Congenital microgastria with limb defect combined with megaduodenum: case report and review of literature

Peter Vasas · Satvinder S. Mudan · Charles A. Akle

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Abstract Congenital microgastria is a rare developmental disorder with only 59 previously reported cases. Most of the patients undergoing reconstructive surgery are in early infancy, providing gastric augmentation to achieve optimal thrive. The Hunt-Lawrence pouch is the standard surgical treatment in infancy, but late surgery in early adulthood requires different approach. This case demonstrates that duodenojejunostomy is safe and effective in the treatment of congenital, functional duodenal obstruction in adulthood.

Keywords Microgastria · Limb defect · Developmental abnormality · Hunt-Lawrence pouch · Duodeno-jejunal bypass

Introduction

Congenital microgastria (CM) is a rare developmental disorder, with only fifty nine previously reported cases. It is an idiopathic disorder which was first described in 1842. [1] Microgastria presents with vomiting, recurrent aspiration pneumonia, and failure to thrive. All previously reported cases underwent operative intervention in early childhood, providing gastric augmentation to achieve adequate nutritional intake. We report a case of an eighteen year old boy, who successfully underwent reconstructive surgery in adolescence.

Clinical report

The patient was born in the Middle East to distantly related parents. No other congenital or inherited diseases were identifiable in the family. The patient's birth weight was 1.6 kg following an uncomplicated pregnancy. At birth he had left arm hypoplasia with an absent left hallux. As an infant he failed to thrive, suffered recurrent respiratory infections, and frequent vomiting. Aged two he underwent a barium swallow and esophago-gastroscopy, which demonstrated a patent gastric tube with marked dilatation of the second part of the duodenum. He underwent duodenal 'tapering' and excision of the redundant duodenal wall. Symptoms were temporarily alleviated.

He represented at the aged twelve. Repeat endoscopy showed a peptic stricture of the lower third of the esophagus, small calibre stomach and a dilated duodenal bulb. Endoscopic esophageal dilatation resulted in resolution of dysphagia and vomiting. Over a period of three months four further esophageal dilatations were performed. Barrett's epithelium was excluded by histopathology. At the age of seventeen the patient was referred to our unit with recurrence of symptoms. Physical examination revealed short stature, low BMI (16.2), absence of secondary sexual characteristics, left sided limb hypoplasia, congenital absence of the thumb (Figs. 1 and 2) and left sided conductive deafness.

Esophago-gastroduodenoscopy revealed a peptic esophageal stricture between 23 and 30 cm. This was endoscopically dilated to 15 mm. The stomach was small and the third part of the duodenum was grossly dilated. Esophageal manometry showed poor peristalsis. Ambulatory pH monitoring showed severe nocturnal reflux and duodeno-gastro-esophageal reflux. Barium contrast study confirmed the abnormally small stomach, a large hiatus hernia, and an extremely dilated

P. Vasas (✉) · S. S. Mudan · C. A. Akle
The London Clinic, University College London,
London, UK
e-mail: vasasdr@gmail.com

Fig. 1 Congenital absence of left thumb

second and third part of duodenum with an abnormally positioned duodenojejunal flexure. The saccular megaduodenum was extending into the pelvis like a sump (Fig. 3).

Genetic investigations revealed normal chromosomes, telomeres, comparative genomic hybridisation, chromosome breakage or platelets (thrombocytopenia with absent radius syndrome).

In view of the previously failed procedure and the recurrence of symptoms, definitive surgery was planned. Triangular tapering duodenoplasty was considered inappropriate because there was no mechanical obstruction [2]. Duodeno-jejunosomy was felt to be the best option considering the functional obstruction. This would also allow ablation of the saccular sump.

During the surgery there was no stricture of 4th part of the duodenum, fibrous band or superior mesenteric artery syndrome. Instead of mobilizing the DJ flexure around the Treitz-ligament, a side-to-side duodeno-jejunal bypass

Fig. 2 X-ray of left hand

Fig. 3 Barium contrast study

(SDJ) was performed along the length of the third part of the duodenum [Fig. 4] [3]. Histology results excluded smooth muscle myopathy and nerve fibre hyperplasia.

His symptoms improved significantly, and a postoperative barium study showed rapid transit of food from stomach into the jejunum. By 4 weeks his albumin and haemoglobin significantly improved (Albumin 25 g/L to 31 g/L, Hb: 10.3 g/dl to 12.7 g/dl). He gained three kg. His esophageal stricture was treated with esophageal dilatation and proton pump inhibitors. Follow-up at 12 and 18 months showed excellent clinical and functional result with significantly improved BMI (16.2 to 18.6).

Fig. 4 Duodeno-jejunostomy

Discussion

Congenital microgastria is a rare disorder, with only 59 previously reported cases. The clinical presentation of microgastria is variable, depending upon the stage at which gastric development has been arrested. The stomach originates as a dilatation of the foregut during the 4th week of embryological development. In the 5th week the stomach achieves its normal contour through differentiated growth. The stomach and the spleen are both derived from the embryonic dorsal mesogastrium; hence association with asplenia is common. The association of limb, cardiac, tracheo-esophageal, vertebral and renal anomalies with congenital microgastria has been attributed to impairment of early mesodermal development, most probably due to gene dysfunction. These associations often reported as VATER/VACTERL syndrome (vertebral, anal, cardiac, tracheo-esophageal, renal and limb defect), [4] although many cases in the literature do not fit the diagnostic criteria for VATER/VACTERL. [5] CM typically presents with postprandial vomiting, failure to thrive, growth retardation, malnutrition, and recurrent aspiration pneumonia in early childhood. [1] The diagnosis is made with contrast studies of the upper gastrointestinal tract, which usually show a narrow, fine tube in the midline, frequently associated with incompetence of the lower esophageal sphincter and consequently megaesophagus. The esophagus dilates to compensate for the absence of the stomach, hence the esophagus returns to normal size after surgery. Because of severe acid reflux changes in the esophageal mucosa are often visible, and the exclusion of Barrett epithelium is of paramount importance. Several biopsy studies in microgastria have shown normal gastric mucosa; however the total cell mass of the stomach is severely reduced, leading to reduced production of acid and intrinsic factor [1]. All previously reported cases have been diagnosed and have undergone operative treatment in early childhood. [1, 6–8] This case describes successful operative management of a patient in adolescence. Before the introduction of gastric augmentation, conservative treatment consisted of frequent small feeds aiming to achieve adequate nutritional intake. [2] Conservative treatment has not been successful, failure to thrive, underdevelopment of secondary sexual characteristics and cognitive impairment were common.

A Roux-en-Y loop created from a jejunal loop may not provide adequate absorption because of dumping syndrome. Joining afferent and efferent jejunal loops together creates a Hunt-Lawrence pouch (HLP), which improves outcome by augmenting the neo-stomach content and reduce dumping into the small bowel. The HLP has

excellent results relieving symptoms of reflux and facilitates nutritional management [4, 8]. All case reports and retrospective studies carried out in this subject have dealt solely with infants. All authors performed HLP at the age of 11–15 months. Most of them reported optimal outcome with adequate weight gain and socio-intellectual development. Longest follow up was 18 years with good outcome [7]. In adulthood a large capacity stomach is not vital to maintain adequate nutritional status. In this case the preoperative studies showed functional obstruction of the fourth part of the duodenum, therefore a side-to-side duodenojejunostomy was chosen as the surgical treatment. The duodenojejunostomy safely ameliorated the functional duodenal obstruction. As the patient was 18 years old, it was felt that creating a HLP would not improve nutritional intake. The duodenojejunostomy provided optimal GI tract continuity, abolished the gastro-esophageal reflux and dysphagia, and enhanced nutritional absorption.

Summary

Duodenojejunostomy is safe and effective in the treatment of congenital functional duodenal obstruction in adulthood. Congenital microgastria presenting in adolescence may be treated with duodenojejunostomy in preference to HLP, although larger patient collective is needed before duodenojejunostomy becomes the standard treatment. There is no consensus on the age at which duodenojejunostomy has improved outcome over HLP.

References

1. Kroes EJ, Festen C (1998) Congenital microgastria: a case report and review of literature. *Pediatr Surg Int* 13(5–6):416–418
2. Spigland N, Yazbeck S (1990) Complications associated with surgical treatment of congenital intrinsic duodenal obstruction. *J Pediatr Surg* 25(11):1127–1130
3. Loire J, Gouillat C, Partensky C (2000) Megaduodenum in chronic intestinal pseudo-obstruction: management by duodenectomy-duodenoplasty. *Gastroenterol Clin Biol* 24(1):21–25
4. Ramos CT, Moss RL, Musemeche CA (1996) Microgastria as an isolated anomaly. *J Pediatr Surg* 31(10):1445–1447
5. Stewart C, Stewart M, Stewart F (2002) Microgastria-limb reduction anomaly with total amelia. *Clin Dysmorphol* 11(3):187–190
6. Hoehner JC, Kimura K, Soper RT (1994) Congenital microgastria. *J Pediatr Surg* 29(12):1591–1593
7. Jones VS, Cohen RC (2007) An eighteen year follow-up after surgery for congenital microgastria—case report and review of literature. *J Pediatr Surg* 42(11):1957–1960
8. Velasco AL, Holcomb GW III, Templeton JM Jr, Ziegler MM (1990) Management of congenital microgastria. *J Pediatr Surg* 25(2):192–197