

A Case of Primary Acquired Gastric Outlet Obstruction in Late Infancy and Childhood

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Abstract

Presentation

We report a case of a 20-month-old who presented with a 3-week history of non-bilious vomiting and metabolic alkalosis.

Diagnosis

Serology demonstrated a hypokalaemic, hypochloraemic metabolic alkalosis. Abdominal ultrasonography and barium studies confirmed the diagnosis of gastric outlet obstruction.

Treatment

Oesophago-gastro-duodenoscopy identified a hypertrophied closed pylorus with evidence of pyloric muscular hypertrophy. A laparoscopic pyloroplasty was performed.

Discussion/Conclusion

Hypertrophic pyloric stenosis is the most common cause of gastric outlet obstruction in children. It typically occurs between 2 and 12 weeks of age and is very rarely found in older children. Primary acquired gastric outlet obstruction in late infancy and childhood is a rare cause of gastric outlet obstruction. Few cases have been reported in the literature outside of northern India. We present this case to highlight the need to consider this diagnosis in the older child.

Introduction

Gastric outlet obstruction (GOO) is defined as a structural abnormality at the level of the pylorus, which impedes gastric emptying and produces characteristic presentation of recurrent, projectile, non-bilious vomiting and hypokalaemic-hypochloraemic metabolic alkalosis.

Hypertrophic pyloric stenosis is the most common cause of GOO in children; with an incidence of between 1.5 and 3 per 1000 live births.¹ This condition most frequently occurs between the ages of 2 and 12 weeks and is very rarely found in children older than this.²⁻³ We report an unusual case of gastric outlet obstruction in an older child of 20 months.

Case Report

A 20-month-old presented with a 3-week history of persistent, projectile, non-bilious vomiting. There was associated early satiety and abdominal bloating with no lower gastrointestinal symptoms.

On examination, the patient appeared well and interactive.

- Observations were within the normal ranges.
- Mild dehydration present⁴ - reduced skin turgor and dry mucous membranes.
- Cardiovascular, respiratory, abdominal and neurological exams were unremarkable.

Serology demonstrated a hypokalaemic, hypochloraemic metabolic alkalosis. Abdominal ultrasonography identified that 'no gastric contents were seen to pass the pylorus.' Fluoroscopic studies showed that barium did not flow distal to the pylorus. Concentric narrowing at the pyloroduodenal junction was noted.

Differential diagnoses included hypertrophic pyloric stenosis, an antral or pyloric web, duplication cyst, annular pancreas, neoplasm, volvulus, Crohn's disease, and chemical injury.

Dehydration and hypochloraemic alkalosis were corrected and a nasogastric tube was placed. Oesophago-gastro-duodenoscopy showed a hypertrophied closed pylorus with evidence of pyloric muscular hypertrophy. The patient underwent laparoscopy and a pyloroplasty was performed. The patient made an uneventful post-operative recovery.

On last review at 20 months post operatively, there had been no complications and optimal weight gain was observed.

Discussion

Gastric outlet obstruction (GOO) in children between the ages of 3 months and 17 years, without a clear underlying cause, is very rare.⁵ In 1997, primary acquired gastric outlet obstruction in late infancy and childhood was named 'Jodhpur disease', after the city in which it was first reported. Forty-two cases have been previously reported in the literature, the

majority from northern India. It predominantly occurs in males with an incidence of 1 in 100,000 live births.⁶ This may postulate that genetic, ethnic, and dietary factors may contribute to its aetiology. The estimated mean age of presentation is 2.9 years.⁷

The aetiology and pathophysiology of this entity remain poorly understood. One of the proposed aetiologies is a deficiency of nitric oxide synthase enzyme within the pylorus. Nitric oxide synthase is the primary inhibitory neurotransmitter that acts on the pylorus and therefore, a deficiency could manifest as a pseudo-obstruction.⁷ Another proposed cause is an abnormality within the interstitial cells of Cajal. These act as communicators between the autonomic nervous system and smooth muscle cells within the gastrointestinal tract. An abnormality could therefore contribute to uncoordinated muscular contraction and delayed gastric emptying.

Some non-surgical treatment options include pneumatic balloon dilatation and botulinum toxin injection. Surgical management remains the gold-standard treatment. The standard technique is the Heineke-Mikulicz pyloroplasty. It is most appropriate for the treatment of smaller strictures.⁸ It involves making a 4-5 cm-long full-thickness longitudinal incision, which is closed in a transversely.⁸ This procedure rarely yields complications; however, complications include leakage at suture site, ongoing vomiting, and gastric dumping syndrome.

In conclusion, this is a rare condition which presents as gastric outlet obstruction in an older child. The management of this condition is primarily surgical, and it yields a very high success rate.

Declarations of Conflicts of Interest:

None declared.

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