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Laparoscopic Repair Of Arare Symptomaticgiant Hernia Of Bochdalek In An Adult – A Case Report

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ABSTRACT:

The incidence of Congenital Diaphragmatic hernias 1:2000 to 1:12500 amongst all live births. (1,2) It is rare adults with a incidence of 0.17%. (3) Presentation in adults is usually asymptomatic and are detected incidentally during imaging studies.Somepatients present when complications occur. Otherwise when symptoms are present, it is usually more of GI symptoms or rarely with respiratory symptoms. Here we report a case of a 39yr old female patient who presented with respiratory symptoms and had a Left side Bochdalek hernia which was treated successfully by laparoscopic mesh repair. Patient is fine after 1 year of follow-up.

Key Words: Diaphragmatic hernia, Congenital, Bochdalek hernia, laparoscopic mesh repair

1. INTRODUCTION:

Bochdalek hernia is a type of congenital diaphragmatic hernia where the defect is in postero-lateral aspect of the diaphragm. About 38% of CDH cases in adults are misinterpreted for plural effusion, lung cyst, empyema, pneumothorax due to its rarity and hence treatment is delayed. Even though a chest x-ray can detect a diaphragmatic hernia, CT scan should be considered the investigation of choice.All diaphragmatic hernias should be repaired at the earliest,once it has been diagnosed, because of the risk of strangulation or volvulus or perforation of its contents. Minimally invasive procedure, either laparoscopically or thoracoscopic approach is safe and effective form of management. Primary closure of the defect with or without mesh reinforcement is recommended. Here we report a case of 39 yr old female patient who presented with breathlessness which was aggravated in supine position for a duration of 6 months.Respiratory system showed decreased air entry on Left axillary, Infra axillary and subscapular areas. CT scan confirmed a Left diaphragmatic hernia.

Successfully performed a laparoscopic mesh repair for a defect of size 7 X 5 cm after reduction of Small bowel, transverse colon, omentum, and left kidney.

2. MATERIALS AND METHODS:

A 39yr old female patient came to our OP with complaints of breathlessness for 6 months duration which was aggravated on supine position. Her symptoms has worsened over the past few weeks. No symptoms suggestive of GI disturbance like nausea, vomiting, constipation, retching, heart burns. Normal bowel and bladder habits. She is mother of 2 children born by caesareansection.Nopast history of trauma to the chest or abdomen. She was born through a normal vaginal delivery and no history of breathing difficulty in the neonatal or in childhood period. No family history of diaphragmatic hernia.
On examination of the patient she was comfortable at rest, vitals were stable, abdomen was not distended, wandering spleen was palpated as aintra-abdominal mass with restricted mobility in left half of the umbilical region extending to epigastrium above and to hypogastrium below. Insinuation of the finger between the coastal margin and the mass was possible. Dull on percussion with surrounding resonance. Onauscultation of the chest, decreased air entry on Left axillary, Infra axillary and subscapular areas. Heart sounds were well heard on the right hemithorax, compared to Lt hemithorax. Dull note heard on percussion of the base of left hemithorax. Chest Xray showed air filled bowel loops in the left hemi thorax. (Fig 1a)

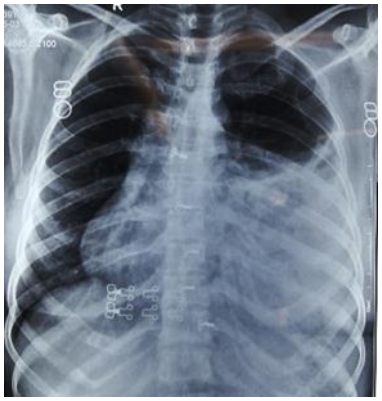

Fig 1a: chest x rayshowed air filled bowel loops in the left hemi thorax

There was no evidence of GERD or hiatus hernia in upper GI scopy.CT of the chest and abdomen with oral and IV contrast confirmed the diagnosis of Left sided Bochdalekhernia. (Fig 1b)

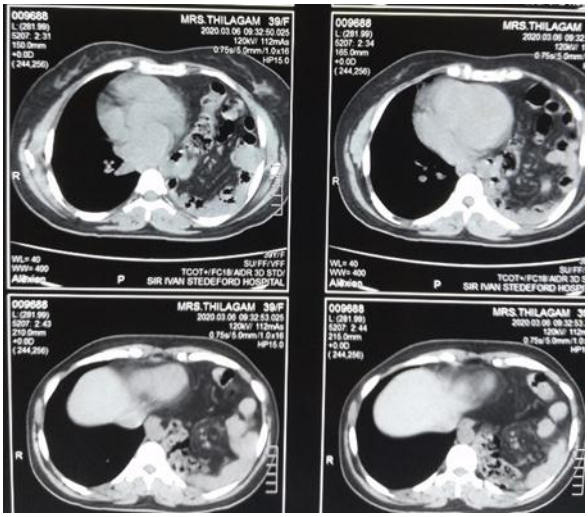

Fig 1b: CT scan chest

Preoperative preparation included intensive spirometry exercise, nebulization.Chest physician opinion was obtained and PFT done to find out her pulmonary reserve.

Laparoscopic diaphragmatic hernia repair was done by creating pneumoperitoneum by open techniqueanddiagnostic laparoscopy showed a tossed spleen in the midline, without any attachment to the diaphragm.Oesophagealhiatus,liver and stomach werenormal.Majority of the small bowel and the omentumand transverse colon were seen towards Lt diaphragmatic area herniating through the defect into the thorax.

A steep reverse trendlenburg positionrevealed the upper rim of the defect in the Lt posterolateral aspect of the diaphragm.After reduction of the small bowel loops, omentum and transverse colon, partial herniation of the Left kidney retroperitoneally was noted and the same was reduced after incising the peritoneum all along the posterior rim of the defect. A posterolateral defect of size 7cm X 5 cmwasdelineated. (Fig 2a) Lt triangular ligament of the liver was divided to mobilize the Lt lobe of the liver to accommodate the mesh.

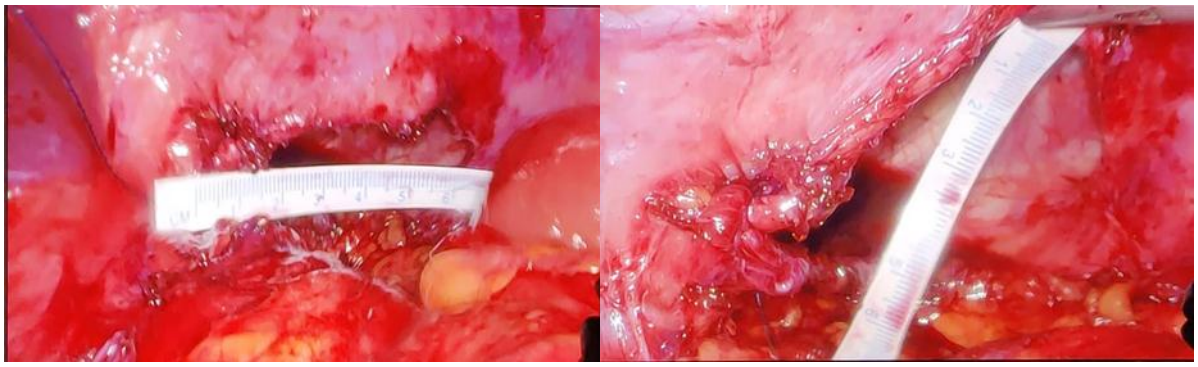

Fig 2a: Defect

On inspection of the Lt thoracic cavity there was no hernial sac and a hypoplastic lung was present. (Fig 2b)

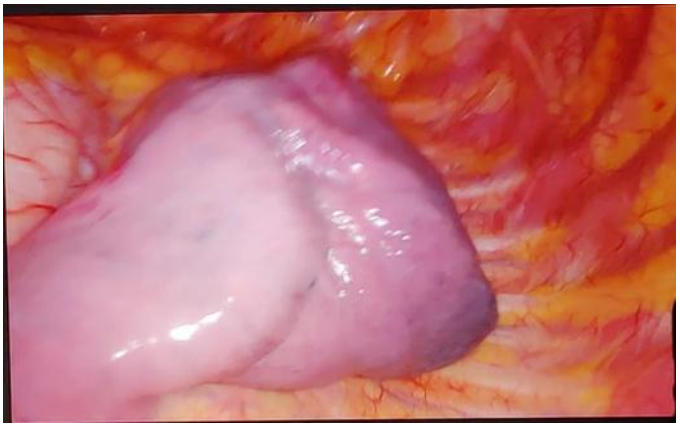

Fig 2b: Hypoplastic lung

Primary closure of the defect done with continuous 1-0 barb polypropylenesutureand reinforced with a 10X 15 cm dual mesh. (Fig 2c,2d)

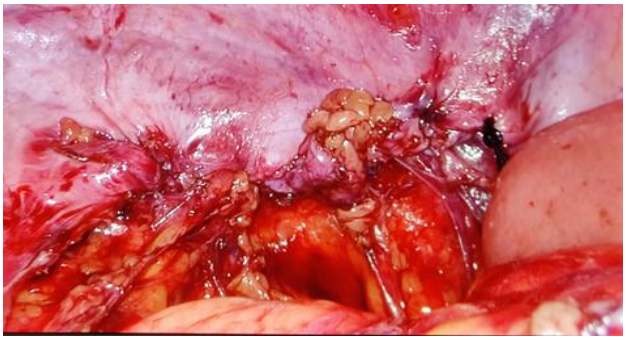

Fig 2c: primary closure

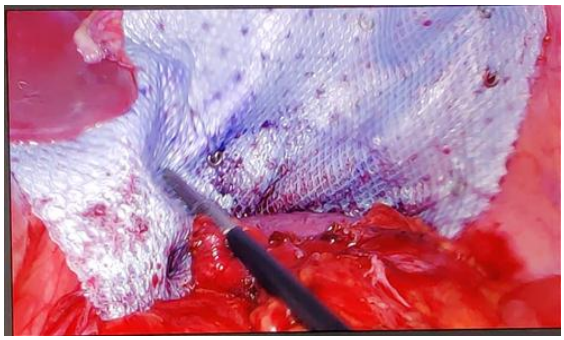

Fig 2d: 10X15 cm dual mesh

Mesh was fixed with non-absorbable helical tackers.The reverse Trendelenburg position helped to reposition the spleen in Left hypochondrium.ICD placed in the Lt thorax and ports closed.Post-operative period was uneventful. Patient was electively ventilated for 1 day.Orals started from 2ndpost-operative day. ICD was draining 500ml initially, gradually decreased over 1-week period. Patient was discharged on 7th day. CT chest 1 month later showed a fully expanded left lung with spleen in normal position. Patient was followed up for 1 year now without any complaints.

3. DISCUSSION:

Bochdalek hernia is a type of congenital diaphragmatic hernia where the defect is inpostero-lateral aspect of the diaphragm which allows the herniation of abdominal contents into the thorax, compressing the lung.It was first described by Vincent Alexander Bochdalek in the year 1848.(4) The cause of this defect is postulated as failure of closure of the Pleuroperitoneal canal which closes normally at 6-7 weeks of gestation period by the process of growth of a pluero peritoneal membrame from dorsal aspect at the root of 12th rib towards the ventral aspect and fusing with the septum transversum and dorsal mesogastrium of the oesophagus.(5)This pleuroperitoneal cannal communicates through the foramen of Bochdalek between the two cavities. This condition usually presents itself in the neonatal period with breathing difficulty. (6) In severe cases it will be difficult for the neonate to eventhrive.
It is not just the defect in the diaphragm and the pressure caused by the herniated contents which is the only pathophysiology behind this disease. Most of the times this disease will be accompanied with Pulmonary Hypoplasia, Pulmonary Hypertension and pulmonary

immaturity and deficiencies in the Surfactant and antioxidant system. All these factors contribute to the compromised respiratory status in the newborn.

Presentation of this BH in adult is rare. The incidence being 0.17% to 6%. Usually in adults the cause of diaphragmatic hernia is post trauma to chest or abdomen involving diaphragm. 70 to 75% of BH occurs on the left side than the right side because during normal development itself the left sided pleuroperitoneal canal closes a bit late compared to the rightside.(7) Right sided BH symptoms won't be much because liver does not allow much of the abdominal contents to herniated through the defect.

Initially the approach towards management was early surgery, within a window period of 24 to 48hs. Now, the recent concepts of management is more against an urgent surgical treatment, which will worsen the Pulmonary Hypertension.

Congenital diaphragmatic hernias include Bochdalek hernia, substernal or parasternalherniaofMorgagni – when the defect is onthe Rt side and Larrys hernia, when defect in Lt substernalarea,Peritoneal - pericardiac hernia. Some include eventration of diaphragm also under this classification. (8)

Aetiology: Even though quinine, thalidomide, nitrofen, phenmetrazine, polybrominated diphenyls have been used to induce CDH in various species, drugs and environmental factors as a cause for CDH in humans is still uncertain. Maternal dietary intake of vitamin A in pregnancy has been found to have a beneficial effect in decreasing the incidence of diaphragmatic hernia in a study from Japan. (9)

Relationship between CDH and retinoid signalling pathway are also being explored. In spite that CDH being a congenital defect, this disease doesn't seem to run in families. Only in less than 2% of cases there is a positive family history.Dietary intake of vitamin A during pregnancy has been found to have a protective effect against development of CDH.Genetic causes like Chromosomal abnormalities, sequence variants, copy number in CDH patients are reported in 7 to 31% cases and in such instances, there are associated other congenital defects.

Clinical presentation:Presentation in adults is usually asymptomatic,as many are able to live a normal life with a BH and hence these are detected incidentally during imaging studies or present with vague gastrointestinal symptoms like abdomen pain, GERD.(7, 10)Obstructive symptoms like vomiting, obstipation occurs when complications occur. Patients can present with respiratory symptoms like breathing difficulty, cough, wheezing.

On examination of the abdomen in case with massive herniation of abdominal contents into the chest, abdomen will be scaphoid and feel empty. Respiratory signs can range from diminished breath sounds on the affected side to absence of breath sounds and even bowel sounds can be heard in the chest.

In adults, due to its rarity, chances of misdiagnosing this clinical entity is common. In about 38% of cases CDH in adult is misinterpreted for plural effusion, lung cyst, empyema, pneumothorax.(11)Even though a chest x-ray can detect a diaphragmatic hernia, CT scan has high sensitivity to diagnose CDH and should be the investigation of choice as it gives details of defect size and contents also.

All diaphragmatic hernias should be repaired at theearliest once it has been diagnosed, because of the risk of strangulation or volvulus or perforation of its contents. Minimally invasive procedure, eitherlaparoscopically or thoracoscopic approach is safe and effective form of management since it reduces the morbidity of the surgery at the same time giving a good result.Even though via thoracoscopy repair of the defect is easy, laparoscopy has the advantage of reduction of contents and additional procedures like fundoplication or gastropexy can be performed if required.Contents of the hernia sac may vary according to the side of the hernia. On left side, stomach, small bowel, transverse colon, spleen along with omentum can herniated. (12) On the right side due to the presence of liver, colon, small

bowel, kidney omentum. After reduction of the contents, Primary closure of the defect with or without mesh reinforcement is recommended. Sometimes if the defect is very large as to not able to achieve a primary closure, then a thick meshPTFE like can be sutured to the borders of the defect all along without a primary closure. A Bochdalek hernia mostly do not have a hernial sac. (13,14)This is in contrast to para sternal hernia of Morgagni and Larry which usually has one, in which situation the sac can be either excised or can be left behind and repair done.If the lung is atelectatic and does not expand after reduction of the contents, a ICD is left insitu and can be removed postoperatively once there is a full expansion of the lung which happens in 2 or 3 days' time depending on the magnitude of the hernia.CDH can be associated with GERD, hiatal hernia and in such situations a Fundoplication procedure can be combined with the repair of hernia.A volvulus of stomach will require a gastropexy after completion of the repair.

4. CONCLUSION:

Bochdalek hernia is a congenital diaphragmatic hernia and occurrence in adult is rare. Doctors should be aware of this condition which can be easily misdiagnosed for a pulmonary pathology. This condition can be successfully treated by minimal access surgery without much morbidity and complications. Our patient is doing well 1 year after surgery without any recurrence.

CONFLICT OF INTEREST:Nil

5. ACKNOWLEDGMENT:

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