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Case Report

A rare case of Bochdalek diaphragmatic hernia with concomitant partial situs inversus

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ABSTRACT

Congenital diaphragmatic hernias clinically presenting in adulthood are exceedingly rare lesions, mainly left-sided defect (Bochdalek). Bochdalek hernias most commonly manifest 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 hernias. Here we have a 32 year old female patient who presented with 10x10 cm diaphragmatic hernia with dextrocardia who was asymptomatic for years.

Keywords: Congenital diaphragmatic hernias, Bochdalek hernia, Morgagni hernia

INTRODUCTION

The topic of Congenital Diaphragmatic Hernia (CDH) has frequently appeared in medical literature since its first description in the early 18th century. CHD is a term applied to a variety of congenital birth defects that involve abnormal development of the diaphragm. Congenital diaphragmatic hernia occurs in 1 out of every 2000-3000 live births and accounts for 8% of all major congenital anomalies. The risk of recurrence of isolated congenital diaphragmatic hernia in future siblings is approximately 2%. Familial congenital diaphragmatic hernia is rare (<2% of all cases), and both autosomal recessive and autosomal dominant patterns of inheritance have been reported.

Congenital diaphragmatic hernia is a recognized finding in Cornelia de Lange syndrome and also occurs as a prominent feature of Fryns syndrome, an autosomal recessive disorder with variable features including diaphragmatic hernia, cleft lip or palate, and distal digital hypoplasia.

The three basic types of congenital diaphragmatic hernia include Bochdalek Hernia (BH), anterior Morgagni Hernia (MH) and hiatus hernia. Congenital hernias resulting from a developmental failure of posterolateral diaphragmatic foramina to fuse properly were first described by Czech anatomist Vincent Alexander Bochdalek in 1848, although the origins of descriptions of diaphragmatic hernia can be dated to writings from as early as 1690. Diaphragmatic hernias through the posterolateral foramen of Bochdalek[1] represent the commonest type of congenital diaphragmatic hernia. The majorities are present during neonatal life and have a poor prognosis, being associated with congenital pulmonary abnormalities. The left-sided BH occurs in approximately 85% of cases. Left-sided hernias allow herniation of both the small and large bowel and intra-abdominal solid organs into the thoracic cavity. In right-sided hernias (13% of cases), only the liver and a portion of the large bowel tend to herniate. The incidence of Bochdalek and Morgagni hernias among adults is very rare. Bilateral hernias are uncommon and are usually

fatal. Presentation of a BH in an adult is exceptionally rare.[2,3]

In 1959 Kirkland published the first review of 34 cases of adult BH, and as of 1992 only 100 cases of symptomatic adult BH have been reported in world literature; however, with the growing use of abdominal CT this abnormality is being increasingly detected in asymptomatic individuals. Morgagni hernia, rarely seen anterior defect of the diaphragm, is variably referred to as a Morgagni, retrosternal, or parasternal hernia. It was first described by the Italian anatomist and pathologist Giovanni Morgagni in 1769.

Accounting for approximately 2% of all CDH cases, it is characterized by herniation through the foramina of Morgagni, which is located immediately adjacent to the xiphoid process of the sternum. The majorities of hernias occurs on the right side of the body and are generally asymptomatic; however, new-born may present with respiratory distress at birth similar to that found in Bochdalek hernias.

Additionally, recurrent chest infections and gastrointestinal symptoms have been reported in those with previously undiagnosed Morgagni hernias. We reported cases of Bochdalek hernias and cases of Morgagni hernias. The aim of this report was to present rare cases of adult presentation of Bochdalek and Morgagni hernias, and to discuss the clinical presentation and management of these rare cases. Because there are limited reviews of these rare hernias in the literature, the majority of these are single patient case reports. This case discusses about a rare presentation of 10x10 cm diaphragmatic hernia in an adult female with Dextrocardia who was asymptomatic for years.

CASE REPORT

This is a case study of a 32 year old female patient who presented with complaints of pain in left hypochondrium, dyspnoea on lying down in supine position post her normal vaginal delivery, recurrent chest infections since a year. On auscultation the heart sounds were heard on the right lateral end. There were minimal basal crepitations in left lower zone of chest in recumbent posture. Per abdomen: Mild tenderness in the left hypochondrium .No any other findings. Her chest x-ray showed classical bowel loops in the left side of mediastinum with dextrocardia (Figure 1).

CT thorax and abdomen (Figure 2 & 3): Large diaphragmatic hernia on the left side with herniation of fundus, body of stomach and the splenic flexure of the colon. Patient was then taken in Operation Theater and through a Roof top abdominal incision (Fig 4) the contents of the hernia were reduced and the sac was opened and the 2 leafs of diaphragmatic (Figure 5) and intercostals muscles were approximated with prolene. An intercostals drain was inserted in the Left pleural cavity.

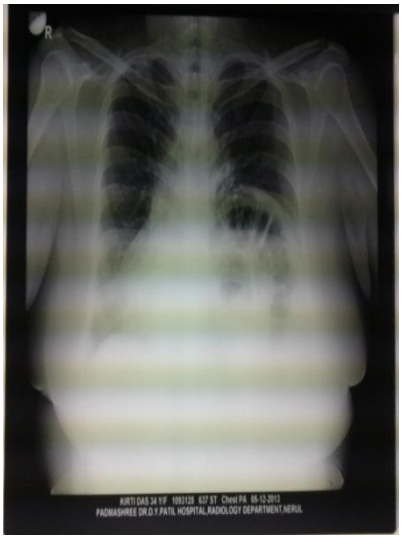

Figure 1: X-ray chest showing bowel loops in the left mediastinum.

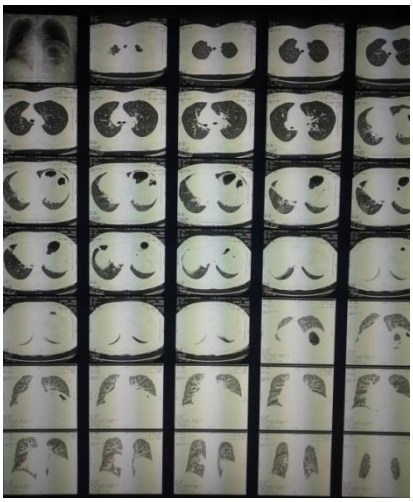

Figure 2: CT thorax.

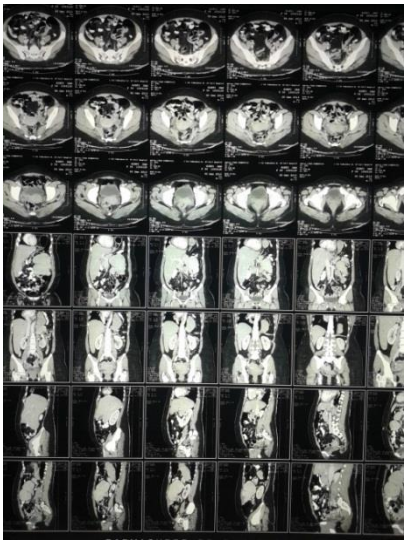

Figure 3: CT abdomen.

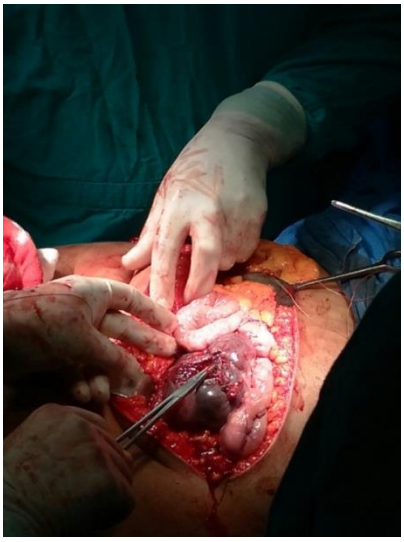

Figure 4: Intra operative image.

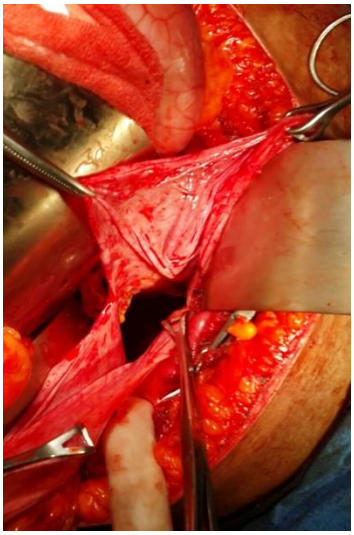

Figure 5: Intra operative image.

DISCUSSION

Bochdalek hernias: Clinical symptoms include dyspnoea, cough, wheezing, thoracic and abdominal pain, ileus, and recurrent chest infections. In right-sided Bochdalek hernias, the contents were liver, colon transversum, and omentum majus. In the left-sided Bochdalek hernias, the contents were gaster, caecum, appendix vermiformis, colon ascendens and transversum, intestinum tenue, omentum majus, and ren. The mean area of the hernial sac was 34.8 square centimeter. Clinical symptoms included dyspnoea, cough, epigastic pain, ileus and subileus, chest and abdominal pain, and recurrent chest infections. In right-sided Bochdalek 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 .Acquired hernias are also called "incidental" or "sub-acute" hernias. Bochdalek hernias most commonly manifest 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 hernias. In the neonate, Bochdalek hernias are one of the leading causes of left-sided hernia and 3 patients (25%) with right-sided hernia were determined. Computed tomography is known to be the most accurate method of diagnosing and evaluating the contents of Bochdalek hernias, especially the smaller ones.

CONCLUSION

Congenital diaphragmatic hernias clinically presenting in adulthood are exceedingly rare lesions. They can occur through an anterior parasternal foramen (Morgagni) or through a posterolateral, mainly left-sided defect (Bochdalek) representing persistence of the pleuroperitoneal canal. The location of the foramina of Bochdalek is defined by the location of the diaphragmatic coronary ligaments bilaterally. Bochdalek hernias occur when these soft-tissue anastomoses fail to close or when they reopen. If the herniation is present from the time of birth, it is termed "congenital". If the herniation forms later, perhaps because of extension of intra-abdominal or perirenal fat into the thorax, it is termed "acquired". The clinical symptoms of diaphragmatic herniation are frequently vague and nonspecific, including chest pains, dyspnoea, and gastrointestinal complaints, abdominal pain, nausea and vomiting, constipation or respiratory distress, chest pain, dyspnoea, and wheezing symptoms, followed by severe attacks and episodes of incarceration with serious consequences. Characteristically, 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. Others will present with serious complications associated with strangulation of herniated viscera, especially when the diagnosis has been missed or treatment delayed. There have been reports of BH presenting with sudden death from intrathoracic complications. Gastric volvulus is one of the rare but recognized complications of BH. Presentation with severe symptoms has been reported in 46% of cases and the mortality in these has been high (32%) because of visceral strangulation.[4] The incidence of hernia with peritoneal sac varies from 10 to 38%.

The current treatment of choice of a BH is surgical repair, even in asymptomatic cases, because of the risk of visceral herniation and strangulation. The surgical approach may be via a thoracotomy, laparotomy, or a combination of the two. In addition to these operative treatments, diaphragmatic defects were strengthened via primary closure or prolene mesh. Of all the types of CDH, Morgagni hernias are relatively rare. They arise from a septum transversarium defect due to the failure of closure of the pars sternalis with the seventh costochondral arch. This defect also is referred to as the space of Larrey, after Napoleon's surgeon, who described

the retrosternal space as an avenue through which pericardial tamponade could be treated. Some authors refer to the potential retrosternal space on the right as "Morgagni's gap" and the space on the left as "Larrey's gap". In medical literature, this hernia was presented titled as Morgagni-Larrey hernia,[5] Larrey hernia, or congenital anterior diaphragmatic hernia. In Morgagni hernias, intra-abdominal organs are herniated into the thoracic space through a right retrosternal fissure in the diaphragm.

Morgagni hernia can be associated with the following syndromes and congenital defects: Down's syndrome, Turner's syndrome, Noonan syndrome, Prader Willi syndrome, tetralogy of Fallot, ventricular septal defects, scoliosis, Morquio syndrome,[6] connective tissue disorders, dextrocardia, chest wall deformities, genitourinary abnormalities, and omphalocele.

The surgical approach is still controversial regarding the operative technique in Morgagni hernias. Some authors advocate the transthoracic or transabdominal approach others the video-assisted endoscopic technique. Preoperative imaging is crucial for delineation of the hernia's nature and the extent of diaphragm defect. Although small hernias can be closed by direct suturing, mesh repair is usually used in cases of large defects or muscle weakness. Recent reports have described successful treatment of these hernias by laparoscopic repair.[7-9]

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