Abstract: Congenital diaphragmatic hernia (CDH) is usually diagnosed within minutes or hours after birth because of pulmonary complications and may be classified as complete or partial. Partial defects of the diaphragm are more common, including Bochdalek and Morgagni. Diaphragmatic agenesis is unusual and may be associated with pulmonary hypoplasia resulting in progressive respiratory failure and death of the neonate. Late presentation of agenesis is exceedingly rare. It usually left sided. Here we are presenting a 33 yr old male with the rarest disease i.e. Right hemidiaphragmatic agenesis and its rarest presentations i.e. Intestinal obstruction and absence of lung hypoplasia.

Keyword: INTESTINAL OBSTRUCTION, AGENESIS OF HEMIDIAPHRAGM, PROSTHETIC MESH REPAIR

AGENESIS OF RIGHT HEMIDIAPHRAGM-A CASE REPORT

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33Y/M male presented with abdominal pain, distension, vomiting and obstipation for one week with no other complaints. Had past history of RTA 3yrs back in which he had fracture tibia managed with POP. No other co-morbidities. Had binge alcohol one day prior to the onset of symptoms. General examination showed dehydration and tachycardia. Abdominal examination showed distension with tenderness over epigastrium, (R),(L) hypochondrium without guarding or rigidity, shifting dullness was present with exaggerated bowel sounds and empty rectum in per rectal examination. Bowel sounds was heard on auscultation of right side chest. With these features we thought of acute intestinal obstruction / diaphragmatic hernia on right side. We proceeded with investigations.

X-ray showed features of bowel loops in CXR on right side and multiple air fluid levels in AXR . USG Abdomen showed dilated bowel loops suggestive of intestinal obstruction.
CT-Chest and Abdomen showed features of Right diaphragmatic hernia with liver and large bowel herniating with obstruction

Revised diagnosis of acute intestinal obstruction due to traumatic diaphragmatic hernia was made and patient was explained about the same. In the meantime patient was absconded from the ward and readmitted 2 days after with abdominal pain and no features of intestinal obstruction both clinically and investigation wise and hence planned for elective hernia repair.

F/S/O Diaphragmatic hernia rt side with herniation of liver and large bowel without obstruction With posterolateral thoracotomy approach thoracic cavity explored and it revealed absence of right hemidiaphragm with liver occupying almost 2/3rd of right sided thoracic cavity with compressed right lung no bowel herniation. Right lung was not hypoplastic. Since the liver cannot be pushed into abdomen and there was no diaphragmatic remnants, prosthetic mesh was placed between the liver and lung and fixed to the interior surface of anterior, lateral, posterior chest wall.

APLASIA OF RTHEMIDIAPHRAGM PROSTHETIC MESH REPAIR
Post-operatively patient was on ventilator support for one day and weaned of the same. Started orals on 4th day, ICD. Post-operatively patient was on ventilator support for one day removed on 9th day and suture removal on 10th day. Patient doing well post operatively.

**DISCUSSION:**

Congenital diaphragmatic hernia (CDH) is a congenital malformation of diaphragm, characterized by a defect in the diaphragm along with herniation of gastro-intestinal loops and other abdominal viscera into the thoracic cavity. In case of extreme maldevelopment of diaphragm, there might be a complete agenesis of diaphragm, usually involving one side, but, sometimes both sides. Classical CDH is characterized by a left-sided posterolateral defect in the diaphragm, and named after Bochdalek who was the first to identify the defect in 1848. The right-sided Morgagnian’s CDH are very rare and reported to be 8-15%. Left-sided CDH has better prognosis as compared to right-sided CDH. Congenital hemidiaphragmatic agenesis (CHDA) is considered as one of the rare congenital malformations of diaphragm and reported to be in 6% of all CDH. Unilateral diaphragmatic agenesis is common on left side as compared to the right side (80-90% on the left side). However, in our case the hemidiaphragmatic agenesis was on right side showing rare side of occurrence of a very rare lesion. Some cases of bilateral CHDA have been reported with extremely high mortality. The etiology of CHDA is largely based on assumptions and speculation, and not well defined to date.

A failure of pleuroperitoneal membrane to close the pleuroperitoneal canal on the involved side is considered the probable etiology; however, others proposed a failure of all the precursors components of diaphragm to develop. The classical presentation of CDH is within first 24 hours of life with clinical features dependent upon the severity of pulmonary hypoplasia and pulmonary hypertension. More severe the lung hypoplasia and pulmonary hypertension, early will be the presentation and worse will be the outcome, and vice versa. The clinical features are usually respiratory distress, cyanosis and vomiting. About 10% of patients with absence of lung hypoplasia and pulmonary hypertension may escape the early presentation, as happened in our case. The patients with CDH may present in infantile age or thereafter; presentation of CDH in adults and even senile persons have frequently been reported in literature. The presentation of CHDA is somewhat similar to the CDH and is considered, by many authors, as an extreme variety of Bochdalek CDH. Similarly as for CDH, CHDA may also appear in adults but the overall incidence is very low.

CHDA may be differentiated into partial and complete agenesis with regard to the defect. In case of partial diaphragmatic agenesis a small rim of diaphragm may be present especially on posterior aspects, whereas, in case of complete hemidiaphragmatic agenesis, no diaphragmatic remnant is present as was found in our case. Chest radiograph cannot specifically diagnose the CHDA but is a modality of choice in case of CDH. CT scan and MRI are important diagnostic modalities for the delineation of the diaphragm and its abnormalities. In our case diagnosis of CDHA was made intraoperatively by the absence of right side hemidiaphragm. The surgical repair of the lesion is very difficult owing to completely absent diaphragmatic tissue or if present not suffice to accomplish the repair. Various authors used different treatment options including, prosthesis, abdominal wall and chest wall muscle flaps, suturing the chest margins with liver, use of pre-renal fascia and some advocated no repair at all. In our case we have done a prosthetic mesh repair with uneventful post-operative period.

Tzelepis et al reported absence of left hemidiaphragm is asymptomatic 22 yr old man. The asymptomatic presentation of Adult diaphragmatic hernia (ADH) suggested that no surgical treatment is necessary as the defect was large enough to permit free movement of visceral structures thus, lessening the chances of strangulation and incarceration. However Sheehan et al reported a 66 yr old woman presented with 3 day history of classical features of large bowel obstruction. At emergency laparotomy, the transverse colon and splenic flexure were located in left hemithorax. The entire left hemidiaphragm was absent and there was no diaphragmatic remnants visible. This was the first reported case of true ADH in an adult and it was associated with lung hypoplasia. Traveline et al reported a 39 yr old woman with asymptomatic ADH and lung hypoplasia discovered accidentally following a routine chest x-ray carried out for another reason. The patient was managed conservatively. Singh and Bose et al reported a case of partial left sided defect that was repaired by suturing a synthetic mesh to the diaphragmatic remnants. No long term follow up data were available to access success of treatment and possible complications. Our case differed from the patients described in previous studies on having the right hemidiaphragmatic agenesis with features of intestinal obstruction and absence of lung hypoplasia which all are rarest of the occurrence and presentations.
References:


