

Case Report

A Rare Case of Completely Herniated Intrathoracic Stomach Presenting as Massive Hematemesis and Shock due to Severe Esophagitis in a Four Year Old Girl

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Introduction

Diaphragmatic hiatal hernias are a rare finding in infants and children. Most of the reported cases are acquired resulting as a complication following Nissen's fundoplication for gastro esophageal reflux [1]. More common causes of upper GI bleeds in children under five years of age are esophagitis secondary to GERD, esophageal varices, gastropathy, gastroenteritis, NSAIDs, Mallory-Weiss tear and Duodenal erosions [2,3]. Completely herniated intrathoracic stomach through a diaphragmatic hernia with severe esophagitis and hemorrhage from ulceration as in our patient, although reported elsewhere is being reported for the first time from Pakistan [4].

Case Presentation

A four-year-old girl admitted through emergency presented with no prior admissions or illnesses. Chief complaint was acute onset of massive hematemesis. Patient vomited dark colored vomitus with sloughs in the vomitus in copious amounts. Immediately she was started on an infusion of omeprazole along with intra venous fluids, transamine injection and vitamin K. Labs were sent for Helicobacter pylori that came out to be negative. She was kept nil by mouth and a nasogastric tube was carefully inserted. Over the next few hours patient deteriorated further and went into shock. Blood pressure was not recordable, pulses were weak. There were no petechial spots, bruises, no visceromagalay, fever or bleeding from any other site. Infusion of dopamine and dobutamine were started and endoscopy was planned. A differential of peptic ulcer disease, esophagitis and

Abstract

Age and clinical presentation of diaphragmatic hiatal hernias (a rare finding in pediatric age group) depends upon the size and site of defect. Severe forms with herniation of bowel loops into the thorax are diagnosed at birth due to severe neonatal respiratory distress and absent breath sounds on auscultation. Upper gastrointestinal (GI) bleed with massive hematemesis in pediatrics usually is a presenting complaint in children with chronic liver disease. Other gastrointestinal diseases such as gastro esophageal reflux diseases (GERD) or gastritis can also present with massive hematemesis. We present here a rare case of a four-year-old girl with an unusual cause for massive hematemesis and hypovolemic shock. Further workup led to diagnosis of a completely herniated intra-thoracic, inverted stomach. Site of bleed was severe esophagitis with ulceration at the lower esophagus near gastric junction.

Keywords: Hematemesis; Diaphragmatic Hiatal Hernia; GERD; Esophagitis; Pediatric Hypovolemic Shock

sepsis were made. Fortunately, the patient didn't succumb to her grave condition and started to improve. FFPS, fresh blood transfusion were arranged and infused she was gradually weaned off dopamine. Nasogastric tube was removed and she was allowed orally. Endoscopy couldn't be done on the planned day as patient had taken her breakfast. On the third day of admission, she was sent for endoscopy and findings of endoscopy were large Para esophageal hiatal hernia. An area of extensive ulceration with whitish sloughs and contact bleed was seen at lower end of esophagus involving gastro esophageal junction. No abnormal mass or varices were seen. Biopsy taken from the duodenum showed scalloped mucosa (Figure 1-4). An HRCT of chest and abdomen was planned and results of the CT showed complete herniation of the stomach into the right hemi-thorax and the stomach was inverted in addition to intrathoracic herniation.

The report on the CAT scan showed a large defect in the diaphragm noted at esophageal hiatus measuring approximately 3cm. Stomach was pushed upward through the defect and was malrotated and inverted. Stomach was lying in the right hemi-thorax posteriorly. Fat density along the displaced stomach was suggestive of herniated omentum as well. No mediastinal shift was present. There was no loss of lung volume. Rest of the findings was normal and final diagnosis of a large congenital hiatal diaphragmatic hernia with herniated inverted and malrotated stomach was made. The cardiothoracic surgeon was consulted and operation was planned (Figure 5-8). This child is still awaiting surgery.

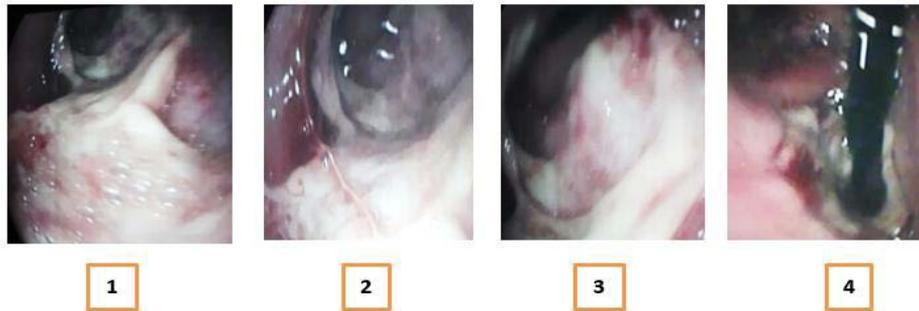


Figure 1-4: Endoscopy of upper GI tract showing, sloughing, ulceration and contact bleeding at lower esophageal end.

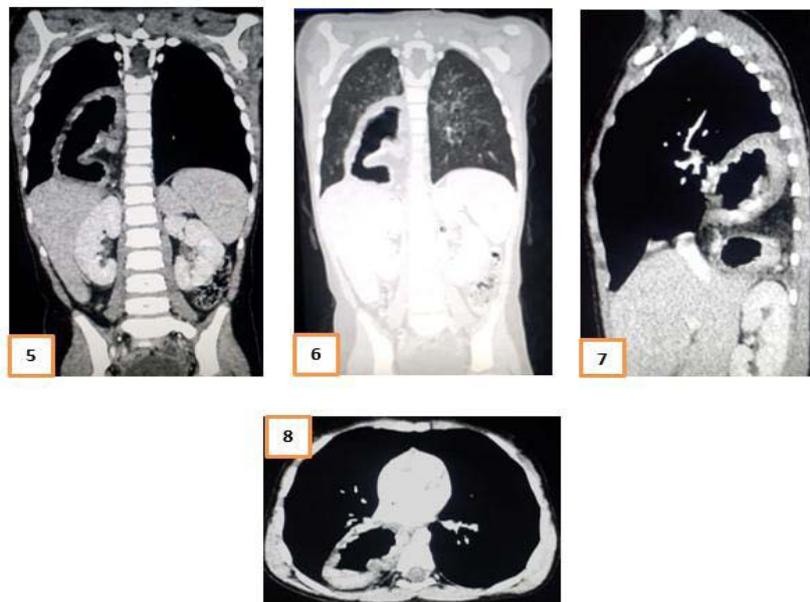


Figure 5-8: HRCT scan of chest showing completely intrathoracic herniated stomach in right hemi thorax. The patient was completely unaware of her condition.

Discussion

Diaphragmatic hernias in the pediatric age groups are divided into congenital and acquired. They constitute about 3.5–5% of all hiatal hernias having a female preponderance (male to female ratio being 1:4) [4-6]. Each year 4% to 10% of all infant deaths from congenital anomalies are caused by diaphragmatic hernias [7].

The esophageal hiatal hernias are divided into 4 types. The sliding hernia is the most commonly found type of esophageal hernia (represents 95% of all hiatal hernias) and is characterized by intrathoracic displacement of the gastro-esophageal junction (Type 1). Type 2 is the rolling or para esophageal hernia, it shows displacement of the stomach fundus and anterior wall. Type 3 is a combination of types 1 and 2; the gastro-esophageal junction is displaced into the chest. Total herniation of the stomach represents the end stage of hiatal herniation and other organs could also be herniated into the chest (Type 4). The incidence of type 4 hiatal hernia compromises 0.3% of all hiatal hernias [8].

Diaphragmatic hernias are found incidentally on radiography or endoscopy and maybe asymptomatic and go unnoticed. An

intrathoracic stomach is the end stage of a hiatal hernia, having a very low incidence and hence is a rare condition [9,10]. It can be asymptomatic as in our patient, though sometimes serious complications such as volvulus of stomach followed by incarceration, bleeding and perforation may occur. In such instances as it is life threatening and surgical intervention is essential [5,11].

A study including total 113 study children, 61 (54%) male conducted in Iran for finding common childhood causes for upper GI bleeding showed that the most important causes of bleeding in upper GI among all admitted patients were prolapse gastropathy (18.6%), esophagitis (15.9%) and esophageal varices, gastritis, and coagulopathy (7.1% for each). Whereas main causes of lower GI bleeding were polyp (32.5%), chronic nonspecific colitis (20.7%), and proctitis (18.2%) [2].

Another study quoted Hiatal hernia as an uncommon condition in children, presenting with symptoms that vary from none to life-threatening condition, thereby making its diagnosis challenging. Two different cases were reported about varying presentations of hiatal hernia, one a fifteen month old girl with partial gastric hiatal herniation brought to the emergency department with the complaints

of respiratory distress, endoscopy showed multiple strictures and ulcers in lower esophagus and stomach mucosa. Ultrasound of the abdomen revealed part of stomach herniating into the thoracic cavity. Barium swallow study confirmed the hiatus hernia (HH) type 1. Second case was a 7-year-old boy presenting with resistant iron deficiency anemia [12]. Familial causes, weakened ligaments and deficient elastin and short esophagus have been suggested as contributing causes to herniation of stomach through a hiatal defect [13].

Conclusions

Children with congenital hiatal hernia may go unnoticed after birth and even during infancy. High suspicion should be kept for hiatal hernias if the child presents with GERD like symptoms or with upper GI bleeds. Regarding diagnosis and subsequent management, guidelines laid by The Board of Governors of the Society of American Gastrointestinal and Endoscopic Surgeons (SAGES) are very helpful and we recommend that they should be followed [14].

Consent

Verbal and written informed consent was taken from the parents.

Authors Contribution

Dr. Saima Gillani: Consultant Pediatrician, Patient workup, diagnosis and management, literature search, conceptualization of study, write-up and proof reading.

Dr. Azmat Ali: Consultant radiologist, Patient workup (Chest HRCT with reporting, provision of films, diagnosis).

Dr. Samia Shaheen: Patient's notes and workup, patient management.

Dr. Waqas Ahmed: Patient workup, patient management.

Dr. Attia Iqbal: Junior consultant, Patient notes, diagnosis and workup.

Miss Fatima Farida: Compilation of notes.

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