ORIGINAL RESEARCH PAPER

Paediatric Surgery

IDIOPATHIC HYPERTROPHIC PYLORIC STENOSIS WITH CONGENITAL DIAPHRAGMATIC HERNIA – A RARE ASSOCIATION

KEY WORDS: Idiopathic Hypertrophic Pyloric Stenosis, Congenital Diaphragmatic Hernia, rare association

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ABSTRACT

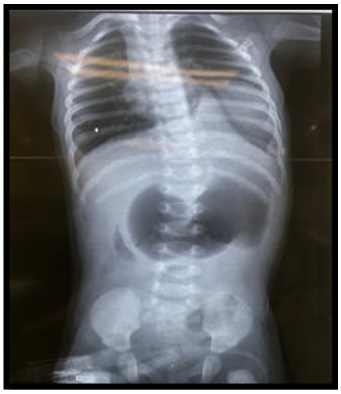
Idiopathic Hypertrophic Pyloric Stenosis (IHPS) has been associated with anomalies (6 to 12.1%) 1,2but has a rare association with Congenital Diaphragmatic Hernia (CDH).The association can be genetic, though it is usually sporadic. We present a case report of a 1 month child with non bilious projectile vomiting with failure to thrive. Radiological evaluation showed IHPS with left CDH which was surgically corrected.

Presentation of IHPS may be atypical when associated with CDH. Early recognition of concomitant anomalies will reduce morbidity in these patients.

Case:

5 week old full term first born male child, on breast feeds, presented with vomiting since 2 weeks. The vomiting was gastric, projectile in nature and immediately after feed. There was history of multiple episodes of respiratory tract infection treated by oral antibiotics. The mother also gave history of significant weight loss as also lethargy and irritability since 2 weeks. Two antenatal ultrasonographies were normal. The perinatal history was not contributory.

The child was lethargic, drowsy, dehydrated and tachypneic on general examination. Anterior fontanelle was depressed with delayed retraction on skin pinch test. On per abdominal examination, there was a soft 2*2cm 'olive' palpable in right upper quadrant. Air entry was decreased on left side. A visible gastric peristalsis going from left to right was visible Haemogram was normal. Serum electrolytes showed hypokalemia and hyponatremia. Renal biochemistry showed an increase in serum creatinine and blood urea nitrogen levels. ABG showed metabolic alkalosis. Xray chest and abdomen showed a elevated left dome of diaphragm.



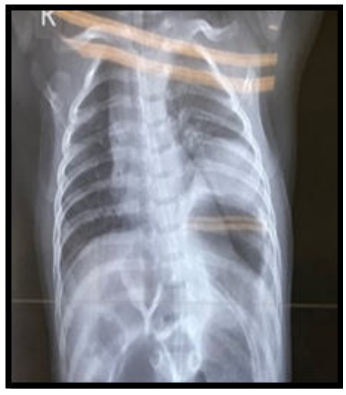


Figure 1 – Xray chest and abdomen showing left CDH

USG thorax and abdomen showed Pyloric canal length of 24 mm, Pyloric canal thickness of 4.3 mm and a Left CDH. 2D Echocardiogram was normal.

After medical stabilization, patient underwent exploratory laparotomy with Ramstedt's pyloromyotomy and left diaphragmatic hernia repair.

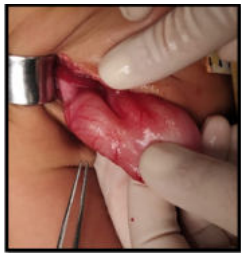


Figure 2 - IHPS

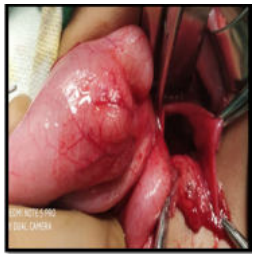


Figure 3 – Left CDH sac

Post operatively patient was maintained on mechanical ventilator on volume controlled mode. The child was extubated and breastfeeds were initiated on post operative day 2. Patient is doing well on follow up and has no symptoms of GERD on one year follow up.

DISCUSSION

Idiopathic Hypertrophic Pyloric Stenosis (IHPS) with Congenital Diaphragmatic Hernia (CDH) is a rare association, exact etiology of which is unknown. The association can be genetic such as Cornelia de Lange syndrome, though it is usually sporadic.

IHPS has been associated with anomalies (6 to 12.1%) [1,2] has a rare association with CDH – only 7 cases reported in world medical literature so far.

A report from the patient National Inpatient Sample (NIS) database and Kids Inpatient Database (KID) reported a 1.4% incidence of pyloric stenosis in CDH[3]. The first series had three patients of operated CDH which were diagnosed to have pyloric stenoses on readmission to the hospital for investigation of vomiting[4]. Of the next three cases reported, only one patient had a posterolateral diaphragmatic hernia and the other two cases were paraesophageal hernias[8]. The next 3 patients were described by Jason Robertson et al described the possibility of IHPS developing after CDH repair and the need for early intervention to prevent long term morbidity.

The diagnosis of IHPS is based on the triad of - projectile vomiting, visible gastric peristalsis and palpable pyloric tumor. With association with other congenital anomalies, the clinical features and physical findings may be atypical rendering the diagnosis difficult.[1]

IHPS has been found to be associated with certain malformations such as malrotation[6], chronic gastric volvulus[7,8], diaphragmatic eventration[7], genitourinary tract abnormalities and esophageal atresia[6,9].CDH is associated with GERD and foregut dysmotility in 45 to 90 % of infants likely to be related to abnormal hiatal anatomy at the GE junction, lack of Angle of His and herniation of the stomach into the chest.[10] Long-term outcomes about feeding in these patients depend on underlying gastrointestinal and neurologic function.

Although the exact etiology of association of IHPS with CDH is unknown, the hypothesis put forward for development of IHPS in a patient of CDH include -

1. Abnormal position of GE junction with stretching and/ or obstruction due to herniation of the stomach into the chest in a patient with CDH [1,4]
2. Subtle changes in gastric function[1]

Awareness of this combination and intraoperative recognition will avoid the problems of delayed diagnosis in the postoperative period with increased morbidity and mortality[4]. Early recognition of concomitant pyloric stenosis may enable patients to tolerate gastric and oral feeds sooner post operatively, possibly reducing complications such as oral aversion, but the long-term outcomes are still largely dependent on the presence or absence of underlying gastrointestinal dysmotility, GERD and/or neurologic function.[10]

CONCLUSION

Presentation of IHPS may be atypical when associated with CDH. Although the association between IHPS and other anomaly is rare, a clinician should have a high index of suspicion for the contribution of associated conditions considering that feeding difficulty is a common problem in patients of IHPS associated with CDH.

Abbreviations

1. IHPS - Idiopathic Hypertrophic Pyloric Stenosis
2. CDH - Congenital Diaphragmatic Hernia
3. GE junction – Gastroesophageal junction
4. GERD – Gastroesophageal Reflux Disease
5. USG - Ultrasonography

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