Case report

Congenital diaphragmatic hernia: A rare cause of obstructive jaundice

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Abstract

Congenital Diaphragmatic Hernia in adults are exceedingly rare. They have been reported to cause dyspnea, gastric reflux and intestinal obstruction . We present the case of a young male with obstructive jaundice secondary to bochdalek hernia of the right hemi-diaphragm. We discuss the aetiologies , presentation and treatment of the disorder.

Case History

A 29 year old male presented to hospital with complains of recurrent jaundice (serum bilirubin 27 direct 17 ), icterus, nausea with on and off abdominal pain. There was no history of trauma. A plain chest xray taken on admission demonstrated a right lung collapse and an elevated right hemi-diaphragm. Ultrasonography of abdomen performed on the same day demonstrated intrahepatic bile duct dilatation and herniation of liver, gall bladder, intestines in right side of thorax.

Due to the dual findings of the chest x-ray and ultrasonography, double contrast computed tomography (CT) of the abdomen and thorax was performed (fig 1). This demonstrated that the right lobe of liver, gall bladder, kidney and intestinal loops , mesentery was within the right chest cavity with kinking of bile duct with collapse of right lung . It also demonstrated intrahepatic duct dilatation as a result of anatomical distortion. There was no obvious pathology in the spleen, left kidney , pancreas ,left hemi-diaphragm.

Computed tomography of the abdomen and thorax demonstrating Mild prominence of intra-hepatic biliary radicals in right lobe of liver, with altered signal intensity on right lobe of liver suggestive of obstructive biliopathy. Dilated left hepatic duct with abrupt narrowing of right hepatic duct at the confluence suggestive of biliary stricture MRI Abdomen
MRI Abdomen showing large diaphragmatic hernia on right side with herniation of bowel loops, fat right lobe of liver, gall bladder and right kidney with underlying collapse of lung.

- Obstructive jaundice reduced within a week of admission without any intervention. It was decided elective repair of diaphragmatic hernia was needed due the risk of bowel strangulation or further biliary complications.

**Intra-operative findings**

- Intra-operatively thoracotomy and paramedian incision was taken for better exposure. Right lobe of liver was found to be cirrhotic with left lobe hypertrophy.
- E/o large defect of size 10×7cm through which right lobe of liver, gall bladder and right kidney were seen to be herniating into thorax. Right lobe of liver was cirrhotic.
- E/o kink in right hepatic duct just proximal to junction with left hepatic duct causing biliary obstruction.
- Right lobe of liver, right kidney & bowel loops reduced into abdominal cavity,
- Cholecystectomy done.
- After reduction of the hernial contents, defect was closed and reinforced with an on laying mesh. Primary closure of the abdominal cavity was done.
Intraoperative photograph showing large defect in the diaphragm with herniation of right lobe of liver and gall bladder in right hemithorax with cirrhotic right lobe of liver.

Postoperatively Patient electively ventilated.

Patient could not be weaned off ventilator due to collapsed right lung and pulmonary hypertension secondary to lung hypoplasia.

On day 3 patient had deranged Liver Function Tests Serum Bili-rubin 12.3(Direct 7.3)

SGOT(354)
SGPT(447)

Died due to acute on chronic liver Failure on postoperative day 4.

Discussion

Congenital diaphragmatic hernia (CDH) is very rare with an incidence of between 1 in 2,500 to 12,000 live births.\textsuperscript{1} The majority (85%) occur in the postero-lateral area of the hemi-diaphragm (Bochdalek hernia), resulting from the persistence of the pleuro-peritoneal canal owing to the non-fusion of the pleuro-peritoneal folds during the eighth week of gestation.\textsuperscript{2} This is almost always an emergency as the vast majority present with respiratory distress and sepsis in the newborn.

CDH in adulthood is exceptionally rare with a rate of 0.17% found incidentally on CT.\textsuperscript{3} Complications resulting from Bochdalek hernias in adults can include bowel obstruction,\textsuperscript{4} gastric reflux\textsuperscript{5} and pancreatitis.\textsuperscript{2} These are almost exclusively left-sided hernias. There have been fewer than 100 cases of adults presenting with a complication of congenital diaphragmatic hernias in the literature and none presenting with obstructive jaundice. There are two reported cases of neonates presenting with obstructive jaundice secondary to a Bochdalek hernia.\textsuperscript{2,8}

Here we present the case of a young male with a rare congenital right-sided Bochdalek hernia. There was a delay in the diagnosis because of the initial presentation of recurrent jaundice.

Investigation by chest x-ray alone is not enough to make the diagnosis although a chest radiograph after nasogastric tube placement could have expedited the diagnosis. CT provided a detailed assessment of the
anatomy and a cause for the obstructive jaundice was established.

Even though CDH has been well described in the literature, the incidence of clinical presentations in adulthood is exceedingly rare and this is the first case of it leading directly to obstruction of the biliary system. Open mesh repair of the hernia is the gold standard treatment option with evidence to back up its safety and efficacy. There are very few documented cases of successful laparoscopic repair of an adult CDH in the literature. One case series suggests that this is a safe alternative treatment modality for CDH presenting past infancy. Current recommendations are that all adults with a CDH undergo repair in order to avoid complications. Possible postoperative complications include abdominal compartment syndrome although there is no evidence in the literature to support this.

Conclusions
CDH is exceedingly rare in adulthood and has been reported to become symptomatic in only a handful of cases. However, since its presence can lead to serious adverse events such as acute intestinal obstruction, or in this case obstruction of the biliary system, it should be investigated fully and repaired rapidly. The diagnosis should be considered in any patient presenting with abdominal pain and an unexplained consolidation on a chest x-ray.

References