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Diaphragmatic Hernia with Atypical Presentation - A Case Series

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Summary:

Congenital Diaphragmatic Hernia is one of the most challenging diagnosis faced by pediatric surgeons. From the time of its first anatomic description more than 300 years ago, CDH has carried a high mortality rate. We aimed to review patients who presented with hernia of diaphragm during the last six months. In this retrospective study, the medical records of three patients treated for diaphragmatic hernias who were admitted to Rajshahi Medical College Hospital between July 2012 and December 2012 were analyzed. Three patients with age of 45 days to 7 years were included in the study. Male to female ratio was 1:2. All patients had left-sided diaphragmatic hernia. Chest X-ray was obtained from all patients which was diagnostic. One patient needed thoracotomy incision. No patient required mesh repair. The mean hospitalization time was 14 days. There was no postoperative death. Diaphragmatic hernia is an uncommon and challenging situation for the surgeon. Prompt diagnosis and treatment prevent serious morbidity and mortality associated with complications such as gangrene and perforation of herniated organ.

Keywords: Diaphragmatic Hernia, Diaphragm, Gastric volvulus, colonic gangrene

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Introduction:

Congenital diaphragmatic hernia (CDH) is a developmental defect of the diaphragm that allows abdominal viscera to herniate into the chest.[1]

The embryologic development of the diaphragm involves multiple, complex

cellular and tissue interactions. The fully developed diaphragm is derived from four distinct components: (1) the anterior central tendon forms from the septum transversum, (2) the dorsolateral portions form from the pleuroperitoneal

membranes, (3) the dorsal crura evolve from the esophageal mesentery, and (4) the muscular portion of the diaphragm develops from the thoracic intercostal muscle groups. The precursors of diaphragmatic structure begin to form during the fourth week of gestation with the appearance of the peritoneal fold from the lateral mesenchymal tissue. At the same time, the septum transversum forms from the inferior portion of the pericardial cavity. The septum transversum serves to separate the thoracic from the abdominal cavities and eventually forms the central tendinous area of the fully developed diaphragm[1-2].

Anatomically, the right side closes before the left. Muscularization of the diaphragm appears to develop from the innermost muscle layer of the thoracic cavity, although it has been proposed that the posthepatic mesenchymal plate is a possible source of muscular tissue. Posterolaterally, at the junction of the lumbar and costal muscle groups, the fibrous lumbocostal trigone remains as a small remnant of the pleuroperitoneal membrane and relies on the fusion of the two muscle groups in the final stages of development for its strength. Delay or failure of muscular fusion leaves this area weak, perhaps predisposing to herniation[3]

Bochdalek first described this area of the posterolateral diaphragm in 1848, and it is for this reason that the most common site for CDH bears his name[1-3] Other types include Morgagni hernia, diaphragm eventration and central tendon defects of the diaphragm. Bochdalek hernia is the result of a congenital defect in the posterior costal part of the diaphragm in the region of the 10th and 11th ribs, which allows free communication between the thoracic and abdominal cavities [4] .

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CDH is estimated to occur in 1 out of 2000 to 5000 births. Females appear to be more commonly afflicted than males. Affected neonates usually present in the first few hours of life with respiratory distress that may be mild or so severe as to be incompatible with life [5,6]. With the advent of antenatal diagnosis and improvement of neonatal care, survival has improved but there still remains significant risk of death and complications in infants with CDH[7]. The overall mortality is still high at most centers[8].The major cause of death is due to two complications: pulmonary hypoplasia and pulmonary hypertension[9]. Experts disagree on the relative importance of these two conditions, with some focusing on hypoplasia, others on hypertension[10].

Because herniation occurs during a critical period of lung development when bronchial and pulmonary artery branching occurs, lung compression by the herniated bowel results in pulmonary hypoplasia. With increasing severity of lung compression, there is a corresponding decrease in the bronchial branching resulting in a reduction of generations of bronchi and lung tissue. In addition, arterial branching is reduced and there is muscular hyperplasia of the pulmonary arterial tree [10-11].

Current ideas view the defect to be an inherent abnormality of lungs which results in a secondary abnormality of the diaphragm or as a failure of the diaphragm to separate the Pleuroperitoneal canal into thorax and abdomen before the midgut returns from umbilicus.The resultant abnormality leads to disordered lung growth. Both lungs are affected,th ipsilateral mo so than the contralateral. Affected infants are bon with a complex interface of pulmonary hypoplasia and pulmonary hypetension. Pulmonary hypoplasia can be severe enough to preclude life outside the womb, while successful management of pulmonary hypertension can lead to a fruitful life[12-14].In this report, we present three cases of diaphragmatic hernia with atypical presentation.

Objectives

Clinical presentation and management of three cases of congenital hernia of diaphragm were discussed to highlight challenges in diagnosis and handling of complications.

Patients and Methods

Three patients with the diagnosis of congenital diaphragmatic hernia treated in Rajshahi Medical College hospital between July 2012 and December 2012 were enrolled to this study. The mechanism, duration of complaints, clinical presentation, kind of visceral herniation, surgical repair, and outcome of the patients have been scrutinized. Diaphragmatic hernia repair and organ reduction were performed in all patients through abdominal approach except one,that required additional thoracic approach.

Case Report-1

A 7-years-old female child presented to our institution with complaints of abdominal pain, non passage of stool for five days, abdominal distension, frequent bilious vomiting, and respiratory distress for three days. The pain was progressive and colicky in nature and became more severe with no definite radiation or shifting. The vomiting was bilious and foul smelling and abdominal distension was progressive. Past medical history was significant and since birth she has been suffering from recurrent respiratory distress especially after running, playing, jogging and was relieved by taking rest. She also suffered from intermittent gastrointestinal symptoms like epigastric discomfort and postprandial colicky abdominal pain. She visited consultant pediatrician and received medication like antibiotics, nasal drops, antihistamines, gastroprokinetcs and antiflatulent for recurrent respiratory tract infections.

Her perinatal history was insignificant and she was delivered by caesarian section and her birth weight was 3.25 kilograms. On general physical examination the patient was restless,dehydrated,febrile, tachypnic with granting respiration. she was moderately anemic, moderately cyanosed and nonicteric. Abdominal examination revealed tense, moderately distended and tender abdomen. Tenderness was more marked over epigastric and left hypochondriac region where rebound tenderness was also present. Abdomen was resonant on percussion, absent bowel sound and no sign of ascites present. Digital rectal examination revealed normal findings. On chest examination asymmetry found between left and right hemithorax, the point of maximum cardiac pulsation and trachea shifted to right. Breath sound was normal on right but present only apex on left side. Laboratory investigations revealed neutrophilic leucocytosis and mild hypokalaemia. Abdominal and chest radiographs in erect postures revealed shifted trachea and mediastinum to right, gas distended bowel loops with air fluid levels in left hemithorax.

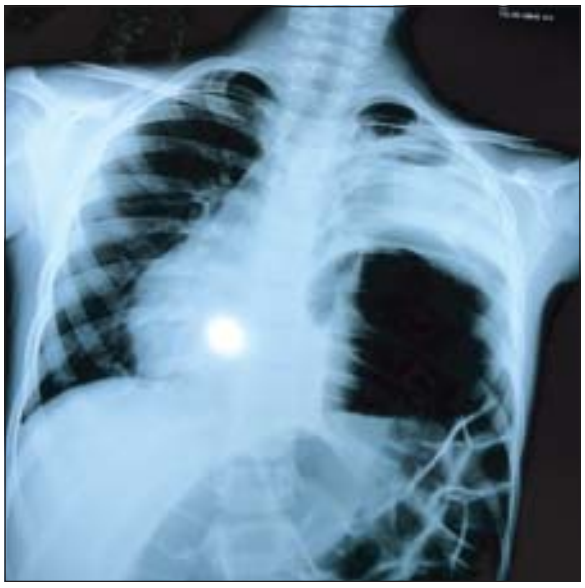

Fig.-1: *Chest radiograph showing ill-defined left dome of diaphragm and multiple air-fluid level in mid &lower zone of left hemithorax and mediastinal shift to right.*

The patient was provisionally diagnosed to have obstructed diaphragmatic hernia. After resuscitation emergency surgery was arranged. abdomen was approached through left subcostal incision. Moderate amount of hemorrhagic fluid found in abdominal cavity and small intestine, caecum, ascending and transverse colon were hugely distended. The left dome of diaphragm was highly elevated. The stomach, greater part of intestine and spleen with its hilum was found in the left hemi thorax. Part of transverse colon was protruded through an aperture of the diaphragm. The viscera were gently reduced from left hemithorax. The reduction of transverse colon was difficult as it found herniated through the diaphragmatic defect and finally it was irreducible. it required an additional thoracic approach and a left sided thoracotomy done. There was a defect on left dome of diaphragm which was about 5 cm in diameter. Herniated intrathoracic part of transverse colon was gangrenous.

After reduction of abdominal contents a tension free diaphragmatic repair was done with 1/0 prolene and the laxed part of diaphragm was plicated. Chest wall was closed, keeping a water seal drain in situ.

The gangrenous part of transverse colon was resected and a double barrel colostomy was performed on right hypochondrium. Abdomen was closed in layers. In immediate post operative period the patient needed ventilatory support in intensive care unit for 24 hours. Later she could maintain a satisfactory oxygen level and post operative chest radiographs revealed optimum lung expansion.

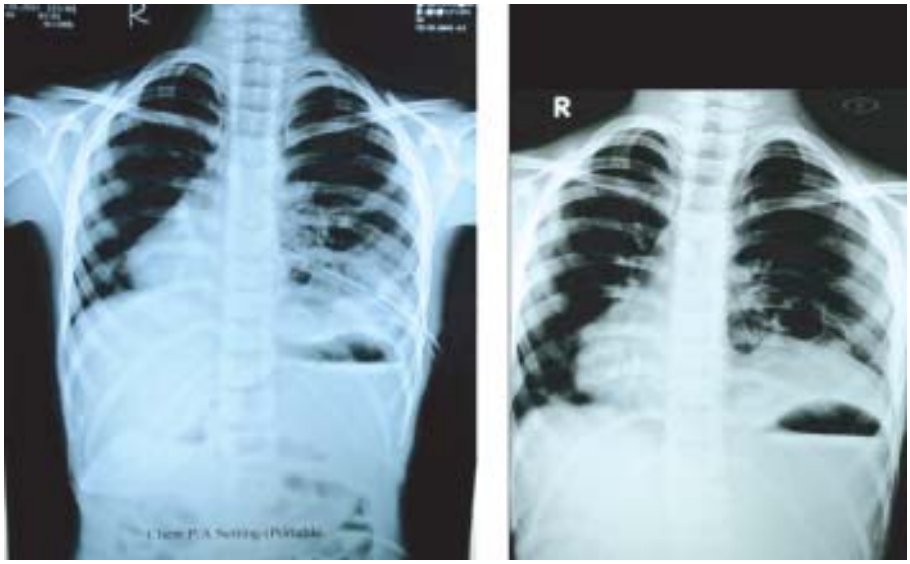

Fig-2: *Post operative day chest radiographs before and after removal of chest drain tube showing extend of lung expansion.*

She was recovering nicely but on fifth post operative day dehiscence of abdominal wound as evident. It was managed by regular dressing and secondary wound closure. She was discharged after three weeks. She underwent closure of colostomy two months later. She had a normal respiratory and gastrointestinal status on next sixth months follow up.

Case Report-2

Two-and a half months old female child presented with history of recurrent respiratory distress and non-bilious vomiting off and on since 2 months. On examination,fullness was present in the epigastrium and left hypochondrium. There was no visible peristalsis. Breath sound was decreased on left lower hemithorax. Laboratory investigations were normal. Chest radiograph revealed multiple air-fluid level into the left hemithorax.

Intraoperatively there was a left posterolateral congenital diaphragmatic hernia with a sac and with organoaxial volvulus of the stomach with the pylorus herniating into the left hemithorax posterior to the fundus.

The contents were reduced, volvulus corrected, sac excised and diaphragmatic defect repaired. Three-point gastropexy was done to anterior abdominal wall. Patient had an uneventful recovery.

Over the last six months she attained a normal growth but experienced three episodes of respiratory tract infections, which improved by medical management. Radiologically there was no evidence of recurrence.

Case Report – 3

A 45 days old male baby presented with the complaints of respiratory distress, and occasional bilious vomiting since birth. The baby was delivered by caesarian section after full term pregnancy. On general physical examination the patient was vitally stable. Abdominal examination revealed mildly distended and tender abdomen in all quadrants with normal bowel sounds. A digital rectal examination revealed normal findings. Laboratory investigations were within normal limits except mild hypokalaemia. Radiographs of abdomen and chest delineated intestinal air shadows in the left hemithorax.

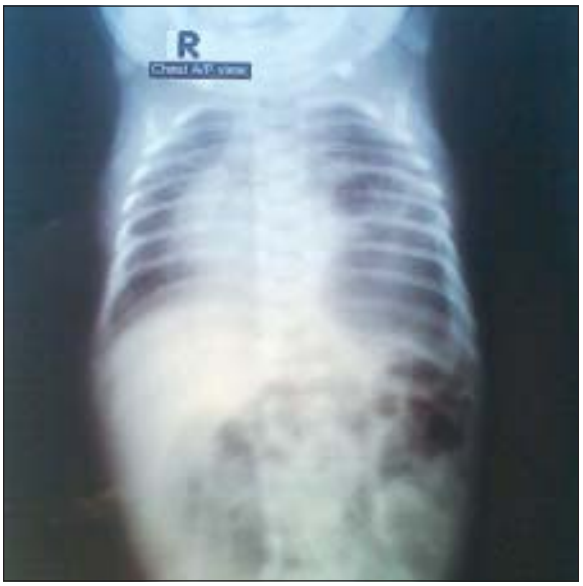

Fig.-3: *Left dome of diaphragm with multiple air-fluid level.*

An exploratory laparotomy was performed by left subcostal incision. The part of stomach, part of small intestine, spleen was found within the left hemithorax herneating through a large defect in posterolateral part of left dome of diaphragm. The left kidney was also found within the thoracic cavity. The stomach, spleen and small intestine were reduced and the kidney was mobilized from the thoracic cavity.

The defect of the diaphragm was repaired with non-absorbable suture material. Patient had an uneventful post operative recovery.

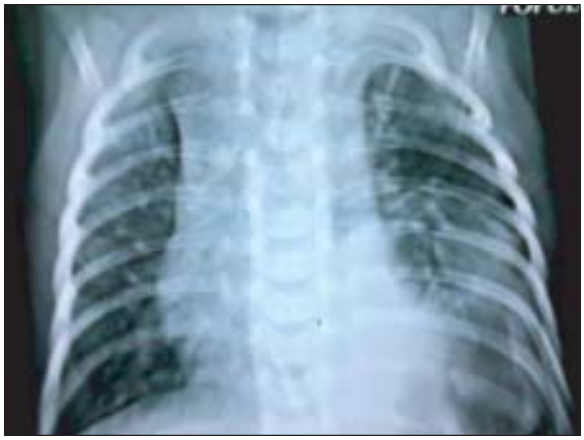

Fig.-4: *Chest radiograph suggestive of diaphragmatic hernia.*

The patient maintained a satisfactory oxygen level and post operative radiograph showed satisfactory lung expansion. The patient was discharged on 8th post operative day. Over the next six months follow up, there was no evidence of recurrence but she developed three episodes of respiratory symptoms,that required medical treatment.

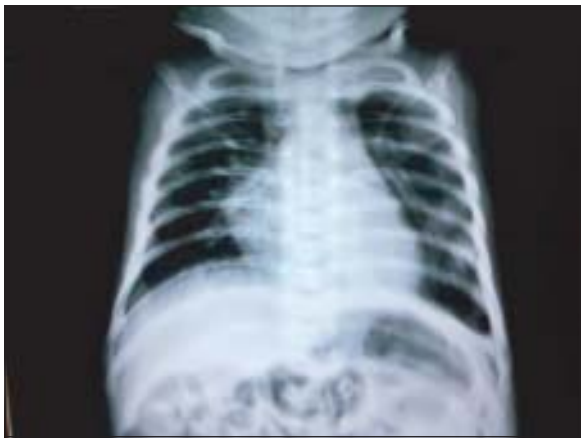

Fig.-5: *Post operative chest radiograph with lung expansion.*

Discussion:

Congenital diaphragmatic hernias mainly present in the neonatal period and are associated with a mortality that has not changed much despite the advances made

in critical care[15]. Rarely, these hernias present later in life, some even in adulthood. There are numerous reports of CDH presenting after infancy. These patients are either asymptomatic or have minimal respiratory symptoms, possibly because the lungs are not hypoplastic. Late-presenting CDH is often difficult to diagnose, and

delays in treatment are common. Moreover, the detection of congenital diaphragmatic hernia may be missed because of intermittent herniation of the abdominal viscera into the thoracic cavity and wide variability in presentation[16]. In addition, Bochdalek hernia may be mistaken for left middle lobe collapse, pneumonic consolidation, pericardial cyst, sequestration of the lung, mediastinal lipoma, or anterior mediastinal mass[17].this might be the reason behind late detection of first case by attending clinician.

Late presenting CDH may present with gastrointestinal tract symptoms that may include intermittent abdominal pain, vomiting, and dysphagia. Respiratory symptoms usually include dyspnea and chest pain. Symptoms may be intermittent or acute depending on the extent of herniation of abdominal viscera into the thorax. An acute presentation is usually due to incarceration, obstruction, or strangulation of the herniated viscera . Diagnosis is ascertained by a combination of chest X-rays, CT and magnetic resonance imaging (MRI), as well as upper gastrointestinal and bowel double-contrast studies[15,18].

A careful analysis of chest films and a thorough search for connecting bowel segments passing through the diaphragmatic defect may help to avoid incorrect diagnosis and an undesirable delay in treatment[19].

We report three cases of delayed presentation of a potentially life-threatening CDH. 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[20].

Transabdominal and transthoracic approaches have been recommended in Congenital Diaphragmatic hernia repair. The abdominal approach is easily performed through an upper abdominal incision. But when the patient has findings suggesting intestinal strangulation and irreducibiity of the hernia, transthoracic repair of diaphragmatic hernia might be required in addition to laparotomy [21].

Gastric volvulus is rare as stomach is secured in place by the gastrophrenic ligaments, esophageal hiatus, retroperitoneal fixation of the duodenum, short gastric vessels and gastrocolic ligament. It occurs only when these attachments are lax or absent. Secondary causes are eventration ofthe diaphragm, diaphragmatic hernia, congenital bands, wandering spleen and paraesophegeal hernia. Primary volvulus is mainly mesentericoaxial in type while secondary volvulus is mainly organoaxial[21,22]. The clinical symptoms depend on the extent or degree of rotation and gastric outlet obstruction. Operative treatment includes reduction, correction of underlying cause and gastropexy. is not necessary for a successful outcome in secondary volvulus[22]. However, many authors prefer to fix the

stomach even after the correction of underlying defects[15,23].

Following surgical repair of the hernia abnormalities of respiratory function was noticed in most of the cases, which is common as reported by other authors. Significant improvement of lung function is expected over subsequent years of life[12,24].

In most centers Congenital Diaphragmatic Hernia has a mortality rate of 30-60%,[8,12]. Outcomes in present study were more favorable due to absence of other congenital abnormalities. Individual rates vary greatly dependent upon multiple factors; size of hernia, organs involved additional birth defects or genetic problems, status of lung growth, type of treatments, timing of treatments and complications[15,24].

In conclusion, even though diaphragmatic hernia is rare, prenatal USG and post natal chest radiography can diagnose most of cases. Prompt diagnosis and treatment can prevent serious morbidity and mortality associated with complications.

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