CASE REPORT

An unlikely cause of severe malnutrition in a 3-year-old girl with previous gastroschisis

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SUMMARY

A 3-year-old girl with previous gastroschisis associated with jejunal and colonic atresia presented with severe oedema, abdominal pain and diarrhoea. Clinically she was malnourished. Serum albumin and concentrations of micronutrients were low. A barium meal examination showed jejunal dilation. A stricture was suspected and the patient was taken to theatre where an 8 cm length of bowel was resected including a jejunal stricture at the point of previous atresia repair. Inside the proximal dilated jejunum was a large trichobezoar (hairball), thought to be acting as a ball valve inside the bowel. This girl made a rapid recovery after surgery. Her nutritional state improved, symptoms resolved and serum biochemistry normalised. She remains well at follow-up with normal blood results, normal albumin and no diarrhoea.

BACKGROUND

Although malnutrition is relatively common worldwide the presentation of a severely malnourished child in the developed world is less common, and usually secondary to a significant disease process.

We identify an unusual cause of malnutrition in a child with gastroschisis and discuss the aetiology and pathophysiology of this presentation.

CASE PRESENTATION

A 3-year-old girl who had previously undergone surgery at birth to correct gastroschisis (see [figure 1](#)) and to repair two atresias (jejunal and colonic) presented to the paediatric gastroenterology department with a 6-month history of increasing oedema, markedly increased appetite and general decline in energy levels with significant lethargy. Her weight had been increasing along the 50–75th centile for the past 2 years; her height was on the 5th centile.

On further questioning there was a history of intermittent abdominal distension during the preceding year, accompanied by abdominal pain and frequent loose stools. Although there was a history of frequent vomiting during the first 2 years of life her mother described infrequent but extremely large vomits over last few months, at times containing food that had been eaten 2–3 days previously.

On examination she was pale but not clinically jaundiced. She had oedematous pale skin and thin wispy hair. There was marked oedema of the face, distension and oedema of the legs and arms bilaterally. Apart from a baseline tachycardia cardiovascular and respiratory examinations were normal. On abdominal examination there was distension

due to ascites but no masses were demonstrated (see [figure 2](#)).

It was noted that she was often miserable and lethargic but frequently requested food, which she would eat extremely quickly.

INVESTIGATIONS

Bloods at presentation showed (normal range in brackets)

- ▶ Haemoglobin: 81 g/dL (110–140)
- ▶ White cell count: $6.3 \times 10^9/L$ (5–15)
- ▶ Mean cell volume: 69.1 fL (75–87)
- ▶ C reactive protein: <1 (normal)
- ▶ Alanine transaminase: 19 iu/L (7–35)
- ▶ Bilirubin: 1 $\mu\text{mol/L}$ (0–20)
- ▶ Albumin: 13 g/L (30–50)
- ▶ Potassium: 3.9 (3.5–5)
- ▶ Sodium: 137 mmol/L (133–146)
- ▶ Urea: 3.8 mmol/L (2.5–6.5)
- ▶ Creatinine: 25 $\mu\text{mol/L}$ (20–70)

Vitamins, minerals and trace elements

- ▶ Calcium (adjusted): 2.15 mmol/L (2.2–2.6)
- ▶ Serum iron: <2 (Low)
- ▶ Ferritin: 2 $\mu\text{g/L}$ (10–170)
- ▶ Transferrin: 1.64 g/L (1.9–2.8)
- ▶ Phosphate: 1.13 mmol/L (0.9–1.8)
- ▶ Folate: 10.9 (3.2–25.9)
- ▶ Vitamin B₁₂: 268 ng/L (normal)
- ▶ Selenium: 0.64 (0.5–1.3)
- ▶ Zinc: 5.4 $\mu\text{mol/L}$ (11–24)
- ▶ Copper: 5.3 (low)
- ▶ 25-OH-vitamin D: 12 nmol/L (<30 nmol/L is consistent with deficiency)

Urine

- ▶ Protein/creatinine ratio was normal, alongside normal urine protein.

Stool

- ▶ Faecal elastase: 101 $\mu\text{g/g}$ (moderate pancreatic insufficiency, <100 is severe pancreatic insufficiency)
- ▶ Negative for *Clostridium difficile* toxin, negative for norovirus and rotavirus, no cysts, ova or amoebae seen on microscopy.

Radiology

- ▶ Barium meal: possible jejunal stricture (see [figure 3](#))

Endoscopy

- ▶ No enteropathy

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of secondary malnutrition in gastroschisis is wide including an underlying enteropathy, liver disease and pancreatic

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Figure 1 At birth with gastroschisis.

insufficiency. Primary malnutrition resulting from neglect must also be excluded.

TREATMENT

The patient was admitted for further assessment. She continued to vomit very large volumes. She was managed with intravenous fluids, human albumin solution (HAS) and parenteral nutrition (including appropriate nutrients and micronutrients). Her oral intake was kept minimal. The use of 4.2% HAS was indicated to treat acute circulatory compromise, manifested as tachycardia and poor peripheral perfusion.

All macronutrient requirements were given via parental nutrition (PN) due to ongoing vomiting and nutritional compromise refractory to enteral nutrition. PN was increased to full requirements gradually over several days to reduce the risk of refeeding syndrome. 100% of the micronutrient and vitamin requirements were replaced parenterally (including iron).

Figure 2 At presentation, demonstrating significant oedema and abdominal distension.

Figure 3 Barium meal demonstrating dilation of the bowel in the area of jejunal anastomosis.

With nutritional repletion the patient's clinical state improved considerably with resolution of her oedema although she continued to vomit if fed orally.

Barium radiology showed dilated jejunum proximal to an area suspicious of a stricture (see [figure 3](#)).

She had a laparotomy to investigate further. A jejunal stricture measuring 20 mm in length was identified at the point of surgical anastomosis for her jejunal atresia repair, with dilation of the proximal bowel to 40 mm. The stricture was resected and 80 mm of bowel removed with subsequent end-to-end anastomosis of healthy bowel (see [figure 4](#)).

On further examination of the dilated jejunum a large spherical trichobezoar was identified just proximal to the stricture.

We hypothesise that this was acting as a ball-valve and intermittently obstructing the gut.

Histology of the resected area (dilated and narrowed area) identified superficial ischaemic necrosis only with normal mucosa observed throughout. This necrosis was thought to be due to compression from the trichobezoar.

Figure 4 Resected jejunum with trichobezoar.

In the immediate postoperative period she was continued on parenteral nutrition. Over the subsequent few days food was reintroduced and tolerated well and she was discharged home.

She has made an excellent recovery with complete resolution of oedema over the course of 1–2 weeks (see figure 5). Her serum albumin is now normal. Weight initially fell after treatment as oedema resolved but has risen from 15.9 kg ($Z=0.09$) during acute illness to 17.1 kg ($Z=-0.06$) at 5-month follow-up. Height at 2 months prior to acute illness was 95 cm ($Z=-1.31$), at the point of discharge from hospital height was 95.5 cm ($Z=-1.68$) but at 5-month follow-up had risen to 99.2 cm ($Z=-1.13$). Z-scores are weight-for-age and height-for-age.

OUTCOME AND FOLLOW-UP

The patient remains well 6-month postprocedure with normal blood parameters including serum albumin. There have been no further episodes of distension/vomiting. She remains on long-term multivitamin/mineral supplementation.

DISCUSSION

This girl was thought to have a ball-valve-type obstruction to her jejunum due to the combination of a trichobezoar and luminal stricture. She presumably developed malabsorption due to bacterial overgrowth of the obstructed bowel segment. This in combination with incomplete passage of dietary intake through the small bowel is likely to have led to her malnutrition.

Despite a weight that was increasing along the 50–75th centile this patient was severely malnourished. The presence of significant oedema was masking tissue loss and falsely reassuring in this situation.

It is interesting that with further history taking that this child has a (long-term) habit of chewing and swallowing her hair.

The differential diagnosis for malnutrition in a patient with gastroschisis is broad and it was important to rule out an

underlying enteropathy alongside disorders not directly relating to the gut, such as pancreatic insufficiency and hepatic failure.^{1 2}

Case reports of long-term malabsorption in intestinal pseudo-obstruction are acknowledged in the literature,³ with many adult cases going on to become dependent on total parental nutrition.⁴

Small intestine bacterial overgrowth is recognised to cause malabsorption of nutrients at all age groups,⁵ older people are at specific risk due to comparatively immotile guts and it should be considered as a cause of malnutrition in these patients.⁶ Gastrointestinal dysmotility is well-recognised as a long-term problem postgastroschisis.

Reasons for the formation of a trichobezoar are numerous⁷ and they can have many adverse effects⁸; in this case it was likely exacerbated by the presence of a Pica secondary to severe iron deficiency seen with the stricture of the jejunum. Hair is a comparatively common cause of an intestinal bezoar; several case reports have detailed obstruction caused by other material blocking the intestine including vegetable material such as rhubarb.⁹

Cases of bacterial overgrowth in the small bowel secondary to pseudo-obstruction have been described in relation to causing malabsorption.¹⁰ A case report of a girl aged 13 suffering from severe nausea, vomiting and anorexia with a diagnosis of Rapunzel syndrome (where a large trichobezoar extended from the stomach to the duodenum without causing obstruction) demonstrates the potential variety and severity of clinical presentation of trichotillomania.¹¹

In summary we describe an unusual and interesting cause of malabsorption in a young girl with gastroschisis presenting with malnutrition.

Learning points

- ▶ This girl was seen in the outpatient department with a chronic history; her clinical appearance prompted her assessment and admission. Clinical examination findings such as recent onset of oedema require immediate further investigation.
- ▶ The diagnosis of secondary malnutrition is based on clinical observations and laboratory investigations including micronutrient status.
- ▶ Stable weight may mask tissue loss in the presence of oedema.
- ▶ The history of gastroschisis with stricturing is of relevance to the acute presentation.
- ▶ Presentation of small bowel strictures is hugely variable. Often they are difficult to diagnose clinically and present with atypical features.

Figure 5 At follow-up, resolution of oedema and a happy child.

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Competing interests None.

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