Case Report

A Late Diagnosis of a Complicated Meckel’s Diverticulum - The First Case of An Enterocolic Fistula Secondary to a Meckel’s Diverticulum in a Paediatric Patient And Review of the Literature

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Abstract
We report a case of acute-on-chronic bowel obstruction in a three year old girl due to an enterocolic fistula secondary to a Meckel’s diverticulum (MD). This is the first case presenting in a paediatric patient. We discuss the varied ways a MD can cause bowel obstruction, look at the presentation of MD as an enterocolic fistula and discuss the management of chronically dilated large bowel in a paediatric patient.

ABBREVIATIONS

MD: Meckel’s Diverticulum; PIES: Protein Induced Enteropathy Syndrome; SB: Small Bowel; LB: Large Bowel

INTRODUCTION

A Meckel’s diverticulum (MD) is a congenital anomaly of the gastrointestinal tract which occurs in 2% of the population [1,2]. It typically presents in those less than two years of age with painless bright red rectal bleeding, however it can present in unusual ways [2]. We describe an atypical presentation of a MD causing acute-on-chronic bowel obstruction and an enterocolic fistula in a three year old female, the first described case of its kind. We look at the ways MD can cause bowel obstruction and also discuss the management of the chronically dilated large bowel.

CASE PRESENTATION

A three year old female child presented acutely with a one week history of deranged electrolytes, abdominal distension and peripheral oedema, on a background of two years of intermittent abdominal pain, progressive abdominal distension associated with bourbourgym and explosive diarrhoea. She was born at term and did well for the first year of life. Due to her gastrointestinal symptoms since the age of one year, food allergy was considered and she was referred to the allergy clinic.

A diagnosis of fish protein induced enterocolitis syndrome (PIES) and cow’s milk protein intolerance was made, and parents were advised to follow a cow’s milk protein free diet and to avoid fish. Despite these measures her symptoms progressed and she presented acutely.

On admission she had a distended abdomen and pitting oedema to the mid-sacrum. She appeared septic, with a tachycardia (heart rate 170 beats per minute), low blood pressure (systolic BP 60-70 mmHg) and pyrexia (temperature 38.8 degrees centigrade). Her blood tests showed hyponatraemia (Na+ 124mmol/l), hypokalaemia (K+ 2.1mmol/l), hypocalcaemia (Ca2+ 1.99mmol/l),...
hypomagnesaemia (Mg 0.6±mmol/l) and hypophosphataemia (phosphate 0.5mmol/l). Her inflammatory markers were raised (CRP 114 mg/l) and her clotting was deranged (PT 25 seconds, APTT 57.4 seconds, INR 2.2). Antibiotics were commenced and she was given a potassium, phosphate infusion and albumin. She had lost 1kg in weight in one month.

Over the twenty four hours following her admission she complained of worsening abdominal pain and a surgical review of her abdomen was obtained. A contrast enema showed extremely dilated proximal large bowel lying upstream of a tight colonic stricture at the level of the proximal transverse colon (See (Figure 1)). The differential diagnoses considered were congenital colonic stricture, inflammatory bowel disease, a band obstruction, complex appendicitis and a late presentation of Hirschsprung’s disease. The decision was made to perform an emergency laparotomy as she became profoundly septic following the contrast enema.

The findings at laparotomy were of an enterocolic fistula between the terminal ileum and the proximal transverse colon, with chronic inflammatory changes causing a tight colonic stricture with a small pin-hole perforation at the site of the fistula (see (Figure 2)). The caecum, ascending colon and proximal transverse colon up to the level of the stricture were hugely dilated, measuring a maximum of 9 cm in diameter at the caecum (see (Figure 3)). The location of the fistula corresponded to the position in the distal ileum where a MD would normally be located, arousing suspicion of the fistula being secondary to a previous meckel’s diverticulitis. There was further small bowel obstruction due to loops of small bowel herniating around the fistula.

It was determined that she did not have a functional cause for the bowel obstruction; therefore the surgical decision was made to preserve the right colon. The fistula was taken down with resection of the colonic stricture and a primary end-to-end colo-colic anastomosis was performed (a 3:1 discrepancy was corrected). A proximal ileostomy was created at the site of the fistula. This was done with a view to defunction the dilated colon.

Histopathology showed the fistula to contain pyloric glands consistent with ectopic gastric mucosa seen in a MD, supporting our theory that this could be a fistula secondary to an inflamed MD.

She had a difficult post-operative course requiring admission to intensive care for management of sepsis. She was discharged home 27 days after the procedure, with the ileostomy. A contrast enema prior to closure suggested the colonic caliber had reduced and the ileostomy was reversed after four months.

At final follow up, one year post procedure she had no evidence of colonic dysmotility. She was passing formed stool twice daily and had gained adequate weight (on the 91st centile). She was also followed up in the allergy clinic. No further dietary restrictions except avoiding fish was recommended.

**Discussion and literature review**

A MD, the commonest congenital anomaly of the gastrointestinal tract [1], is a true diverticulum containing all layers of the bowel wall. It is a remnant of the vitello-intestinal duct that connects the fetal mid-gut to the yolk sac. This duct typically involutes by the 5th week of gestation but failure of involution leads to a persistent connection between the small bowel and umbilicus resulting in a range of pathologies [1-3].
A MD is present in 2% of the population, with an equal incidence between males and females [1]. It is usually symptomatic before the age of two years. It is found on the antimesenteric border, two feet from the ileocecal valve and is two inches long. Two types of ectopic mucosa (gastric or pancreatic) can be found at the base [1,2].

It presents in a variety of ways. It is the most common cause of painless rectal bleeding in infants less than two years of age [1,2]. This is due to mucosal ulceration caused by acid secreted from ectopic gastric mucosa at the base of the MD. It, and various remnants of the vitello-intestinal duct can cause bowel obstruction – this is the commonest mode of presentation in adults [1]. It can also present with inflammatory complications – mimicking appendicitis if the diverticulum becomes inflamed, and can lead to perforation or stricture formation.

An enterocolic fistula causing a bowel obstruction is an unusual presentation of a MD in a paediatric patient and this is the first case of an enterocolic fistula secondary to a MD in a child. Similar presentation of a large bowel obstruction in an adult would raise suspicion of an inflammatory, neoplastic or ischaemic process [4]. These are rare in the paediatric population therefore the clinical presentation in this child was perplexing. In children the common causes of large bowel obstruction can be considered as functional or mechanical.

Mechanical causes of large bowel obstruction in the paediatric age group include colonic or rectal atresia or stenosis, volvulus, foreign body ingestion, adhesions from previous surgery or an acute inflammatory process and a congenital band obstruction. Functional causes include Hirschsprung’s disease [5].

The management of large bowel obstruction in children depends on the underlying aetiology. Given the degree of colonic dilatation in our case, a similar feature is seen in neonatal patients presenting with colonic atresia - a congenital cause for colonic dilatation. In this condition a right hemicolecotomy and ileocolic anastomosis is a well described management option [6], due to the dilated colon proximal to the atresia being of very poor quality and the high risk of anastomotic leak if a primary anastomosis is performed. There is also a theoretic risk of the chronically dilated colon not being able to regain normal motility. Papers advocate an individualised approach to management depending on the location of the atresia. Primary anastomoses have been performed when there is minimal size discrepancy between proximal and distal bowel. A staged procedure with a defunctioning ileostomy to allow the proximal dilated bowel to return to a normal size and function prior to anastomosis has been shown to be an effective alternative avoiding the need for a hemicolecotomy and loss of the ileocecal valve [7,8]. However there is no published literature on the management of acquired dilated large bowel due to obstruction in children. In similar situations in adults a hemicolecotomy is usually performed due to the high risk of neoplasia. In our patient, due to neoplasia being an unlikely cause, the decision was made to preserve the chronically dilated right colon.

In a comparable case a 57 year old adult patient with a diverticulo-colic fistula underwent a right hemicolectomy. Histology confirmed the fistula contained gastric mucosa consistent with a MD. There was no explanation as to why a hemicolecotomy was performed [9].

Due to the rarity of this presentation of an enterocolic fistula in a paediatric patient, we were interested to perform a literature search looking for further rare presentations of MD. Using the search criteria of “MD”, “vitelline duct”, “fistula” (gastric, intestinal, rectal or enterocolic) we searched Embase and Medline. Sixteen articles of MD resulting in fistulae were identified. These were all case reports. Details of these cases including ours are shown in (Table 1).

The commonest pathology was ileo-vesicular and entero-cutaneous fistulae. Two cases were of a fistula with the rectum [10,11] and one with the appendix [12]. There were only two reports of an enterocolic fistula secondary to a MD [9,13] and both of these were in adult patients. Our case is the only described enterocolic fistula in a child. We wanted to look at the two enterocolic fistulae in further detail to look at the management of those cases in comparison to our patient. One of the adult cases is as discussed above, by Lo et al [9] whose operative finding bears resemblance to our patient. However the operative management was different and that patient underwent a hemicolecotomy and our patient did not.

In the other adult case of a diverticulo-colic fistula as reported by Reimer et al in a 54 year old male the fistula was connected to the sigmoid colon [13]. Exploratory laparotomy showed a fistula at the site of the MD going to the middle third of the sigmoid colon. They performed a resection of the diverticulum and adhesiolysis. In this case, histology did not confirm ectopic gastric mucosa, but the fistula was at the junction of the distal third and proximal two-thirds of the ileum, the expected site for a MD.

Although the most common mode of presentation of a MD in a child is profuse painless bright red rectal bleeding, bowel obstruction caused by a MD in the paediatric population is a common finding in the literature [2, 14]. A MD can cause bowel obstruction in a variety of ways [2]. There have been case reports of obstruction caused by internal herniation around a Meckel’s band [14]. Additionally the band can cause compression of the bowel leading to obstruction, thought to be more common in a shorter band [15]. It can act as a lead point for intussusception and cause obstruction in this way [16]. Inflammation of the diverticulum causing adhesional obstruction has also been reported [17-19]. The MD itself has been seen to wrap round the ileum leading to obstruction [20]. There have been reports of foreign bodies and faecoliths in the MD causing bowel obstruction [21-23]. This is the first report of a bowel obstruction secondary to an enterocolic fistula and stricture in a child.

We have performed a resection of the fistula and adjacent stricture and a primary colonic anastomosis, avoiding the need for a right hemicolecotomy. A defunctioning ileostomy allowed the dilated colon to gradually decompress prior to stoma closure. So far, with limited follow-up, the successful outcome of our patient suggests resection and anastomosis is a feasible option.
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<td>Our case</td>
<td>3 year female</td>
<td>Diverticulo-transverse</td>
<td>Resection of fistula and primary anastomosis with defunctioning ileostomy</td>
<td>7 months</td>
<td>Pyloric glands consistent with ectopic gastric mucosa</td>
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<tr>
<td>Lo et al, 2015 [9]</td>
<td>57 year male</td>
<td>Diverticulo-transverse</td>
<td>Right hemicolecotomy including distal small bowel incorporating the fistula tract</td>
<td>Discharged day 5 post-operatively. No long term follow up</td>
<td>Terminal ileal – transverse colon fistula, containing non-specialized gastric mucosa. Evidence of active inflammation</td>
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<td>Reimer et al, 1988 [13]</td>
<td>54 year male</td>
<td>MD-sigmoid colon</td>
<td>Uneventful recovery</td>
<td>MD acute and chronic inflammatory changes, no ectopic tissue</td>
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<td>Yang et al, 2012 [12]</td>
<td>18 year male</td>
<td>MD-appendix</td>
<td>Segmental resection of ileum and appendectomy</td>
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<td>Watt et al, 1985 [10]</td>
<td>19 year male</td>
<td>Giant MD containing an enterolith - rectum</td>
<td>Resection and small bowel anastomosis, closure of rectal fistula</td>
<td>Full recovery</td>
<td>No gastric or pancreatic mucosa</td>
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<td>Arıturk et al, 1991 [24]</td>
<td>4 year male</td>
<td>Ileo-vesicular</td>
<td>Wedge resection of 12cm Meckel’s and closure of fistula tract</td>
<td>19 months post operation – well, no urinary symptoms</td>
<td>Normal mucosal epithelium; no ectopic tissue</td>
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<td>Boussida et al, 2013 [25]</td>
<td>66 year female</td>
<td>Ileo-vesicular</td>
<td>Terminal ile-ectomy, diverticulectomy including resection of a portion of the bladder. Primary ileo-ileal anastomosis</td>
<td>Uneventful post-operative recovery, no long term follow up</td>
<td>Mild Meckel’s diverticulitis, no evidence of ectopic gastric or pancreatic tissue</td>
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<td>Dearden et al, 1983 [26]</td>
<td>81 year female</td>
<td>Ileo-vesicular</td>
<td>Meckel’s diverticulectomy, excision of fistula and repair of bladder</td>
<td>Uneventful post-operative recovery, no long term follow up</td>
<td>Inflamed MD; no heterotropic tissue identified.</td>
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<td>Grazioti et al, 2002 [27]</td>
<td>40 year male</td>
<td>Vesico-ileal (caused by ingested bone in MD)</td>
<td>Laparotomy; ileal resection, resection of section of bladder, removal of foreign body</td>
<td>No complications at 6 months follow up</td>
<td>No mention of histological analysis of fistula tract</td>
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<td>MacKenzie et al, 1989 [28]</td>
<td>30 year female</td>
<td>Vesico-ileal</td>
<td>Diverticulectomy; cuff of the bladder taken including the vesicular end of fistula.</td>
<td>Uneventful post-operative recovery, no long term follow up</td>
<td>Mild Meckel’s diverticulitis; sharp junction; no heterotropic tissue</td>
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<td>Petros et al, 1990 [29]</td>
<td>22 year male with Crohn’s disease</td>
<td>Vesico-ileal (plus other fistulae to bladder secondary to Crohn’s)</td>
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<td>Uneventful recovery</td>
<td>Mature gastric epithelium in the MD, not affected by Crohn’s disease</td>
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<td>Gilman et al, 2015 [30]</td>
<td>40 year male</td>
<td>Entero-cutaneous fistula via umbilical hernia</td>
<td>Diverticulectomy with excision of fistula tract and umbilicus</td>
<td>No follow up</td>
<td>Histopathology &quot;approved&quot; Meckel’s</td>
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<tr>
<td>Cranshaw et al, 2000 [31]</td>
<td>52 year male</td>
<td>Entero-cutaneous, 11 days post appendi-ectomy</td>
<td>TPN and Octreotide. Re-operated on 49 days, diverticulectomy.</td>
<td>Total inpatient stay 56 days. No long term follow up</td>
<td>Granulation tissue but no heterotropic tissue identified</td>
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<td>Hatipoglu et al, 2014 [32]</td>
<td>39 year male</td>
<td>Entero-cutaneous fistula of MD via a urachal cyst</td>
<td>Laparotomy and Meckel’s diverticulectomy with sparing of the adjacent bowel, resection of UC and umbilical entero-cutaneous fistula tract</td>
<td>Uneventful post-operative recovery, discharged 6 days post procedure</td>
<td>Focal ulcerated area in the small intestine with inflammation. Gastric ectopic tissue in MD, mucosal ulceration adjacent to the diverticulum. Submucosal oedema in small intestinal tissue</td>
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in the management of dilated bowel secondary to an obstruction caused by an enterocolic fistula, with the return of function of the chronically dilated large bowel.

CONCLUSION

We describe the first case of an enterocolic fistula secondary to a Meckel's diverticulum in a 3 year old female causing bowel obstruction. This is an extremely unusual presentation of a Meckel's diverticulum, previously only seen in the adult population. It reminds us that this rare entity can present atypically in a child and should be considered as a cause of bowel obstruction.

This case is useful to help guide the intra-operative management of chronically dilated large bowel in a child.

REFERENCES

fistulization of umbilical hernia. Arq Bras Cir Dig. 2015; 28: 152-153.