RIGHT SIDED CONGENITAL DIAPHRAGMATIC HERNIA OF MORGAGNI TYPE IN ADULTS: A RARE ENTITY

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Abstract

Congenital diaphragmatic hernia (CDH) is an anomaly of infants. It is associated with other anomalies, including pulmonary hypoplasia, malrotation of the gut and patent ductus arteriosus (PDA). Hence it is essential that it is diagnosed early. Sometimes, it can remain unrecognized till adulthood, if no other associated developmental anomalies, like pulmonary hypoplasia, occur. In adults, it is diagnosed when intestinal contents herniate into the thorax causing pulmonary and intestinal complications. It is mostly present on the left side because of late fusion of the pleuro-peritoneal membranes, and the absence of the liver on the left side. Moreover, it is commonly due to trauma in adults. A case report of a right sided diaphragmatic hernia of Morgagni type in an adult was discussed.

Keywords: *Congenital diaphragmatic hernia, Pulmonary hypoplasia, Pleuro-peritoneal membranes, Morgagni.*

Introduction

Diaphragmatic hernia in adults is mostly traumatic. Congenital diaphragmatic hernia (CDH) presenting in adult life is rare (1). The delayed presentation is due to the absence of associated anomalies. It presents when intestinal contents herniate into the thorax causing pulmonary or intestinal complications. It occasionally presents in emergency, as incidence of gut ischaemia or strangulation is high.

CDH occurs in approximately 1 in 2000 to 1 in 5000 live births (2). Males and females appear to be equally affected. CDH is of two main types: postero-lateral or Bochdalek variety (most common) and anterior or Morgagni type (less common) (3).

We report a case of a middle-aged female who presented with symptoms of shortness of breath and abdominal pain, and was subsequently diagnosed with a congenital diaphragmatic hernia on the right side of the Morgagni type.

Case Report

A 55 year old lady, a housewife, with no known comorbidity, presented in OPD with complaints of progressive increase in shortness of breath and abdominal pain, for the past one year. Initially, it was compatible with her daily life activities, but for the past few days she became dyspnoeic even on walking for short distances. She had colicky abdominal pain intermittently, which was aggravated by intake of meals and relieved by defaecation. There is no history of fever, cough, haemoptysis, orthopnoea, chest pain, palpitation, weight loss, melaena or trauma. On examination, her vitals were normal with decreased air entry on the right side.

Chest X-ray was performed, which revealed gaseous shadows in the bowel with increased elevation of the right dome of the diaphragm (Figure 1). With a working diagnosis of diaphragmatic hernia, CT scan of the chest was performed, which confirmed the findings of a defect in the diaphragm of about 5 cm, with herniation of a healthy transverse colon (Figure 2).

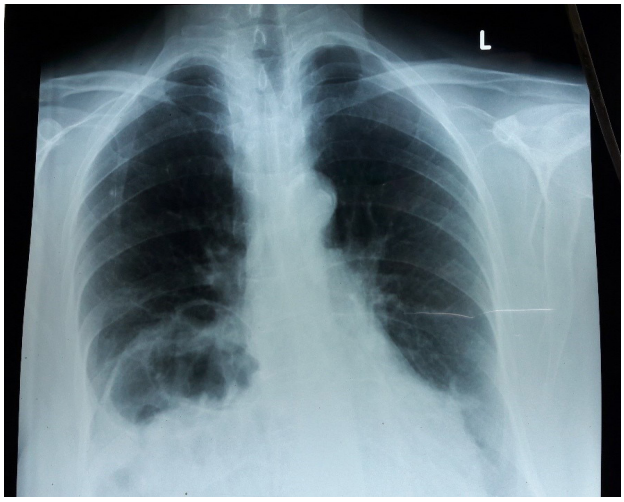

Figure 1: Chest X-ray revealing gaseous shadows in the bowel with increased elevation of the right dome of the diaphragm

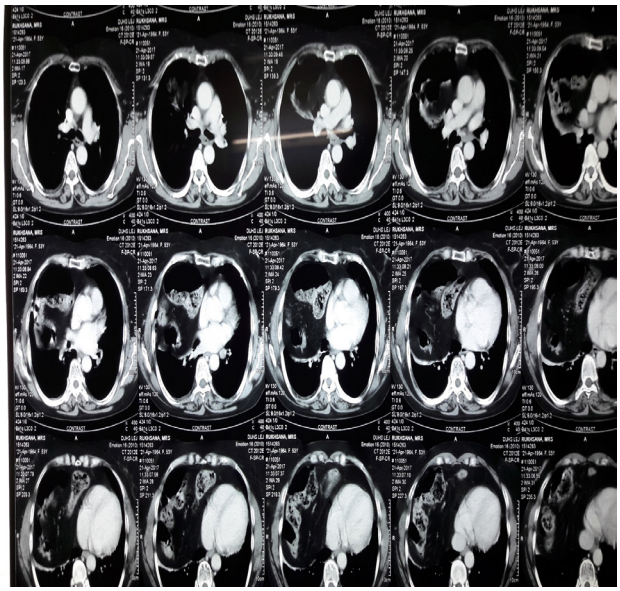

Figure 2: CT chest scan showing a defect in the diaphragm of about 5cm with herniation of healthy transverse colon

There was a defect on the right side of the diaphragm anteriorly, containing transverse colon which was easily delivered to the thorax (Figure 3,4). During surgery, the diaphragmatic defect was closed with polypropylene mesh. The patient's postoperative admission stay was uneventful and she was discharged on the third postoperative day. On the follow up visit, after two weeks, the patient was normal with significant improvement of her symptoms. Her wound healed, and normally stitches were removed.

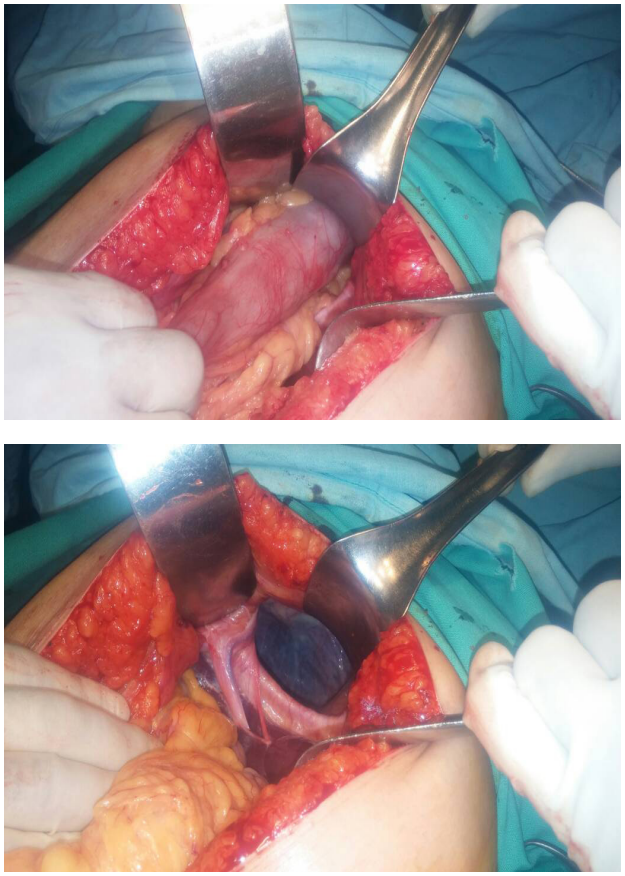

Figure 3 and 4: Surgical repair of diaphragmatic hernia using transabdominal approach

Discussion

Congenital diaphragmatic hernia (CDH) presenting in adult life is rare (1). The delayed presentation is due to the absence of associated anomalies. It presents when intestinal contents herniate into the thorax causing pulmonary or intestinal complications. It occasionally presents in emergency, as incidence of gut ischemia or strangulation is high.

The diaphragm is a muscular dome-shaped structure which separates the thoracic cavity from the abdominal cavity, and which consist of a central tendon and a peripheral muscular structure (4). Embryologically, it is formed from four structures: pleuro-peritoneal membranes, dorsal mesentery of the aesophagus, septum transversum and a body wall musculature. Pleuro-peritoneal membranes form the central dome, septum transversum forms the central tendon, dorsal mesentery of the esophagus forms the crus of diaphragm and the body wall musculature forms the peripheral musculature (3,4,5). The internal thoracic artery penetrates the diaphragm just lateral to the sternum in the anterior retro-sternal gap. This gap is termed Larrey's gap on the left and Morgagni's gap on the right (3,6).

The diaphragm has three main openings, one each for the vena cava, the aesophagus and the aorta, respectively, besides having small openings like Morgagni's or Larrey's gap, as described above. If during development, the four components are not fused together properly, then diaphragmatic hernia is likely to occur (7).

During the embryonic period, physiological rotation of the intestine occurs. If all the components of the diaphragm have not fused properly before return of the extracoelomic intestine to the abdomen, malrotation of the midgut might occur (1,8).

CDH is of two main types: postero-lateral or Bochdalek variety (most common) and anterior or Morgagni type (less common) (3).

The left sided Bochdalek variety is more common (90% occurance) (3,7,8). Morgagni type is more common on the right side, and this also occurred in our case in which transverse colon is presented. Morgagni type of hernia more commonly causes pulmonary symptoms while Bochdalek variety causes intestinal symptoms; however, in our case the patient had both types of symptoms (6,7). Bochdalek variety contains mostly stomach, spleen, pancreas, while Morgagni type contains transverse colon and partly liver, as hernial contents (3,6,7). In our case, only transverse colon was present.

CDH in adults is diagnosed late because of delayed presentation. Decreased breath sounds and presence of bowel loops in the chest cavity, on chest auscultation and Chest X-ray, prompts the clinician to acquire more sophisticated imaging modalities, like CT scan, to confirm the diagnosis.

The treatment of CDH in adults is mandatory, even if it is diagnosed incidentally, there is increased chances of strangulation of the bowel (3,4,6). The approach can either be transabdominal or transthoracic. The transabdominal is the more preferred approach as it is more convenient to handle the gut. The small defect in the diaphragm can be closed with a monofilament suture; however a larger one demands a mesh repair. Depending upon the expertise, it can be done laparoscopically.

We utilized the open transabdominal approach. The defect was of about 5cm, and so, we repaired it with a polypropylene mesh.

Conclusion

CDH in adults is a rare developmental anomaly. Prompt diagnosis and treatment is mandatory to avoid abdominal thoracic complications. The thoracic approach is equally as good as the abdominal approach but contributes a significant morbidity. Abdominal laparoscopic approach would be ideal, in expert hands.

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