Isolated Dextrogastria Simulating Congenital Diaphragmatic Hernia: A Diagnostic Dilemma

Man Mohan Harjai, Inna Kedle Indrajit and Monal Kansra

We report eventration of the right hemidiaphragm with isolated asymptomatic dextrogastria in a newborn. The diagnosis of this rare association could not be made clinically or by various imaging techniques. The final diagnosis could only be ascertained at laparotomy. The pathology was corrected by plication of the right hemidiaphragm, anterior fixation of the stomach, and Ladd’s procedure. [Asian J Surg 2010; 33(1):59–62]

Key Words: congenital diaphragmatic hernia, congenital hiatus hernia, dextrogastria, eventration of diaphragm, multiple congenital anomalies

Introduction

Congenital anomalies in the position or attachment of the proximal portion of the alimentary tube are exceedingly rare and usually occur as a part of general transposition of the viscera. Isolated dextrogastria is the rarest of all visceral transpositions and usually coexists with eventration of the right hemidiaphragm. We report an isolated case of asymptomatic dextrogastria with eventration of the right hemidiaphragm, which resulted in a diagnostic dilemma between congenital diaphragmatic hernia and congenital hiatus hernia. Another peculiarity of this case was that neonate had associated multiple congenital anomalies.

Case report

A 1.4-kg preterm newborn infant was born at 34 weeks of gestation by spontaneous vaginal delivery with severe birth asphyxia. The baby was resuscitated and cried after 20 minutes. Examination revealed multiple congenital anomalies in the form of microcephaly, prominent occiput, micrognathia, large low-set ears, beak-shaped nose, ulnar deviation of hands with crossing of fingers, micropenis, and rocker bottom feet. He was managed in the neonatal intensive care for prematurity. Routine and complete X-ray of the infant revealed air fluid level in the right lower chest, which was suggestive of right-sided congenital diaphragmatic hernia (Figure 1).

The baby was transferred to our tertiary care centre for management of congenital diaphragmatic hernia. Chest roentgenogram with contrast revealed a right-sided stomach, or dextrogastria without any outlet obstruction. Ultrasonography was suggestive of an anatomically located right dome of the diaphragm with good movements and no diaphragmatic defect. Contrast-enhanced computed tomography of the abdomen and chest could not delineate the diaphragm properly but showed crossing over of the oesophagus at the T4–T5 level towards the right, with the stomach lying at the base of right lung (Figure 2). Magnetic resonance imaging revealed a right-sided stomach lying above the liver (Figure 3). The possibility of congenital
hiatus hernia or some rarer entity such as dextrogastria was considered. In view of the diagnostic dilemma, surgery was performed after optimal stabilization.

On exploratory laparotomy, the left hypochondrium was empty; the left lobe of the liver was hypoplastic; the first part of the duodenum was found emerging between the right and left lobes of the liver. There was complete transposition of the stomach to the right side; the stomach was lying above the liver along with the spleen and attached to it distal pancreas. Eventration of the right dome of the diaphragm was noted. There was associated malrotation of the midgut in the form of Ladd’s bands; the caecum lay in the left iliac fossa. Plication of the eventration, anterior fixation of the stomach and Ladd’s procedure was carried out. Postoperative X-rays showed anatomical relocation of the stomach with normal contours of the right hemidiaphragm (Figure 4). The patient succumbed in the postoperative period from associated multiple congenital anomalies.

Discussion

Total situs inversus is moderately uncommon and occurs 1 in 6,000–8,000 cases. Partial or isolated visceral transposition is also rare and most often involves the heart. Isolated situs inversus of the stomach, with otherwise normal positions of the thoracic and abdominal viscera, is an extremely rare anomaly that occurs in two distinct forms. In type 1 dextrogastria, the stomach lies completely behind the liver, but the chest appears normal (Figure 5); in type 2 dextrogastria, the stomach lies above the liver in association with eventration of the diaphragm (Figure 6).
Embryologically, failure of the foregut to rotate normally results in type 1 dextrogastria. If there is associated failure to complete descent of the foregut from the chest, dextrogastria with eventration (type 2) will occur. Generally, both types of dextrogastria produce no symptoms and are discovered accidentally. Eventration of type 2 dextrogastria causes abnormalities on routine chest radiography and can simulate abscess, right-sided hiatal hernia, or other pathology at the right base. Contrast studies can show redundancy of the lower oesophagus in type 2 dextrogastria, with a high position of the stomach, as seen in our case. In both types, the gastric fundus and body are located posteriorly and appear as a mirror-image reversal on frontal projection. The duodenal bulb loop, duodenojejunal junction, and small and large bowels are all in their usual locations. In both forms of dextrogastria, the ligament of Treitz and all other viscera are in their usual position, but in our case, the spleen with the attached tail of the pancreas were pulled into the right thorax, along with the stomach. As a result of its close relationship with the liver, the right-sided stomach might produce interesting and confusing changes in liver scans. The diagnostic dilemma prevailed in our case until laparotomy, despite extensive evaluation by various imaging modalities.

Most cases of dextrogastria in the literature have been reported in adulthood. Previously, only four cases of dextrogastria in infancy were reported in the English language literature. One of these cases was duodenal web in which dextrogastria was detected incidentally by upper gastrointestinal contrast study; in the other three cases, the symptoms were recurrent vomiting and developmental retardation. Underlying eventration of the hemidiaphragm is known to occur in about one-sixth of all children who present with gastric volvulus. There was no evidence of gastric volvulus or gastric inlet or outlet obstruction in our case. The stomach is usually relatively fixed at the oesophageal hiatus at the pylorus, and is prevented from abnormal rotation by its ligamentous attachment. Absence or attenuation of these anatomical anchors results in movement of the stomach within the wide sub-diaphragmatic space under the eventrated diaphragm; this creates the potential for gastric volvulus. The coexistence of associated rotational abnormalities of foregut and midgut with dysmorphic features was another peculiarity in our case. Similar coexistence with rotational anomalies has been reported only three times before in the English language literature. Plication of the diaphragm with anterior gastric fixation via gastropexy or gastrostomy and abdominal exploration for
associated gastrointestinal anomalies is the most appropriate approach. We performed plication of eventration with partial wrapping of the fundus and gastropexy. Recurrence of gastric volvulus has been known to occur even following gastropexy.1

References