Congenital diaphragmatic hernia (Bochdalek’s hernia) revealed late in adulthood

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The aim of this retrospective study was to report the cases of diaphragmatic hernia revealed in adulthood and discuss the difficulties of diagnosis. We retrospectively reviewed all the files of patients operated for a period of 15 years. Each individual patient study was identified, reviewed and classified. We noted the position and the side on which the diaphragmatic hernia arise and the contents of the sac. We performed a graphic representation review for each patient included in this study and we noted also the patient’s sex, age, and clinical signs. For a period of 15 years, 11 adults (05 men and 06 women) were operated. The average age was 53 years (17-87 years). All our patients were symptomatic. A dyspnea was present in 5 patients. The diagnosis was not made on chest X rays in 8 cases, it has been made on CT scanner. MRI and CT scan were necessary for diagnosis in 3 cases. The hernia was in the left side in 10 cases and on the right side in 1 case (9%). The contents of the hernia were, most often, the colon (8 times), the stomach (5 times) and the small bowel (5 times). The post operative period was simple in 9 cases. We recorded one death. Bochdalek’s hernia is not rare, a diagnosis of an adult diaphragmatic hernia should always be kept in mind in patients with overlapping abdominal and respiratory symptoms with or without history of trauma. Simple chest X-ray may help in diagnosis. Surgery remains the only treatment for this kind of pathology.

Keywords: Bochdalek’s hernia, congenital diaphragmatic hernia, respiratory signs, diagnosis difficulties, surgical treatment.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a birth defect of the diaphragm. The most common type of CDH is a Bochdalek hernia; other types include Morgagni hernia, diaphragm eventration and central tendon defects of the diaphragm. CDH is a life-threatening pathology in infants and a major cause of death due to two complications: pulmonary hypoplasia and pulmonary hypertension.

In 1848, Bochdalek described the improper fusion of the posterolateral foramina of the diaphragm, a congenital defect that now bears his name. Well-known in neonates, Bochdalek’s hernia is the most common form of diaphragmatic hernia, occurring in approximately 1 in 2500 births and twice as often in males as in females. Bochdalek’s hernia results from failure of normal development of the posterolateral diaphragm during embryogenesis. Separation of the developing thoracic and abdominal cavities occurs during the eighth week of gestation, when the opening between the chest and abdomen (the pleuroperitoneal canal) closes. The herniated bowel passes through the defect, filling the chest cavity and causing hypoplasia of the left lung and a shift of mediastinal structures to the right side. It is not uncommon for other abnormalities to be associated with Bochdalek’s hernia; these may include neural tube defects and cardiovascular abnormalities (Pediatric imaging teaching files, 2006). In classic cases, Bochdalek’s hernias typically arise on the left side, contain fat or omentum predominantly and the kidney rarely, and do not necessarily lead to symptoms. Identification of previously undiagnosed Bochdalek’s hernia in adults occurs most frequently when the patients are undergoing CT for reasons that appear to
be unrelated to the (frequently asymptomatic) hernia—typically for surveillance of cancer or assessment of vague symptoms.

MATERIAL AND METHOD

This is a retrospective study of 11 cases of Bochdalek’s hernias. All our patients were collected in the departments of general surgery over a 15 year period from January 2000 to December 2015. This study concerned 6 men and 5 women. The average age was 53 years old (extremes: 17-87 years). The sides of the diaphragmatic hernia, the associated complications were determined using imaging techniques. Therapeutic methods with short and long term outcomes were recorded. Bochdalek’s hernia is defined as a posterolateral diaphragmatic interruption. It was differentiated from eventration by direct visualization of a diaphragmatic interruption or foramen located in the posterolateral position. Patients who met the criteria were included in the cohort, and all others were excluded. The estimation of hernia size (small or large) was more important than the volume of the hernia sac itself.

RESULTS

All our patients were symptomatic. Isolated or associated clinical signs leading to the discovery of diaphragmatic hernia were exclusively respiratory type of dyspnea in five patients, digestive type of repetitive vomiting in four patients and mixed (respiratory and digestive) in the last two patients. The chest X-ray has to suspect the diagnosis by showing a posterolateral opacity or fluid level pictures to intra thoracic anterior projection in 8 cases (Photo 1 and 2). Upper gastrointestinal opacification by nasogastric tube was performed in 1 case and showed the presence of intrathoracic stomach in position (Photo 3). The thoracoabdominal computed tomography (Photo 4) performed in 10 patients, confirmed Bochdalek’s hernia diagnosis in 9 cases and suspicion of hiatal hernia in the other case. For 2 patients, magnetic resonance imaging (MRI) showed a large posterior diaphragmatic defect. All patients were operated abdominally. Midline laparotomy umbilical addition was performed in all cases. The hernia was left in 10 cases and right in 1 case. The content of the hernia was, most often, colon (8 times), stomach (5 times) and small bowel (5 times). The closure of the diaphragmatic defect posed no particular problem and the establishment of prosthesis was never necessary. The postoperative course was uneventful in 10 cases. We recorded one death on the 5th postoperative day by organ failure (woman of 87 years with heart disease). Some patients had more than one abdominal organ contained at least partially within the hernia. All the Bochdalek’s hernias contained at least some fat or omentum.

DISCUSSION

The origins of descriptions of diaphragmatic hernia can be dated to writings of 1690 (Fine et al., 1987). Congenital hernias resulting from a developmental failure of posterolateral diaphragmatic foramina to fuse properly were first described by Bochdalek in 1848 (Salacin et al., 1994). The position of the foramina of Bochdalek is defined by the location of the diaphragmatic coronary ligaments bilaterally which when they fail to close or when they reopen Bochdalek's hernia occurs.

The hernia is called congenital when it occurs in the first few weeks of life and acquired when forms later. A Bochdalek’s hernia (80% to 90%) is one of two forms of a congenital diaphragmatic hernia, the other form being Morgagni hernia (1% to 6%) (Ameet et al., 2009). Bochdalek’s hernias occur more commonly on the posterior left side (85%) and right side (15%). Diaphragmatic hernias may be congenital or secondarily acquired due to blunt or penetrating trauma to abdomen. Congenital diaphragmatic hernia, (incidence one in 2000 to one in 5000 live birth) Incidence of traumatic diaphragmatic hernia resulting from severe blunt or penetrating abdominal trauma varies from 0.8% to 5% (Shailesh et al., 2013).

The overall prevalence of asymptomatic BH in adults is 6% (Ilkins et al., 1994). From all patients with a congenital BH only 5% will be diagnosed in childhood or adulthood (Osebold and Soper, 1976). New born who are born with a Bochdalek’s hernia are more than likely to have another birth defect caused by the hernia. About 20% of those children born with a Bochdalek’s hernia, also have a congenital heart defect. In addition, infants born with this condition may also have other abnormalities. “Between 5-16% of infants have a chromosomal abnormality (Diaphragmatic Hernia, 2006). In most cases, left-sided hernias or Bochdalek’s hernias have a ratio of 3:2 of males to females. In other words, Bochdalek’s hernias are more common in men. Bochdalek's hernia most commonly manifests during the patient's first few weeks of life. Diagnosis beyond the first 8 weeks of life is estimated to represent 5-25% of all Bochdalek's hernias (Nitecki and Bar-Maor, 1992).

In the neonate, Bochdalek's hernia is one of the leading causes of respiratory distress and remains one of the most common congenital anomalies of the thorax (Puri and Wester, 1997). Most neonatal Bochdalek’s hernias are left-sided (Langer, 1998). In adults, most Bochdalek's hernias are likely to be asymptomatic, and thus the finding of the condition is incidental (Langer, 1998; Fine et al., 1987). The symptoms are typically vague in those patients who do experience them at the time of diagnosis. The patients may present with chest
Photo 2. Intra thoracic fluid level of the left hemithorax

Photo 3. Upper GI-Opacification:
Presence of the stomach in left thorax

Photo 4 (a and b). CT scan
a. The colon and small bowel are intrathoracic
b. Ascension of the stomach, omentum and small bowel

Photo 1 (a and b). Chest x-ray
a. Posterolateral opacity of the left hemithorax
b. Right sided diaphragmatic hernia and pneumonia
pain or describe symptoms that are generally referable to the gastrointestinal tract (Fine et al., 1987; Hines and Romero, 1983). Sener et al. 1995 made a distinction between large and small hernias and noted that in most patients Bochdalek's hernias are small and contain only a modest amount of fat (Ahrend and Thompson, 1971). Most Bochdalek's hernias occur on the left side, allowing protrusion of the abdominal viscera into the chest. The herniated bowel passes through the defect, filling the chest cavity and causing hypoplasia of the left lung and a shift of mediastinal structures to the right side. It is not uncommon for other abnormalities to be associated with Bochdalek's hernia; these may include neural tube defects and cardiovascular abnormalities. Adult Bochdalek’s Hernias can present in two ways. They can give rise to vague, mainly gastrointestinal (Hines and Romero, 1983; Perhoniemi et al., 1992; Osebold and Soper, 1976), abdominal pain, nausea and vomiting, constipation) or respiratory, (Nouheim, 1998; Osebold and Soper, 1976) (chest pain, dyspnea, wheezing) symptoms, followed by severe attacks and episodes of incarceration with serious consequences. As usual these symptoms can be intermittent, as herniated viscera can spontaneously reduce causing symptom regression. In such cases, radiological investigations demonstrate reduction of the hernia with symptom resolution (Niwa et al., 2003). Others will present with serious complications associated with strangulation of herniated viscera, especially when the diagnosis has been missed or treatment delayed (Fingerhut et al., 1984). There have been reports of Bochdalek's Hernias presenting with sudden death from intrathoracic complications. Gastric volvulus is one of the rare but recognized complications of Bochdalek’s Hernias (Robb and Reed, 2006). Presentation with severe symptoms has been reported in 46% of cases and the mortality in these has been high (32%) because of visceral strangulation (Bochdalek's hernia, 2006). Formerly unrecognized Bochdalek's hernia in the adult patient is often diagnosed incidentally to other problems. Symptomatic cases of Bochdalek's hernia in adults are fairly rare, although case studies are found in the literature (Pediatric imaging teaching files, 2006). Supposed causes for late-presenting hernias include congenital herniation, blunt or penetrating trauma, physical effort (including sexual intercourse), pregnancy, labor and delivery, sneezing or coughing, and even ingestion of a large amount of meal (Chandarakant et al., 2015; Nitecki and Bar-Maar, 1992). If patients with late presenting hernias typically had previous CT or MR imaging studies that had been obtained over time, the causes of individual cases of hernia and the evolution of this entity could be discerned. Unfortunately, in most cases—as with all the patients in our study—no CT or MR images obtained before presentation and no follow-up images exist for comparison.

Adult Bochdalek’s hernias are very rare with less than hundred cases reported in the literature. With the wider use of computed tomographic scans and other imaging studies, asymptomatic Bochdalek’s hernias in older patients are increasingly discovered as incidental findings. In adults this type may be asymptomatic and incidentally diagnosed on chest radiographs, or it may present with dyspnea, recurrent chest infections and gastrointestinal symptoms (Gale, 1985).

The literature reports a left-sided predominance for Bochdalek's hernia, with left-sided occurrence of the hernia accounting for 70-90% of cases (Wilbur et al., 1994; Bétrémieux et al., 1995). These hernias may contain fat, retroperitoneal structures, or intraperitoneal contents, although the latter two conditions are exceedingly rare (Mehdi et al., 1995). In the instance of intrathoracic abdominal organs, a peritoneal sac is noted. The size of the hernia seen on cross-sectional imaging does not necessarily correspond to the size of the diaphragmatic defect, which may be substantially larger (Mehdi et al., 1995). The incidence of hernia with peritoneal sac varies from 10-38%. In right-sided
Bochdalek's hernias, the contents are predominantly the liver, the kidney, and fat. A left-sided hernia may contain enteric tract, the spleen, the liver, the pancreas, the kidney, or fat. Colon-containing hernias are rare and usually occur through left-sided defects (30). Bilateral Bochdalek's hernias were found by Gale (Bétrémieux et al., 1995) to represent a prevalence of 3-6%. Of note, congenital Bochdalek's hernia is usually an isolated anomaly a contradistinction to many other congenital defects in which multiple anomalies are found together (Wilbur et al., 1994). Traumatic diaphragmatic hernias may occur due to blunt (68-75%) or penetrating (25-32%) abdominal traumas (Wilbur et al., 1994). Diaphragmatic injuries are more common on left side as right hemi-diaphragm is protected by liver. Traumatic diaphragmatic hernia may have acute presentation in poly-trauma (may even present with hydro-pneumothorax with or without hollow viscous perforation) or they may present many years after initial injury (Gale, 1985). Diaphragmatic eventration on the other hand is abnormal elevation of intact diaphragm due to congenital muscular hypoplasia or due to phrenic nerve injury. Diaphragmatic eventration is rare (incidence <0.05%), being more common in males. It can be unilateral or bilateral, but it usually involves the left hemi-diaphragm (Wilbur et al., 1994). Diaphragmatic rupture occurs in as many as 25% of blunt thoracic and abdominal traumas (Mehdi et al., 1995) and traumatic diaphragmatic rupture is more common on the left side of the body (Reber et al., 1998). Such injury may be diagnosed initially, but it may also go undiagnosed (Naunheim, 1998). Some of the undiagnosed patients may never be identified as having Bochdalek’s hernia, whereas others will present with symptoms related to the herniation (possibly with strangulation of abdominal contents) (Montresor et al., 1999; Schumpelick et al., 2000). Other patients will go on to have the hernia identified on images performed for other, unrelated indications. Cases also have been reported in literature with ICD insertion leading to iatrogenic perforation in patients with undiagnosed diaphragmatic hernia (Wadhwa et al., 2014). Because of the severity of complications, each Bochdalek’s hernia diagnosed should be systematically treated (operated) even if it is asymptomatic (Habib et al., 2002). Surgery is the only treatment currently available. If surgery is recommended, a discussion with the surgeon is indicated to verify the planned surgical approach and ascertain whether there are other problems that will be surgically corrected at the same time. Imaging studies are performed to assess the hernia contents and to evaluate the presence of associated abnormalities. As noted by Alam and Chander, the organs that most commonly herniated into the chest through the diaphragmatic defect are: Stomach; ileum; colon; and spleen (Alam and Chander, 2005). Repair of Bochdalek's hernia in an adult may be via either an open abdominal or a thoracic approach, although hand-assisted thoracoscopic repair has been successfully undertaken (Mousa et al., 2006). Mesh (such as polypropylene mesh) may be used in the repair of the diaphragmatic hernia.

Postoperative care when an open approach is used would follow a typical plan of care for a patient undergoing repair of a diaphragmatic hernia. Prevention of respiratory complications and pain management are key nursing responsibilities. The complicated Bochdalek’s hernia must be treated urgently because of the risk of cardiorespiratory decompression, necrosis or gastrointestinal perforation. The incision of choice is midline laparotomy thereby make a complete inventory of the abdominal cavity, the reinstatement of the herniated organs, resection of the hernia sac and closure of the diaphragmatic defect (Fingerhut et al., 1978; Habib et al., 2002). In this study, the patients are aged from 17 to 87 years, had a Bochdalek’s hernia, confirmed by the operative findings and the absence of thoracoabdominal trauma can evoke a diaphragmatic rupture. The clinical histories of our patients show the difficulty of diagnosing of Bochdalek’s hernia in its later appearance. The clinical manifestations of this hernia are respiratory, digestive or mixed Chest pain; Difficulty of breathing; abdominal pain; Features of intestinal obstruction; and symptoms similar to those of gastroesophageal reflux disorder.

In this cohort of patients we shown that the diaphragmatic hernia in adults can remain asymptomatic for a long time before causing clinical signs, mixed, respiratory lung compression, as is the case of our seven patients, and digestive by strangulation viscera as is the case of four patients. Respiratory symptoms such as dyspnea, chest pain, cough moving towards primitive cardiopulmonary diseases. One patient had dyspnea placing on the hydro pneumothorax account. Gastrointestinal signs such as vomiting, epigastric pain, slow transit, hematemeses, melena, moving towards primitive digestive diseases, but can also be secondary to a herniation of Bochdalek’s hernia. One of our four patients had presented vomiting liable for hiatus hernia. The digestive symptoms often evolve for months or years, as in our population, and are triggered or exacerbated by the increase in intra-abdominal pressure after a meal supine or during exercise.

On chest x ray, the Bochdalek’s hernia can appears in chest radiographies as single or multiple intestinal air, frankly projecting above the diaphragm; by the presence of an intrathoracic of a gastric air pocket or a heterogeneous solid posterolateral mass (Habib et al., 2002; Zenda et al., 2000). This review is shown in 9 cases. Nevertheless, several sources of error are possible with chest radiography. The base of the thorax image can be interpreted as lung, pleural or diaphragmatic tumor on the AP view, mediastinal tumor on the lateral view, emphysema bubble pneumonia or lung abscess as in one of our patients (Habib et al., 2002). If in doubt, the high or low digestive opacification identifies the hollow viscera contained in Bochdalek's...
hernia and the seat of the hernia neck (Habib et al., 2002; Bujanda et al., 2001). However, Bochdalek’s hernia can be confused with a hiatus hernia or diaphragmatic hernia. CT and MRI appear to be the most successful exams because they can identify intra thoracic viscera, specify the seat of the hernia neck, and rarely discover a contralateral Bochdalek’s hernia (Habib et al., 2002). These exams are contributing in all cases. If it is not sought, the Bochdalek’s hernia can however be overlooked on CT (Steenhuis et al., 1994). The polymorphic clinical symptoms and non-specific and the limitations of chest radiography explain the diagnosis of HB is often asked adults on the occasion of a respiratory or gastrointestinal acute complication even after the patient’s death. The collapse of the lung causes acute dyspnea rest as is the case of three of our patients (Habib et al., 2002; 41). It may be associated displacement of the mediastinum and decreased venous return of veina cava that can cause cardiac arrest as is the case of one of our patients (Habib et al., 2002). The most common digestive complications are gastric volvulus, as in 5 cases, and the strangulation of the small intestine or colon manifested by an occlusive syndrome (Habib et al., 2002; Reed et al., 1987). More rarely, the strangulation of the digestive tract can cause bleeding ulcer, ischemic or diastatic perforation (Fingerhut et al., 1978; Habib et al., 2002; Zenda et al., 2000). The perforation of the digestive tract is manifested by pyopneumothorax and septic shock with sometimes brutal death. For this, surgical cure of all Bochdalek’s hernia must be done even without symptoms before the onset of complications.

CONCLUSION

A diagnosis of an adult diaphragmatic hernia should always be kept in mind in patients with overlapping abdominal and respiratory symptoms with or without history of trauma. Simple chest X-ray may help in diagnosis but still not sufficient, however contrast enhanced CT scan and barium studies should be done in patients with doubtful complicated presentation and before planning any surgical intervention. Further terrible presentation such as obstruction and strangulation of bowel loops including pyopneumothorax may be seen in patients with delayed post-traumatic diaphragmatic hernias. Definitive surgical approach helps in reducing morbidity and mortality in adult diaphragmatic hernia patients.

REFERENCES


