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Case Report

Unusual associations of bilateral Morgagni hernia in a child with Down's syndrome: a case report

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ABSTRACT

Morgagni hernia is a rare form of congenital diaphragmatic hernia. It is associated with Down's syndrome as well as other congenital anomalies. Pediatric patients usually present with respiratory manifestations, the clinical presentation of Morgagni hernia is non-specific. Therefore, Morgagni hernia is mainly diagnosed by radiological imaging. The rarity of this type of hernia along with the vague clinical presentation can lead to missed diagnosis.

Keywords: Morgagni hernia, Down's syndrome, Diaphragmatic hernia

INTRODUCTION

Morgagni's hernia (MH) is characterized by retrosternal herniation of the abdominal contents through a diaphragmatic defect in a sternocostal triangle. It presents 3-5% of all diaphragmatic hernias.[1]

MH is a rare diaphragmatic hernia with nonspecific symptoms and variable presentation. The hernia can be discovered incidentally, or it can present with nonspecific gastrointestinal or respiratory symptoms and, in severe cases, with respiratory distress requiring support.[2,3]

Right-sided anterior diaphragmatic defect is much more common than the left-sided and bilateral types. It is associated with different congenital anomalies. Down's Syndrome and congenital cardiac diseases are the most common associated pathology.[3,5-7] Whereas other malformations such as intestinal rotation, omphalocele and chest wall deformities are also found along with MH.[4]

Colon is commonly herniated in the defect but the small intestine, stomach and the liver have also been reported.[16]

We reported a rare and unique case of a bilateral Morgagni hernia associated with malrotation and duodenal web in a child with Down's syndrome. To our knowledge, the pattern of anomalies present in our patient has not been previously described.

CASE REPORT

A 1-year 8-month-old boy, known case of down syndrome, came to outpatient department for ritual circumcision. He had mild chest infection for which chest X-ray was done. Incidentally diagrammatic hernia was discovered and patient was admitted for further work up (Figure 1a).

CT scan chest and abdomen was done that showed a large 4 cm anterior right hemidiaphragm defect with omental fat and part of large bowel herniated through it into the right hemithorax, displacing heart and mediastinal structures to the left side, causing mass-effect on the

pyloric canal and proximal duodenum. Focal 1×1 cm hyperdense component within the dilated/distended second part of the duodenum.

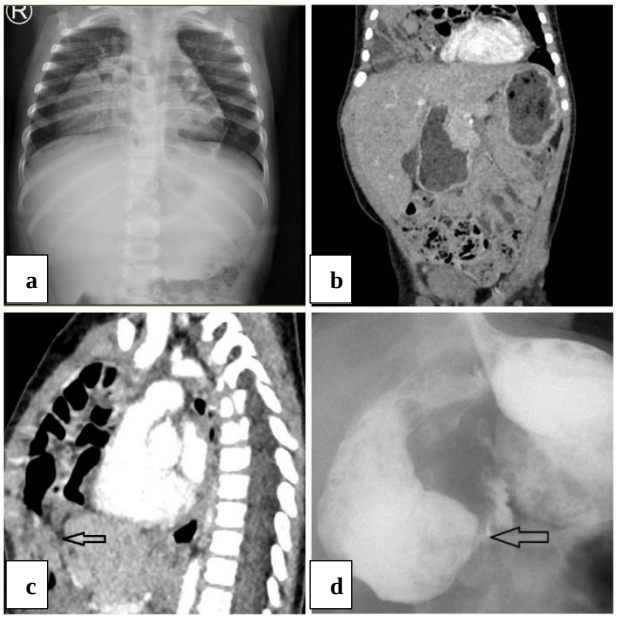

Figure 1: (a) Chest X-ray, diaphragmatic hernia; (b) CT scan showing right hemidiaphragm defect with bowel herniated through it into the right hemithorax, displacing heart and mediastinal structures to the left side, causing mass-effect on the pyloric canal and proximal duodenum; (c) component within the dilated second part of the duodenum; (d) showing duodenal obstruction suggestive of web.

Also focal narrowing at the second part of duodenum was seen with the proximal distention (Figure 1 b and c).

Upper GI contrast study was done that demonstrated distended stomach, whereas, contrast passed freely into the C-loop of duodenum, transient passage of contrast to second part of the duodenum through small opening with small crescent filing defect suggestive of duodenal web, The contrast reached into the jejunum with normal calibre (Figure 1d).

Patient underwent surgery electively. Per operatively we found two wide anterior defects in the right and left hemidiaphragms with bridge in between. Only transverse colon was seen bulging in the right hemithorax within hernial sac, while on the left side only hernial sac was found with no content (Figure 2a).

Reduction of transverse colon to abdominal cavity, starting with the right side along with excision of both hernial sac. Both defects closed with silk O interrupted sutures (Figure 2b).

Stomach, pylorus, 1st and 2nd part of the duodenum were found to be hugely dilated with Ladd's band compressing 2nd part of the duodenum. DJ was in normal position and no narrowing of mesentery seen. After excision of Ladd's band and straightening of duodenum, still there was persistent distension of stomach and proximal duodenum and contents could not be emptied distally. On the base of this finding, longitudinal duodenotomy and excision of fenestrated web was done. The duodenotomy was closed with vicryl 4/0. Rest of Ladd's procedure was completed as usual. Finally, circumcision was done.

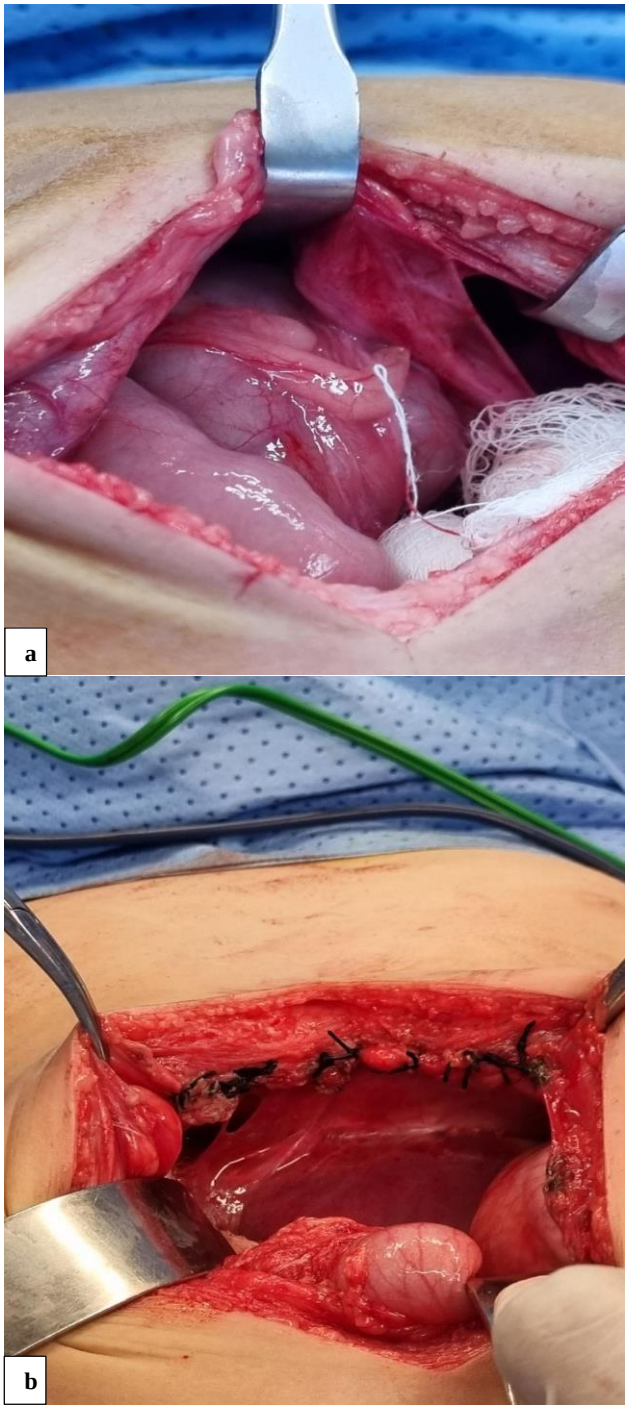

Figure 2: (a) Bilateral anterior defect in diaphragm separated by bridge in between; (b) surgical repair of defect.

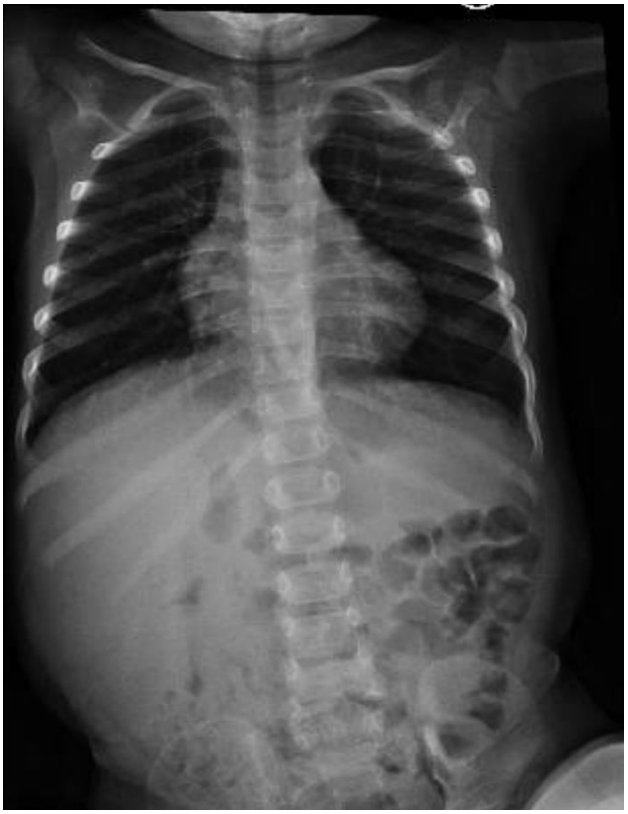

Figure 3: Postoperative chest X-ray.

The patient tolerated the procedure well, and he was discharged from the hospital 12 days after surgery. He was last seen in outpatient department 6 month post surgery and was thriving well (Figure 3).

DISCUSSION

The MH is a rare anomaly that represent about 5% of all congenital diaphragmatic hernias. They occur when abdominal content herniates through triangular parasternal gaps.[2]

In the literature, there are variabilities in the incidence of the side of hernia. Lamas-Pinheiro et al and Escarcega et al found bilateral hernias in the majority of their series (57% and 77% respectively).[4] Whereas Al Salem et al reported 8% patients had bilateral and Golden et al reported only 7.7%.[5] Due to the rareness of the condition, as well as the nonspecific presentation, the diagnosis is usually delayed. Al Salem et al reported frequent chest infections in most of the patients 81% and 3.8% were managed for GERD, whereas, Golden at al found almost a similar frequency of symptomatic and asymptomatic patients.[3,4] Among the symptomatic the most common presentation was respiratory tract infection. The most commonly herniated structure was the colon in the literature, however omentum and small intestines have also been found with high incidences.[3-6]

Down syndrome and congenital cardiac diseases are the most common associated pathologies.[3-7]

The connection between Down syndrome and MH is interesting. Honoré et al reported a highly significant association between the occurrence of MH and DS.[13] Pokorny et al also reported that three out of five infants with MH had DS. Al Salem et al found 28.3% of patient with Down syndrome.[3]

The association of CDH with malrotation is of 42-80% due to abnormal rotation of the embryological midgut secondary to the herniation.[8,9] The association between congenital diaphragmatic hernia with duodenal atresia is rare with only two previous cases reported.[14-16] As far as we know, this is the first reported case of a Down syndrome patient with bilateral MH with duodenal fenestrated web (windsock deformity) with malrotation.

CONCLUSION

We present a child of Down syndrome with a bilateral MH, Duodenal windsock deformity and malrotation. This case is unique in that it demonstrates a rare presentation of a rare pathology. A literature search for MH reveals several case reports throughout the years with varying patient ages and presentations. To our knowledge, this is the first report of a MH, malrotation and duodenal fenestrated web in a Down syndrome patient.

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Ethical approval: Not required

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