Concurrent hypertrophic pyloric stenosis and gastroschisis: An unusual presentation of a rare association

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A B S T R A C T
Gastroschisis and hypertrophic pyloric stenosis rarely occur in the same patient, with few reports of this unusual association published to date. We describe a neonate with prenatally diagnosed gastroschisis treated with interval silo reduction and closure. Following definitive repair, the patient had a 30-day interval of difficulty feeding and occasional emesis that progressed to projectile vomiting. Upper GI series was consistent with hypertrophic pyloric stenosis. Upon surgical exploration, multiple gastric adhesions were found to cause a 180° organoaxial gastric volvulus with aberrant relocation of the pylorus to the left upper quadrant. Following adhesiolysis and reduction of the volvulus, a Ramstedt pyloromyotomy was performed on the hypertrophied pylorus. Postoperatively, the patient’s diet was advanced as tolerated, and he was discharged home on postoperative day 10.

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In traditional cases of hypertrophic pyloric stenosis, the olive is typically palpable in the right upper quadrant, consistent with the normal location of the pylorus. However, in rare cases, an aberrantly located pylorus can lead to a delay in diagnosis, and therefore in treatment.

1. Case report

A 1-day-old newborn male was born at 36 4/7-weeks gestational age via vaginal delivery with prenatally diagnoses gastroschisis. The pregnancy was otherwise notable for early cannabis use in the 1st trimester and preterm premature rupture of membranes. Apgar scores were 8 and 9, and initial vitals and laboratory studies were within normal limits. Physical exam demonstrated a 1.5 × 2 cm abdominal wall defect to the right of midline with herniation of the entire bowel, stomach, and bladder. The patient was taken emergently to the operating room, where a protective silo was constructed. All hernia contents were viable upon completion of the procedure. Gradual silo reductions were performed every 48 h over 6 days, ultimately resulting in complete reduction of the gastroschisis. The abdominal wall was closed on day of life (DOL) 7. Three days later, the patient was successfully extubated, tolerated high-flow nasal cannula with good respiratory function, and feeds were advanced as tolerated.

On DOL 20, the patient developed occasional nonbloody, non-bilious emesis. An upper GI series performed at that time failed to demonstrate the passage of contrast into the duodenum. However, the gastric outlet obstruction was judged functional rather than obstructive, likely related to slowed gastric motility from the recent gastroschisis repair. The plan at that time was to hold feeds temporarily until tolerated and to repeat upper GI series in two weeks, with the expectation for gastric motility to return in time. Feeds were gradually advanced as tolerated with the addition of metoclopramide to his medication regimen.

On DOL 47–49, the patient once again developed increased emesis. An abdominal X-ray demonstrated gastric distension, and abdominal ultrasound showed likely pyloric stenosis, for which radiology recommended upper GI series (Fig. 1). Repeat upper GI series demonstrated extrinsic compression of the gastric antrum and gastric distention consistent with hypertrophic pyloric stenosis (Fig. 2).

On DOL 52, the patient was taken back to the operating room for Ramstedt pyloromyotomy. Multiple gastric adhesions resulting in a loose nonincarcerating 180° organoaxial gastric volvulus with aberrant relocation of the pylorus to the left upper quadrant were found. Adhesiolysis resulted in reduction of the gastric volvulus and relocation of the hypertrophied pylorus to its usual position in
the right upper quadrant. A Ramstedt pyloromyotomy was then performed.

Postoperatively, the patient’s diet was advanced as tolerated, and he was discharged home on DOL 62.

2. Discussion

In traditional cases of hypertrophic pyloric stenosis, the olive is typically palpable in the right upper quadrant, consistent with the normal location of the pylorus [1]. However, in the case described above, an aberrantly located pylorus led to a delay in diagnosis, and therefore in treatment. While not conclusive, we postulate that the child’s recurrent emesis was mostly the result of the pyloric stenosis rather than the gastric volvulus given the presence of the child’s projectile emesis as well as the loose nonconsticting nature of the volvulus felt in the operating room. However, the gastric volvulus was likely the cause of the observed stomach distention on abdominal X-ray done on DOL 49. Despite the unclear cause of symptoms, this unusual presentation of hypertrophic pyloric stenosis with a left upper quadrant olive is a rare entity and warrants discussion.

Although the etiology of the adhesions and resultant organo-axial volvulus may have been related to silo creation and interval reduction, this particular complication has not yet been reported in the literature. An alternative explanation is that the volvulus was due to the large gastrochisis, which included both the bladder and stomach. Gastrochisis has been well associated with abnormal rotation of the bowel, but the frequency of volvulus appears to be rare [2]. Abdelhaveez et al. conducted a retrospective study of 128 patients with gastrochisis assessing the incidence of malrotation and volvulus. They reported malrotation in 29 patients (22.7%), but did not find volvulus in any patients [2]. As such, the presence of gastric volvulus with gastrochisis is a rare finding of this case.

Additionally, this case is distinctive for the occurrence of hypertrophic pyloric stenosis following gastrochisis in the newborn. Upon review of the literature, there are very few documented cases of these two pediatric conditions occurring together. The rarity of this association is demonstrated by Saxena et al. [3], who reported that of the 70 gastrochisis patients treated at their center over a 15-year period, only one had concurrent pyloric stenosis. Murthi and Nour [4] reported a similar case of antenatally diagnosed gastrochisis followed by pyloric stenosis diagnosed at day of life 23.

3. Conclusion

This unusual case of pyloric stenosis following reduction of gastrochisis demonstrates how this relatively common pediatric condition can present in an unusual manner (i.e., with an aberrantly located olive not readily apparent on physical examination). Therefore, given that postsurgical feeding difficulties may be due to rare obstructive etiologies such as pyloric stenosis, a high level of suspicion is warranted in any neonate with ongoing emesis to avoid delays in diagnosis and treatment.

Conflict of interest statement

The authors have no conflicts of interest to disclose.

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