Quiz

Correct answer to the quiz. check your diagnosis

Brown bowel syndrome (intestinal lipofuscinosis) – a case report and review of the literature

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Brown bowel syndrome (BBS) is a very rare condition occurring in association with malabsorption syndromes. It is characterised by deposition of granular, brown pigment (lipofuscin or ceroid) in intestinal smooth muscle cells. Rarely BBS can be complicated by distention of any segment of the bowel. We present a case of BBS associated with massive dilation of first loop of the small intestine and moderate dilation of the sigmoid colon with functional intestinal disturbances requiring surgical intervention in an 11-year-old boy.

Key words: brown bowel syndrome, lipofuscin, malabsorption, intestinal dilation.

Introduction

Brown bowel syndrome (intestinal lipofuscinosis, intestinal ceroidosis, ceroid pigmentation) is brown discoloration of the intestinal wall caused by deposition of insoluble, light brown, granular pigment (lipofuscin) within the smooth muscle cells of the muscularis propria and muscularis mucosae of any bowel segment [1]. It occurs in patients with various malabsorption syndromes including celiac disease and Whipple's disease, causing deficiency of fat soluble vitamins, especially vitamin E [2]. Brown bowel syndrome is a rare entity which may be asymptomatic but also may have serious clinical implications. One of them is massive intestine dilation causing sub-obstructive symptoms necessitating partial intestinal resection [3]. Although BBS is very rare, it should be taken into consideration in cases with major intestinal dilatation in patients without mechanical obstruction suffering from a malabsorption syndrome.

Case report

An 11-year-old boy (born on time with signs of evisceration, operated on the first day of life) diagnosed with unspecific malabsorption syndrome, IgA deficiency and unspecific myopathy was admitted to the hospital. He had a history of recurrent diarrhoea and symptoms of functional intestinal disturbances suggestive of partial intestinal obstruction lasting for several months prior to admission. The above listed symptoms worsened during the last three weeks. The boy also complained about colicky abdominal pain.

Physical examination revealed an afebrile, cachectic boy with body mass as well as height below the 3rd percentile. His abdomen was distended but bowel sounds were easily heard. Ultrasound and computed tomography showed the first small intestinal loop distended up to 10 cm at a distance of 15 cm. The sigmoid colon was slightly extended as well. The dilated intestinal loop was palpable through the skin. Lapa-

rotomy revealed a dilated, darker than usual first jejunal loop. Peristaltic movements of the dilated loop were preserved, but they were described as severely disorganised. No perforation was seen. Resection of an 18 cm long segment of the small intestine was done. The material was sent for histological examination.

Microscopically, small intestinal wall structure was preserved and intestinal villi of normal length were seen (Fig. 1). At higher magnification light-brown, granular pigment was visible in longitudinal and circular muscle coats of muscularis propria and in muscularis mucosae of the intestinal wall (Fig. 2A-B). The same pigment was present in macrophages within sinusoids of the small mesenteric lymph node. Brown pigment stained strongly with periodic acid Schiff (PAS) (Fig. 3) and was resistant to diastase digestion. Stains for iron and melanin were negative. The diagnosis of BBS (intestinal lipofuscinosis) was made. The patient's postoperative course was uncomplicated.

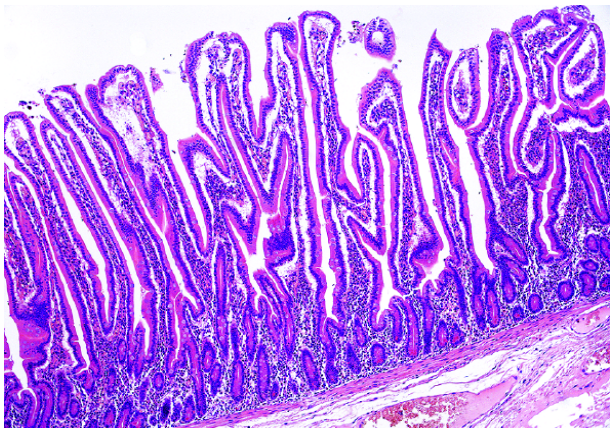

Fig. 1. Intestinal villi of normal length, HE, 10×

Discussion

When brown discoloration of any part of the gastrointestinal tract is macroscopically visible, one of the intestinal pigmentation conditions must be suspected. Intestinal pigmentation is seen in BBS, Whipple's disease and melanosis coli. In BBS granular pigment is accumulated mostly in smooth muscle cells of muscularis propria and muscularis mucosae. Rarely accumulation of pigment is also present in mesenteric lymph node macrophages. The most frequently affected part of the bowel is the small intestine. In Whipple's disease and melanosis coli brown pigment is present almost exclusively within macrophages in intestinal lamina propria. The pigment in BBS is lipofuscin [4, 5]. It stains strongly with PAS and is resistant to diastase digestion. It also stains with methylene blue, methenamine silver and Masson-Fontana [6, 7]. In some cases of BBS intestinal wall discoloration may be very discrete or even invisible on gross inspection and the diagnosis is made only on the grounds of histological findings in a bowel specimen or in biopsy material.

Brown bowel syndrome was first described by Wagner [8]. Later the association between BBS, malabsorption syndromes and vitamin E deficiency was found [9]. Foster proposed that BBS is a kind of smooth muscle mitochondrial myopathy resulting in lipofuscin accumulation within smooth muscle mitochondria, which occurred in patients with vitamin E deficiency [5]. Vitamin E acts as a mitochondrial antioxidant. Accumulation of lipofuscin (ceroid) usually occurs in smooth muscle of the small or large intestine and in lymph node macrophages. Rarely brown, granular pigment can be seen in smooth muscles of the oesophageal wall and in walls of small vessels [6, 10]. Vitamin E plays a role in protection and stabilisation of mitochondrial membranes and prevents membranes' damage by free radicals. Vitamin E deficiency accelerates peroxidation of unsaturated membrane lipids and leads to mitochondrial malfunction and degeneration with lipofuscin accumulation in intestinal smooth muscle cells. Damaged mitochondria are not able to provide a sufficient energy supply for smooth muscle, which may result in impaired function of smooth muscle and muscle myopathy with subsequent decrease in intestinal tone leading to massive dilatation

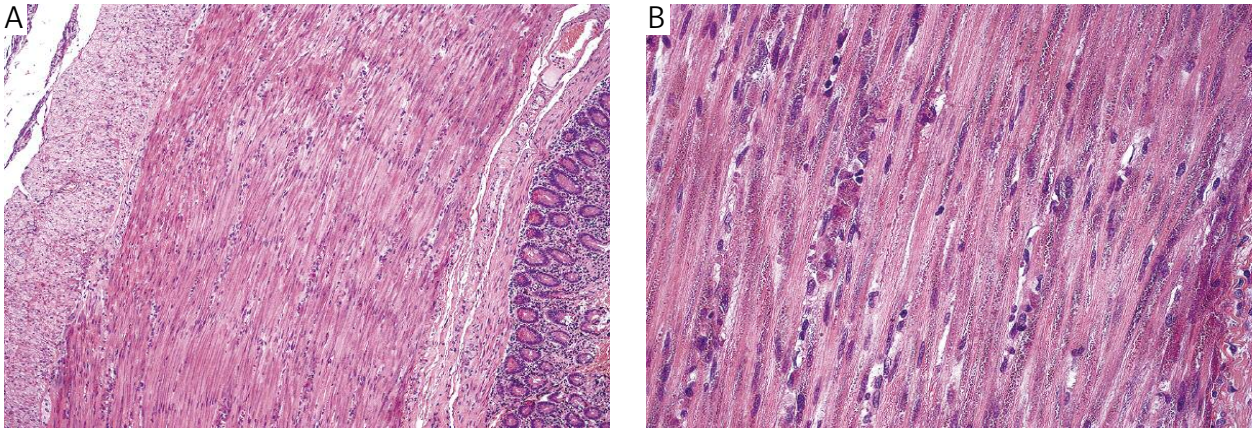

Fig. 2A-B. Brown, granular pigment in longitudinal and circular muscle coats of intestinal wall, HE, A: 20×, B: 40×

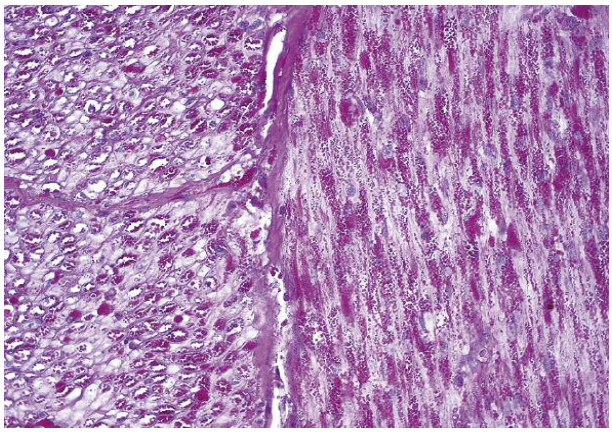

Fig. 3. Strong staining of pigment with periodic acid Schiff (PAS), 40×

of any part of the bowel. Therefore BBS may present clinically as pseudo-obstruction necessitating surgical intervention [2, 10-13].

Brown bowel syndrome is three times more common in males than in females (Table I). It is found in association with various malabsorption syndromes and nutritional disturbances, causing vitamin E deficiency, especially coeliac disease, chronic pancreatitis, liver cirrhosis, Crohn's disease, tropical sprue, intestinal lymphangiectasia, Whipple's disease and cystic fibrosis (Table I).

Brown bowel syndrome is not a primary disease, but rather a consequence of long-standing malabsorption of fat-soluble vitamins, especially vitamin E. Treatment with vitamin E supplementation in most cases will not

Table I. Clinical manifestations associated with brown bowel syndrome diagnosis

Ref. no.	Patient's		Clinical diagnosis on admission to hospital	Clinical manifestations
	age	sex		
2	58	M	coeliac disease	acute rectal bleeding, steatorrhoea, distal colon dilatation
2	68	F	coeliac disease	weight loss
2	68	F	coeliac disease	intestinal obstruction without mechanical cause
3	34	M	coeliac disease	watery diarrhoea, weight loss, dilatation of the colon
6	30	M	protein losing enteropathy	diarrhoea, steatorrhoea
6	53	M	coeliac disease	steatorrhoea
6	37	M	chronic pancreatitis (total pancreatectomy 15 years ago)	incidental finding
6	71	M	coeliac disease	steatorrhoea
7	11	F	jejunal atresia at birth (operated in neonatal period)	massive dilatation of small bowel
7	10	M	jejunal atresia at birth (operated in neonatal period)	copious vomiting (because of adhesive intestinal obstruction)
10	52	M	chronic pancreatic insufficiency	massive dilatation of small bowel chronic steatorrhoea
11	39	M	Whipple disease	chronic steatorrhoea, intestinal obstruction, megaoesophagus
12	53	M	stricture in sigmoid colon	malnutrition, electrolyte disturbances
12	63	F	jejunal diverticula	weight loss, abdominal pain
12	61	M	postradiation enteritis and adhesions	diarrhoea
12	78	M	chronic pancreatitis	incidental finding
14	67	F	Crohn disease complicated by intestinal obstruction	nausea, vomiting
15	58	F	jejunoileal bypass for weight reduction	incidental finding
16	31	M	alcohol abuse, chronic pancreatic insufficiency	ileal intussusception (vomiting, abdominal pain)
17	58	M	coeliac disease	rectal bleeding, abdominal pain
18	47	M	pulmonary tuberculosis	weight loss, abdominal pain, vomiting
19	–	–	coeliac disease	radiologic changes suggesting lymphoma
20	52	M	malabsorption syndrome	incidental finding
our case	11	M	malabsorption syndrome	massive dilatation of small bowel

reduce the amount of already deposited lipofuscin in muscle cells but it can prevent further progression of lipofuscin accumulation and subsequent complications.

The identification of lipofuscin accumulation and BBS diagnosis had two implications for the patient: a malabsorption syndrome should be searched and the vitamin E level should be checked.

Conclusions

Brown bowel syndrome is a rare condition which probably is a non-specific morphologic marker of other systemic diseases causing vitamin E deficiency. In children and young adults, especially in cases with heavy lipofuscin accumulation, it can be associated with massive intestinal dilation. BBS should be considered in patients with malabsorption syndromes without inflammatory bowel disease when massive intestinal dilation occurred.

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