Volvulus Complicating Dextrogastria in an Infant

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We report eventration of right hemi-diaphragm resulting in gastric volvulus of the right-sided stomach in an infant. The diagnosis of this rare association was made with contrast CT scan. Patient was initially managed with reduction of stomach, plication of right hemi-diaphragm, anterior gastropexy and Ladd’s procedure, but required re-laparotomy after two months for recurrent volvulus.

Keywords: Dextrogastria, Infant, Volvulus.

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heart sounds were normal. Liver was palpable three-finger breadth below the lower coastal margin. Rest of the abdominal examination was unremarkable.

The hematological and biochemical investigations including blood gas studies were normal. Chest roentgenogram showed elevated right hemidiaphragm with haziness and a gas shadow below it. Upper GI contrast study showed right-sided stomach with normally positioned duodenum. Contrast enhanced CT scan of abdomen showed right-sided stomach lying above the liver and below the markedly elevated right hemi-diaphragm.

Exploratory laparotomy performed through right sub-costal incision revealed the right-sided stomach going behind the normally located liver under the membranous, elevated right hemi-diaphragm; it was normally rotated about its sagittal plane. Malrotation of gut was evident because of the Ladd’s bands and the cecum positioned in the left lumbar region. Reduction of stomach in its normal position, plication of right hemidiaphragm, anterior gastropexy and Ladd’s procedure was done. Patient was discharged on the 8th post-operative-day.

Patient remained asymptomatic for next two months. He turned later with episodic postprandial non-bilious vomiting of two-day duration. The upper GI contrast study was repeated, which revealed gastric volvulus. The patient was re-explored; gastric volvulus was corrected, revision anterior gastropexy and Nissen’s fundoplication were performed. The postoperative period was uneventful.

The child had been asymptomatic on two and a half years follow up.

Discussion

Total situs inversus is moderately uncommon occurring in 1:6000 to 1:8000 cases. Partial or isolated visceral transposition is also rare and most often involves the heart. Isolated dextrogastria is rare of all visceral transpositions and can occur in two distinct forms(1), type-1 stomach may lie completely behind the liver, but the chest appears normal and type-2 stomach lie above the liver in association with eventration of diaphragm and may simulate abscess, hydropneumothorax, right-sided hiatus hernia or other pathology of right lung base. Only isolated cases of either type are reported. Type-1 is apparently even rarer than type-2 (incidence <1:100000).

Generally, the dextrogastria produces no symptoms and is first noted as incidental finding on chest roentgenogram, liver scans, during abdominal surgeries, on autopsies. Most of the cases in existing literature have been reported in adulthood(1-6). There had been only three cases of dextrogastria in English literature reported to present in infancy hitherto(7-9). One of the these cases was of duodenal web where dextrogastria was incidentally detected on upper GI contrast study(9); in other two cases, the symptoms were that of recurrent vomiting and developmental retardation. In other patient, recurrent chest infections were also a predominant feature.

An underlying eventration of hemidiaphragm is known to occur in about one-sixth of all the children presenting with gastric volvulus(10). The stomach usually is relatively fixed at the esophageal hiatus at pylorus and is prevented from abnormal rotation by its ligamentous attachment. Absence or attenuation of these anatomical anchors results in the mobility of stomach within the wide sub diaphragmatic space under the eventrated diaphragm and makes it potential for gastric volvulus. Mesentero-axial is the most common type of gastric volvulus observed in the pediatric age group; the same was the finding in our case.

The coexistence of rotational abnormalities of both foregut and midgut was another peculiarity in this case. Similar coexistence has been reported only twice before in the English literature(3,7).

Eventration of the diaphragm associated with gastric volvulus always warrants surgery. Plication of the diaphragm with anterior gastric fixation via gastropexy or gatrostomy and abdominal exploration for associated gastrointestinal anomalies is the most appropriate approach. Though the recurrences following surgical correction by gatrostomy fixation for volvulus are not recorded, the recurrences have been known to occur gastropexy. We have treated the recurrence by gastropexy with Nissen’s fundoplication as we keeping the possibility of GERD in mind.


Hemangiomas are the most common tumor of infancy, occurring in up to 10% of all infants(1). In majority of cases, they are not associated with other abnormalities. Occasionally, they may indicate the presence of systemic malformations. The association of cerebrovascular and facial arterial anomalies and the hemangiomas was first noticed by Pascual-Castroviejo in 1978(2). Later Reese, et al.(3) in 1993 described the association of facial hemangiomas with Dandy Walker and other posterior fossa malformations(3). The acronym PHACE was proposed by Frieden, et al. in 1996(4).

Case Report

A 20-day-old female neonate, delivered at term by LSCS and born out of non-consanguineous marriage presented reddish skin lesions involving the face and the scalp at 2 days of life, associated with progressive erosion of the upper lips since 5 days of life. She also had two episodes of tonic convulsion that subsided spontaneously. The baby was on breast feed since birth but was having feeding difficulties few days prior to admission. Ultrasound scan done at 30 weeks of gestation had revealed Dandy Walker cyst.

On examination the weight of the baby was 2.65 kg, length 52 cm and head circumference

PHACES Syndrome with Congenital Hypothyroidism

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The acronym PHACE syndrome stands for Posterior fossa malformations, Hemangiomas, Arterial anomalies, Coarctation of the aorta and other cardiac defects, Eye abnormalities, Sternal clefting and/or a supraumbilical raphe. We report a 20-day-old neonate who presented with a large facial hemangioma and Dandy-Walker cyst. This case is unusual because the cyst was detected antenatally and there was associated congenital hypothyroidism, which has been described very rarely with this condition.

Key words: Dandy Walker cyst, Hemangioma, PHACES.