Rare Case Report – Congenital Diaphragmatic Hernia Presentation in Adult

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Abstract Congenital diaphragmatic hernia (CDH) which mainly occurs in the newborn or in childhood with severe respiratory distress and high mortality, is rarely found in adults (Yamaguchi et al. *Ann Thorac Cardiovasc Surg* 8:106–108, 2002; Dalencourt and Katlic *Ann Thorac Surg* 82:721–722, 2006; Fraser et al. *Endosc Percutan Tech* 19:e5–e7, 2009; Kanazawa et al. *Surg Today* 32:812–815, 2002). These patients are been accustomed to adjust their lifestyle to manage symptoms associated with frank herniation of the large bowel and liver inside the diaphragmatic hernial sac. Bowel above the liver surface especially the transverse colon is suggestive of a Chilaiditi's syndrome in these group of patients. Diagnostic laparoscopy plays an important role for diagnosis of diaphragmatic hernia in some cases over other investigations like CT scan and ultrasonography. Chilaiditi's syndrome has no surgical line of treatment but a symptomatic diaphragmatic hernia requires surgical correction. Liver as the main hernial content has been reported only in three cases throughout the world (Goh et al. *Am J Surg* 194: 390–391, 2007; Luo et al. *Hepatobiliary Pancreat Dis Int* 6: 219–221, 2007; Bosenberg and Brown *RA Curr Opin Anaesthesiol* 21: 323–331, 2008). A case of a 27 year old female patient

presenting with a symptomatic congenital diaphragmatic hernia is reported.

Keywords Diaphragmatic hernia

Case Report

A 27 year old female with three normal deliveries and routine lifestyle, presented with pain in the abdomen and severe breathlessness. Patient was symptomatic since day 1. Pain which was dull aching increased to shooting type. Breathing was abdominothoracic with a respiratory rate of 36/min; pulse was 120/min, B.P. 80/60 mm of Hg. There was abdominal tenderness in the epigastric and umbilical region with oxygen saturation of 84%.

Clinical diagnosis of peritonitis with septicaemia was made. Patient was resuscitated in the ward with 100% O₂ and IV fluids with propped position. Patient settled and O₂ saturation came up to 94% in 2 h.

X-ray abdomen and chest standing showed no free gas under diaphragm but right dome of diaphragm was elevated with colonic shadow visible over the liver surface area (Fig. 1).

As patient responded well to conservative line of treatment and clinically was not suggestive of peritonitis it was decided to continue the treatment and investigate further.

A CT scan abdomen and pelvis (IV and oral contrast) was reported by the radiologist as a case of Chilaiditi's syndrome.

The diagnosis was confirmed with a diagnostic laparoscopy which showed a frank diaphragmatic hernia on the right leaflet of the diaphragm anteriorly. The defect was 8 × 8 cm through which the right lobe of liver and transverse

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Fig. 1 X-ray chest showing right diaphragmatic hernia

colon were ascending into the sac. Right side of diaphragm showed no movements on laparoscopy.

The high abdominal pressure during the laparoscopy surgery did not favour a good repair of the diaphragm; hence the patient was posted electively for an open diaphragmatic hernia repair, by the Chevron incision, under GA.

The defect was only covered by a pleuroperitoneal membrane with good diaphragmatic muscles around the defect. The right lobe of the liver and transverse colon were pulled down (Fig. 2) from the sac inside the abdominal cavity using haemostats, after which diaphragmatic movements were well established. The sac was completely excised the diaphragm sutured from inside to the intercostal space taking care to see that each interrupted number one prolene suture went through and anchored to the intercostal

Fig. 2 Suturing the defect of diaphragm

muscle. An ICD tube was kept inside the pleural cavity before closing the defect.

Postoperative period was uneventful other than pain at the site over the anchoring sutures to the costal margin of the diaphragm (Fig. 3).

Discussion

Congenital diaphragmatic hernia (CDH) has an incidence of 1:3000–1:5000 per live births [7]. It is believed to be caused by failure of diaphragmatic closure, and usually presents with respiratory distress, and 40–50% of mortality in the neonate [4]. Adult presentation is rare [8]. Clinically, right-sided diaphragmatic hernias are less common than the left-sided ones. The percentages of right-sided CDH and left-sided CDH vary, with published incidences ranging from 8 to 24% right-sided CDH, 73%–90% left-sided CDH [9]. Additionally, majority of herniated organs are the omental fat, bowel, spleen, stomach, kidney, and pancreas. Liver and colon as the herniated organ is extremely rare. This may be owing to the protective effect of the liver on the right side [5]. Another theory suggests that right-sided hernias rarely occur because the right side of the pleuroperitoneal canal closes earlier [10]. CDH is usually recognized during the neonatal period with respiratory compromise and pulmonary hypoplasia. An adult with CDH may present with a wide range of acute or chronic respiratory or gastrointestinal symptoms or may be completely asymptomatic [11]. Imaging is essential for definitive diagnosis of as the clinical symptoms are frequently

Fig. 3 Postoperative x-ray

vague and non-specific. A chest radiograph which demonstrates gas or fluid-filled viscera above the diaphragm may reveal that a hernia contains the stomach, or the large or small bowel. Diagnosis is usually difficult if the herniated organs do not contain gastrointestinal tissue [12]. The herniated part of the liver was initially suspected to be a pulmonary lesion. It has however been reported that the multidetector row CT scan with coronal, sagittal and 3-dimensional reconstructions raises the CT's sensitivity to CDH [5].

Hernia repair is typically performed through the thoracic or abdominal route. Small diaphragmatic defects, are usually repaired by primary repair with non-absorbable sutures [1]. For large defects, prosthetic patches or tissue-engineered grafts are used to avoid causing excessive tension after repair [2, 7]. With the development of surgical techniques, operations of CDH occur through minimally invasive techniques such as laparoscopy or thoracoscopy [3].

Conclusions

A rare case of right-sided CDH of the liver and colon in an adult is presented. X-ray and CT were significant for making the diagnosis. However the pre-operative diagnosis of CDH, especially for rare cases, is difficult to achieve on OPD basis. Rare hernias like this should be kept in mind when coming to a diagnosis [6].

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