Late Presentation of Congenital Diaphragmatic Hernia with Malrotation of Midgut in Adult

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Abstract

18 years old female admitted with complaints of abdominal pain, breathlessness and constipation for 2 days. On examination patient had tachycardia, tachypnea, abdomen symmetrically distended with tenderness on left side. Heart sounds S1 and S2 was heard on right side of the chest, decreased air entry on left side. X ray and CT chest showed diaphragmatic hernia with bowel obstruction. Patient was taken for emergency procedure content reduced and defect was closed, respiratory support given. Clinical and radiological investigations helps in diagnosing the congenital diaphragmatic hernia early and intervention before respiratory failure increases the survival rate in both neonates and adults.

Keywords: Malrotation of midgut, respiratory distress, adult, x ray.

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INTRODUCTION

The aim of this case report is to provide information on the clinical symptoms and signs, diagnosis and management of the late presentation of congenital diaphragmatic hernia in adult. Congenital diaphragmatic hernia is a major malformation occasionally found in newborns and babies. Congenital diaphragmatic hernia is defined by the presence of an orifice in the diaphragm, more often to the left and posterolateral, which permits the herniation of abdominal contents into the thorax. In our case the patient presented with small bowel obstruction due to diaphragmatic hernia Bochdalek type with malrotation of midgut.

CASE REPORT

The subject is an 18 years old female admitted with complaints of abdominal pain and breathlessness for 2 days with history of constipation and has not passed flatus for 2 days. Patient was first born child and neonatal period was uneventful with no previous significant medical and surgical history. On general examination patient is conscious, afebrile. Tachycardia and tachypnea present, abdomen symmetrically distended with tenderness in left hypochondrium, lumbar and iliac fossa region. On Cardiovascular system examination S1 and S2 heard on right side of the chest, Respiratory system examination revealed decreased air entry and bowel sounds heard on left side.

X ray chest and abdomen showed bowel shadows in left chest, on CT chest showed mediastinal shift to right side, collapse of left lung, bowel loop in left chest. CT abdomen revealed Ileum, transverse colon proximal descending colon herniating into the thorax. Jejunum, Transverse & ascending colon appeared dilated, Distal descending colon is collapsed and recto sigmoid on right side.

Fig-1: Plain X ray of chest and abdomen showing bowel loops in left hemithorax
Due to small bowel obstruction and respiratory distress, the patient was taken for emergency procedure through left subcostal incision. The peritoneal cavity was entered, and it was found that the jejunum was dilated, ileum, cecum with appendix, and transverse colon were found herniating into the thorax. The content was reduced, and a defect of size 6.5 cm x 4 cm was noted in the posterolateral surface of the diaphragm, which was closed by using non-absorbable suture material prolene in two layers.

Intercostal drainage tube was inserted on the left side. The patient was on mechanical ventilation for 4 days after that she was weaned gradually and oral feeds started.

**DISCUSSION**

Incidence is 1 in 2000 to 5000 live births and common in the left side around 80% and bilateral in < 5% of the cases. Hernia sac is present in 20% and overall survival rate 70 to 90%. Incidence in adults is 0.1 to 6% and most cases are sporadic.

**Associated anomalies**

CNS lesions, Omphalocele, Esophageal atresia, cardiovascular lesions, Part of trisomy 21, 13, 18.

**Pathophysiology**

CDH is due to failure of closure of the pleuropertitoneal canal in the developing fetus. Usually, the pleuropertitoneal cavity becomes separated by the developing membrane during 8 to 10 weeks of gestation. When this process fails, the pleuropertitoneal canal does not close, and a posterolateral defect in the diaphragm results. It impairs growth of the ipsilateral lung and causes pulmonary hypertension.

**Clinical features**

Respiratory distress at birth (tachypnea, grunting, use of accessory muscles – cardinal sign), Dyspnea, Cyanosis, Scaphoid abdomen, Increased chest wall diameter, Bowel sound heard in chest, Decreased/absent breath sound on side of hernia, Shifting of apex beat to the opposite side.

**Surgical treatment**

Mostly surgery was done 48 hrs of birth after stabilization. Preferred approach is subcostal abdominal incision both laparoscopic and thoracoscopic repair can be done. Content reduced and defect closed by interrupted non-absorbable sutures, if present the hernia sac should be excised. If primary closure is not possible – abdominal/thoracic muscle flaps can be used. For tension free repair native tissue / Prosthetic material
Synthetic patch (GORE TEX) and biological mesh are used.

**Prognosis**

Overall survival 67% in newborns and poor prognosis in major anomalies, severe pulmonary hypoplasia - need of ECMO. In adults prognosis is good if surgery is done before developing respiratory complications.

**CONCLUSION**

Congenital diaphragmatic hernias are usually diagnosed in neonatal period it may present as late in adults with incidence of 0.17 to 6%. Clinical and radiological investigations help in diagnosing the condition, early intervention before respiratory failure increases the survival rate in both neonates and adults. In our case early diagnosis and early intervention saved the patient before developing complications.

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