphragm was absent, and hypolucency of the right lung was found on a routine preoperative chest X-ray examination. A magnetic resonance imaging revealed a herniation of the small bowel, colon, and right kidney into the right hemithorax, as well as a hypoplastic right lung.

Various methods of repair have been suggested in infants with hemidiaphragmatic agenesis, who survive beyond the neonatal period. Neville and Clowes (9) used a lobe of the liver to repair a right-sided agenesis. Holocomb (10) reported the use of a thoracic wall flap, and Rosenkrantz and Cotton (11) the use of a pedicled abdominal muscle flap. Eichelberger et al (12) repaired diaphragms with polyester fiber (Dacron), whereas Geisler et al (13) used Marlex. Lacey et al (14) reported that Silastic prosthetic material was inert and not advisable, whereas the polypropylene mesh, Marlex or Prolene, was found to be firmly incorporated by fibroelastic ingrowth, and was satisfactory as a diaphragmatic substitute. Laetitia and Bax (15) conducted a retrospective study to answer the question what happens over time with prosthetic patches used for closing congenital diaphragmatic defects. In 30 children who underwent a patch resection, Teflon was used 24 times, Goretex 5 times, and lyophilized dura once. Seven patients died shortly after the operation as a result of lung hypoplasia. In 2 of the remaining 23 patients, followed for a median period of 60 months, patch-related complications occurred. One patient had a recurrence of the hernia, and the other one developed a patch abscess. On the basis of their study results, Laetitia and Bax concluded that prosthetic patches used to close congenital diaphragmatic defects behaved well. Our experience of hemidiaphragmatic agenesis repair in the adult indicates that polypropylene mesh is an excellent diaphragmatic substitute.

References


