LATE PRESENTATION OF CONGENITAL DIAPHRAGMATIC HERNIA IN ADULTS: “OUR EXPERIENCE”
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ABSTRACT: Congenital Diaphragmatic Hernia (CDH) usually presents with respiratory distress in the neonatal period. Late onset CDH is less common and is associated with a wide range of clinical symptoms. The authors report two cases with late presentation of congenital diaphragmatic hernia. Two previously healthy patients presented to the hospital with vague gastrointestinal and cardio respiratory symptoms and were investigated with chest radiographs and computed tomographic scans of thorax which suggested the diagnosis of Diaphragmatic Hernia. Contents of the hernia (viz. spleen and bowel loops in the first case and omentum in the other case) were reduced and mesh repair was done. Post-operative recovery was uneventful. These cases throw light on the diagnostic challenges of this disease due to its diverse clinical presentation, the importance of prompt diagnosis and intervention. Early correct diagnosis and treatment are associated with an excellent clinical outcome.

KEYWORDS: Congenital diaphragmatic hernia, Respiratory distress, Neonatal period.

INTRODUCTION: Lazarus Riverius first described a congenital diaphragmatic hernia (CDH) in 1690 found incidentally in a 24 year old man at postmortem. CDH commonly presents in the neonatal period with respiratory distress. Late-presenting CDH is rare and poses considerable diagnostic challenges. CDH 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%. Late presentation of CDH is uncommon, accounting for 5-30%.

Congenital diaphragmatic hernia has been classified into 3 basic types:
• Bochdalek (Posterolateral defect): most common variety seen in up to 70-90% cases, more commonly seen on the left side (85% of cases) than the right side (13% of cases). Bilateral variety being rare and fatal.
• Morgagni or parasternal or retrosternal (Anterior defect) seen in up to 2-6% cases, more commonly seen on the right side than left side.
• Hiatus hernia seen in up to 14-24% of cases.

Late-presenting CDH is less common and the majority present with non-specific gastrointestinal or respiratory symptoms later in childhood or even in adult life. The aim of this report is to present rare cases of adult presentation of bochdalek and morgagni hernias and to discuss the clinical presentation and management of these rare cases.
Case Capsule 1:

- 26 year old male presented to our hospital with recurrent upper abdominal pain vomiting and difficulty in breathing immediately on taking food on and off since 2 years. On examination abdomen was scaphoid, non-tender. A Dull note on percussion was present over lower two third of the left hemithorax. There was decreased air entry on left side of chest on auscultation, bowel sounds were heard in left hemithorax. Routine blood investigations i.e., Complete blood count, Random blood sugar, Liver function test, Renal function test were within normal limits. Chest radiograph PA view showed shift of mediastinum and cardia to the right; air fluid levels in lower two third of left hemithorax; obscured left costophrenic angle which forced the physicians to give a diagnosis of empyema. CT scan of thorax showed spleen, splenic flexure of colon and small bowel loops in left hemithorax and shift of mediastinum and cardia to the right.

A Diagnosis of Large Left sided Diaphragmatic hernia (Bochdalek hernia) was made.

After all pre-operative preparations, patient was taken up for Diagnostic Laparoscopy which showed herniated contents and lot of adhesions at Diaphragmatic hiatus. Decision was taken to convert to open thoraco-laparotomy and thoraco-abdominal incision placed in 8th intercostal space (Figure 3). To our surprise, Spleen was found higher up near the apex (Figure 4) deriving blood supply from intercostal vessels. Hence decision was taken to do a splenectomy. Left lung was found to be hypoplastic (Figure 5). Hernial contents were reduced. Repair was done with composite mesh (parietex) placed from thoracic side. Multiple prolene stay sutures were placed anchoring the mesh to the diaphragmatic surface. ICD was placed in 8th intercostal space. Patient withstood the procedure well. Post-operative period was uneventful, ICD was removed on 12th Post-operative day and patient was discharged on 18th Post-operative day.

Fig. 1: Chest X-Ray PA View

Fig. 2: CT scan of thorax
Follow up chest X-Ray taken 8 months post-surgery showed complete expansion of left lung.

**Case Capsule 2:** 75 year old woman presented to our hospital with dyspnoea on exertion, respiratory catch on lying down and palpitations on and off since 10 years. She visited a number of cardiologists and was even stented twice in the past. She had no history of trauma in the past. On examination, there was decreased air entry over lower half of right hemithorax. Heart sounds were well heard. Routine blood investigations were within normal limits. Her previous chest x-rays were normal. CT scan thorax of t showed right paracardiac defect with herniated omentum in right paracardiac region.
A Diagnosis of Right Paracardiac Morgagni Hernia was made and after preoperative assessment patient was taken up for surgery. Laparoscopic approach was undertaken. Contents i.e omentum was reduced back into peritoneal cavity, defect was repaired by placing a dual mesh from abdominal side and anchoring it by using tackers. Surgery was uneventful. Patient withstood the procedure well, post-operative period was uneventful and patient was discharged on 10th Post-operative day.

DISCUSSION: 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. The diaphragm forms from the septum transversum which grows downwards and backwards to meet the dorsal mesentery of the foregut. Pleuro-peritoneal folds then develop on each side of the septum and extend posterolaterally dividing the chest from the abdomen. A small triangular posterior portion, the pleuro-peritoneal canal is the last area to close. The diaphragm is completed at the ninth week by an ingrowth of muscle fibres from cervical myotomes into the pleural and peritoneal folds. This coincides with the return of intestine from the umbilical stalk. Delayed closure of the diaphragm (the left is normally a week later inclosing than the right), or early return of intestine to the abdomen may result in a hernia. If this happens before the pleuro-peritoneal canal closes the hernia has no associated peritoneal sac, but if later in the embryological sequence, 'after canal closure and before muscle ingrowth is completed, a sac will be present. Left-sided hernias are more common than right because of the later closure of the pleuro-peritoneal canal and the protective effect of the liver.

A left-sided hernia may contain enteric tract, the spleen, the liver, the pancreas, the kidney, or fat. Colon-containing hernias are rare. Colon and spleen were the contents in our case also.

Morgagni’s hernia was first described by the Italian anatomist and pathologist Giovanni Morgagni in 1769. It arises from a septum transversarium defect due to the failure of closure of the Pars sternalis with the seventh costochondral arch. Comer et al most often found the hernial sac containing transverse colon, omentum, liver, and, less frequently, small bowel or stomach. Similar contents were found in our 2nd case. Morgagni hernia can be associated with the following syndromes and congenital defects: Down’s syndrome, Turner’s syndrome, Noonan syndrome, Prader Willi syndrome, TOF, VSD, omphalocele.

Herniation of a solid organ e.g. liver, spleen, is recognized as a risk factor for radiological misdiagnosis of late presenting CDH because the typical appearance of bowel herniation may be sparse or obscured leading to a diagnosis of lung disease instead. A previously normal chest radiograph does not exclude late onset CDH. The radiological findings may lead to misdiagnosis of pneumothorax, pleural effusion or pneumonia. In one series, 18% of the cases of CDH were subjected to insertion of chest drain. A chest radiograph following the passage of a nasogastric tube is useful in facilitating the diagnosis of CDH.

CONCLUSION: The variable clinical features of CDH presenting beyond the neonatal period may result in clinical and radiological misdiagnosis. CDH with complicating mediastinal shift and
respiratory distress requires urgent gastrointestinal decompression and respiratory support. The most significant factor in achieving diagnostic success is to consider it early in the differential diagnosis to avoid misguided or delayed therapy.

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