

Case Report

Obstructed right Morgagni hernia with multiple congenital anomalies: a rare presentation

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ABSTRACT

Congenital diaphragmatic hernias occur in 1 in 3500 live births. Among the congenital diaphragmatic hernias, morgagni hernias are the rarest accounting for 2-3% of all diaphragmatic hernias. They occur through a congenital defect in the anterior part of the diaphragm just behind the sternum. In most cases the defect in the diaphragm is small and the patient may remain asymptomatic. The occurrence of obstruction or strangulation in these hernias is a rare occurrence. We report a rare case of right sided Morgagni hernia in a 14 years old boy who presented with features of subacute intestinal obstruction. Chest X-ray showed a large right sided diaphragmatic hernia with stomach and bowel loops compressing the right lung. A contrast enhanced computed tomography (CT) scan indicated large right sided diaphragmatic hernia containing gastric volvulus with twisted small bowel loops causing right lung collapse, and congenital anomalies of the right kidney and liver. The patient underwent exploratory laparotomy with an upper midline incision with reduction of hernia and primary repair of the diaphragmatic defect found in the anterior aspect of right diaphragm without placement of a mesh. An ICD was placed in right thoracic cavity to allow the lung to expand. Due to inadequate lung expansion a second ICD was placed in the right third intercostal space on the 9th post-operative day and removed on the 18th post-operative day. Patient was discharged with a single ICD on 20th post-operative day which was removed on first follow up. Patient was asymptomatic and recovered well subsequently.

Keywords: Morgagni hernia, Congenital diaphragmatic hernia, Obstructed diaphragmatic hernia

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a severe developmental defect characterised by herniation of abdominal contents into the thoracic cavity resulting in compression of the developing lungs.¹ These are mostly diagnosed in the new-born or early childhood. It occurs in 1 in 3500 live births.^{1,2} About 5-10% individuals remain asymptomatic during childhood and symptoms appear later in life.³ Its aetiology is poorly understood and is thought to be both due to genetic and environmental factors.¹ Affected genes include transcription factors such as GATA4, ZFPM2, NRF2 and WT1.¹

CDH are classified depending on the site of defect into: Bochdalek hernia-defect in posterior aspect of diaphragm and Morgagni hernia- defect in the anterior aspect of diaphragm.

Bochdalek hernia accounts for more than 85% of all congenital diaphragmatic hernias while Morgagni hernia accounts for only 2-3% of all diaphragmatic hernias.⁴

Morgagni hernias were first described in 1796 by Giovanni Batista Morgagni as substernal or retrosternal defects in the diaphragm.³ They are also called as Larrey hernias as the defect is also referred to as the space of Larrey.³ It seen to occur more commonly in females and is more common

on the right side. Literature on Morgagni hernia is scarce with lack of well-defined treatment protocols.

CASE REPORT

A 14 years old boy presented to our hospital with complaints of abdominal pain since 4 days and vomiting since 2 days. Patient also had loss of appetite since 4 days. Patient had no history of trauma in the past. On examination the patient had tachycardia and tachypnoea with mild abdominal distention. There was no tenderness over the abdomen. Auscultation revealed increased bowel sounds over the abdomen as well as the right side of the chest. A nasogastric tube was inserted which drained 500ml of gastric content. An urgent chest radiograph revealed large right sided diaphragmatic hernia with stomach and bowel loops as its content completely filling the right thoracic cavity and compressing the right lung with mediastinal shift to the left.

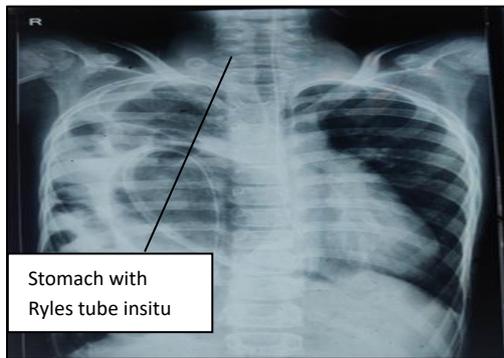


Figure 1: X-ray showing hernia.

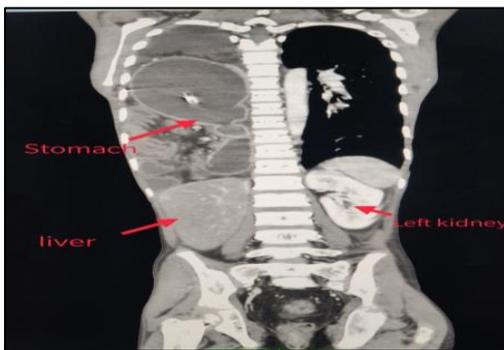


Figure 2: CT scan showing right diaphragmatic hernia.

The patient was taken for an urgent contrast enhanced computed tomography (CT) scan of the abdomen and chest which revealed upward migration of the stomach into the right thoracic cavity and its malrotation along its long axis resulting in inversion of the greater curvature. There was dilatation of the stomach and the 1st and 2nd part of duodenum with twisting of the small bowel loops and their mesenteric vessels. The right lung was compressed and displaced supero-medially with shifting of the trachea and mediastinum to the left. These features were suggestive of

right-side diaphragmatic hernia with gastric volvulus with gastro duodenal obstruction.



Figure 3: CT coronal view showing right ectopic kidney and liver.

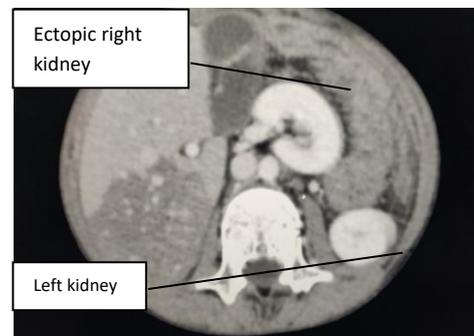


Figure 4: CT abdomen axial view showing right ectopic kidney.

The CT scan also revealed that the liver was displaced inferiorly and aligned vertically extending up to the right iliac fossa with its left lobe in the right hypochondria. There was also thrombosis in the right branch of the portal vein. There was a right ectopic kidney located anteriorly in the abdominal cavity in the left para midline at L2-L4 vertebral level with its hilum facing right postero-laterally. The origin of the right renal artery was normal.

The patient was explored through an upper midline laparotomy incision extending from the xiphisternum to the umbilicus. On exploring the caecum and appendix were found to be lying in the midline above the umbilicus with partial gut malrotation. The stomach, duodenum, part of the small bowel and the transverse colon were found to have herniated through a large defect of approximately 10x5 cm in the anterior part of the right diaphragm. The liver was found to be vertically aligned extending up to the right iliac fossa with the left lobe in the right hypochondria. A right ectopic kidney was found anteriorly to the left of the midline with the hilum towards the right.

The hernia content was reduced through the defect through which the right lung could be seen to be collapsed. After

reduction of the hernia contents a chest tube was inserted in the right thoracic cavity. The defect in the anterior part of the right diaphragm was repaired with continuous interlocking prolene sutures. A single abdominal drain was placed in the pelvis and the abdomen closed. The patient was kept on controlled ventilation for 48 hours then weaned off and extubated. He was kept on continuous oxygen support.

Due to inadequate lung expansion and persistence of right sided pneumothorax a second chest tube was inserted in the right 3rd intercostal space in the mid-axillary line on the 9th post-operative day. This helped in better lung expansion. Patient's skin staplers were removed on 15th post-operative day. The chest tube later inserted was removed on the 18th post-operative day. The abdominal drain was draining considerable amount of serous collection and hence was removed later on the 19th post-operative day. The patient was discharged with a single chest tube in-situ on 20th post-operative day in healthy condition. The chest tube was removed on first follow-up 7 days later after chest radiographs showed complete lung expansion. He was healthy and asymptomatic on subsequent follow-ups with no complications seen even after 1 year of surgery.

DISCUSSION

Anterior defects in the diaphragm were first discovered in 1796 by Giovanni Batista Morgagni and described as Morgagni hernias.^{2,3} These are the rarest of all congenital diaphragmatic hernias accounting for only 3% of cases.^{4,6} The pathophysiology of diaphragmatic hernias is not well known. The congenital defect in the anterior part of the diaphragm is thought to be due to mutation in certain genes encoding for transcription factors responsible for the development of the diaphragm such as GATA4 and NR2F2.¹ Genetic causes have been identified in approximately 30% of cases.¹ Morgagni hernias occur due to failure of fusion of the fibro-tendinous elements of diaphragm, that is, sternal and costal attachments, which leaves behind a muscle-free area known as the costosternal trigone or the space of Larrey or Morgagni's foramen through which the hernia occurs eventually.⁷

Morgagni hernias are more common on the right side than the left due to extensive pericardial attachments on the left diaphragm.⁷ In 90% of cases Morgagni hernias are seen on the right side while in 2% cases on the left side and almost 8% cases are bilateral.⁷ It has been reported that almost 70 % cases of Morgagni hernias are females.³ Although cases of Morgagni hernia can present in early childhood with respiratory symptomatology, most of the cases remain asymptomatic and are detected later in life.⁷ In most of the cases symptoms are usually respiratory followed by gastrointestinal.⁶ Symptomatic adult cases of Morgagni hernia are very rare with majority of the symptomatic cases presenting as life threatening obstruction or strangulation. In our case the patient was a young boy with the hernia was on the right side and presented with obstruction.

Diagnosis in most of the cases can be established by plain chest radiograph. CT can be used for better assessing the contents of the hernia sac. The diagnostic accuracy of both CT and X-ray when used together is 80-90%.³ CT scan is the preferred method of choice for diagnosis of diaphragmatic hernias.⁵ Magnetic resonance imaging (MRI) can be used in some cases to differentiate the hernia from mediastinal masses and intrathoracic tumours.⁷

Management of Morgagni hernia mainly depends on its presentation. Life threatening cases of obstruction or strangulation are managed by laparotomy and reduction of the hernia with repair of the defect. Upper midline laparotomy is preferred over thoracotomy.^{2,6} Although some surgeons advocate the repair of the defect in even asymptomatic cases to prevent future complications, successful non-operative management has also been seen.³ This mainly depends on the size the defect. Repair of the defect can be done either by primary suturing or with prosthetic meshes or both.³ Primary repair is most commonly done.⁶ If a mesh has been placed it should cover the entire hernia defect with overlap of all sides as a safety margin.³ However it has been seen that primary suturing is preferred when working near the pericardium to avoid injuring it.³ Moreover the placement of a mesh can cause more harm than good if bacterial contamination is present.³ No mesh was placed in our case and only primary repair with non-absorbable sutures was done. There has been a recent trend towards laparoscopy for identifying and repairing the diaphragmatic defect but conversion to open laparotomy is seen in most complicated cases of obstruction or strangulation especially if bowel necrosis has set in.

Post-operative stay of the patient in our case was prolonged due to inadequate lung expansion for which a second chest tube had to be placed. In most other cases if there are no respiratory complications patient can be discharged early. Post-operative period is usually uneventful if repair of the defect is properly done, bowel and lung are healthy and there is no bacterial contamination.

CONCLUSION

Morgagni hernias are an extremely rare type of congenital diaphragmatic hernias with little literature describing their presentation and management. Occurrence of large Morgagni hernia with complications such as obstruction or strangulation is unusual and its management is not well defined. Laparotomy is most commonly done for reduction of the herniated contents and closure of the defect especially if there are signs of respiratory distress or bowel obstruction. Our case highlights the difficulties in diagnosis and management of an unusual case of large right sided Morgagni hernia with intestinal obstruction and congenital anomalies of the liver and kidney.

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