Pseudoobstruction in Ceroidosis

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Diffuse deposition of ceroid pigment in the muscularis propria of the gastrointestinal tract in a patient with a long history of malabsorption of unknown origin is reported. The deposition of this waste pigment is not reversible and is related to prolonged depletion of vitamin E. Progressive dilatation and hypomotility of the entire gastrointestinal tract are demonstrated by radiographic studies and possibly related to infiltrate of ceroid pigment in the smooth muscle cell with resulting functional impairment. In the differential diagnosis of ceroidosis with other disease, scleroderma has the closest roentgenographic similarity. Pseudoobstruction of the small bowel which can develop must be treated conservatively to avoid unnecessary bowel resection.

Ceroidosis is diffuse accumulation of ceroid pigment in the muscularis propria of the gastrointestinal tract [1–3]. It is not a primary disease but a consequence of long-standing malabsorption. The main clinical manifestations are chronic steatorrhea, malabsorption, and intestinal obstruction without mechanical cause.

Although the literature contains numerous descriptions of ceroidosis, the radiologic features of this condition have not been described.

Case Report

A 39-year-old black male with a diagnosis of Whipple's disease. was admitted to the Cleveland Clinic in April 1964 following laparotomy at another hospital. On physical examination, the abdomen was distended. A diagnosis of intestinal obstruction was made and a laparotomy performed. At laparotomy, the small bowel, large intestine, and the mesenteric lymph nodes appeared to have extensive brown coloration. An ileotransverse colostomy bypassing 4 feet of completely atonic terminal ileum was performed.

Pathologic diagnosis was ceroid pigment accumulation in smooth muscle cells of the muscularis propria of the jejunum, terminal ileum, and colon. The patient was discharged with a diagnosis of malabsorption and intestinal ceroidosis. During further evaluation between October 1964 and January 1965, cystic fibrosis was ruled out by a normal sweet test with pilocarpine iontophoresis; secretion-induced pancreatic amylase, bicarbonate, and juice volume were normal. Alpha tocopherol levels were 58 and 64 μg/100 ml (normal, 500 μg/100 ml).

In May 1968 he was readmitted for intestinal obstruction which was managed conservatively. In January 1970 he was readmitted because of chronic steatorrhea, peripheral edema, and cachexia. Broad-spectrum antibiotic therapy was begun because of suspected blind-loop syndrome. No improvement was noted.

The patient has been followed as an outpatient for the past 11 years. Abdominal distention and severe malabsorption persist, despite therapy.

Radiologic Findings

A barium study done in 1964 showed dilatation of the stomach, duodenum, and small bowel, with thickening of the valvulae conniventes; no nodular lesions were recognized (fig. 1). Diagnosis of mechanical small bowel obstruction was made. Examination of the large intestine showed megacolon (fig. 2).

Striking progressive changes were noted in the barium examination done in 1974. Megaesophagus without peristalsis was present (fig. 3). Massive dilatation of the stomach and duodenum was present with loss of normal mucosal folds (fig. 4*A*). The 24-hr film showed that barium was still in the small bowel and in a very dilated large bowel (fig. 4*B*). A diagnosis of pseudoobstruction was made.

Discussion

Reports of diffuse accumulation of ceroid pigment in the muscularis propria of the small and large intestine have appeared over the past 30 years [4–7]. The macroscopic findings can be so striking at operation or postmortem that the term brown bowel syndrome has been proposed [3].

Ceroid is a lipofuchsin pigment and it is related to long-standing vitamin E defiency [1, 8–10]. Ceroidosis has been found in a variety of diseases such as cystic fibrosis, biliary atresia, celiac diseases [10, 11], sprue [4, 12, 13], chronic pancreatitis [9], granulomatous enterocolitis [14], resection of the stomach and small bowel [2, 8], biliary cirrhosis [8], intestinal lymphangiectasia [15], and Whipple's disease [8].

Both ceroid and the material found within the macrophages of the lamina propria in Whipple's disease react positively to periodic acid–Schiff (PAS); this can lead to an erroneous diagnosis as in this case initially. Melanosis coli should be differentiated also; this involves only the mucosa of the colon, appears secondary to prolonged use of cascara, and is reversible [3]. Ceroidosis is not reversible. The patient showed no improvement after tocopherol administration. This has also been demonstrated in animal studies [3].

It has been postulated but not proven that ceroid deposits affect the function of smooth muscle. The clinical and radiologic manifestations of intestinal pseudoobstruction would thus be explained in our case and in all cases of cystic fibrosis, sprue, and chronic pancreatitis [9]. Radiographically, the most striking features in our case were hypomotility and massive dilatation of the entire gastrointestinal tract. No organic occlusion was present.

The diffuse deposition of ceroid is believed to be the cause of these radiologic findings, which are strikingly similar to those in scleroderma. In both conditions megaesophagus, megaduodenum, and megacolon are present, but the absence of skin changes and the typical histological

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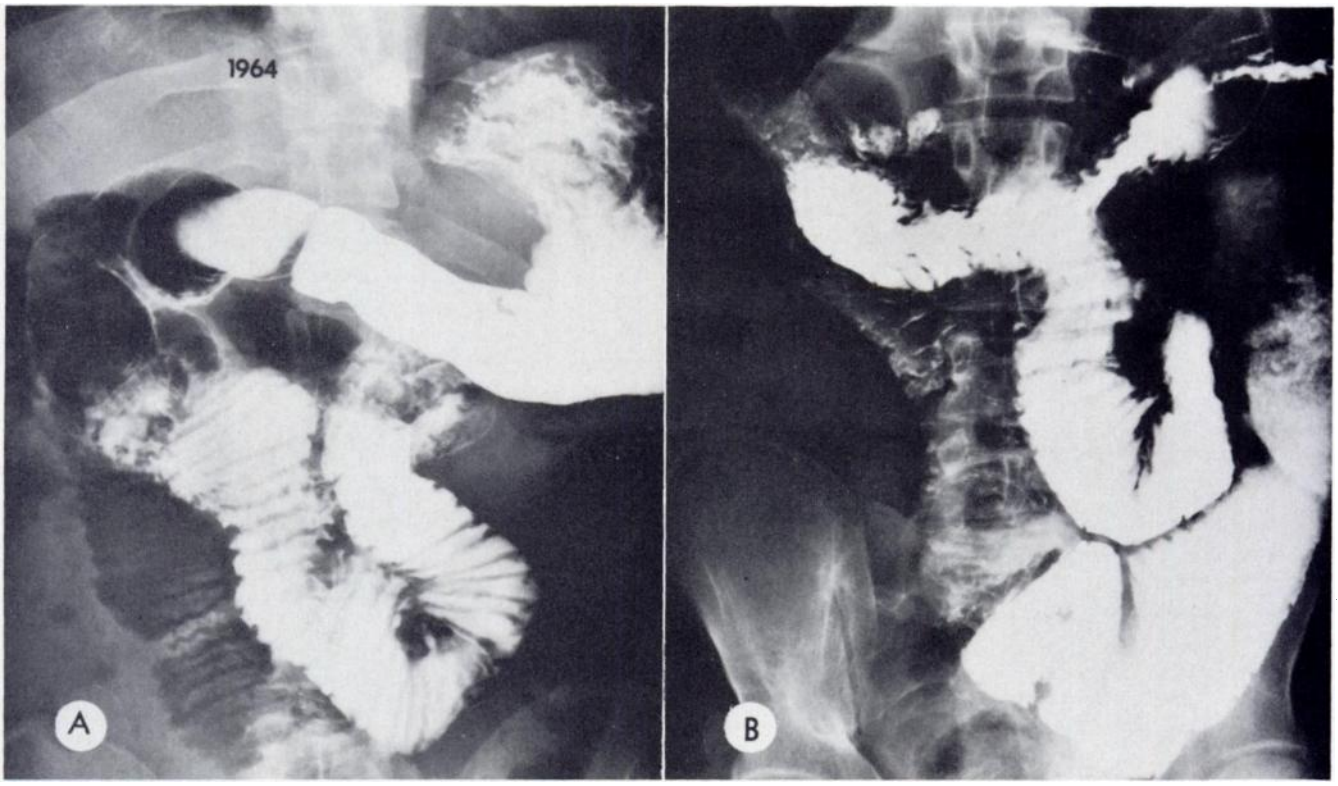

Fig. 1.—*A*, Barium study showing dilatation of stomach, duodenum, and proximal jejunum with thickening of valvulae conniventes. *B*, Follow-up 8½ hr later showing barium still in diffusely dilated jejunum and proximal ileum. Thickening of valvulae conniventes again apparent.

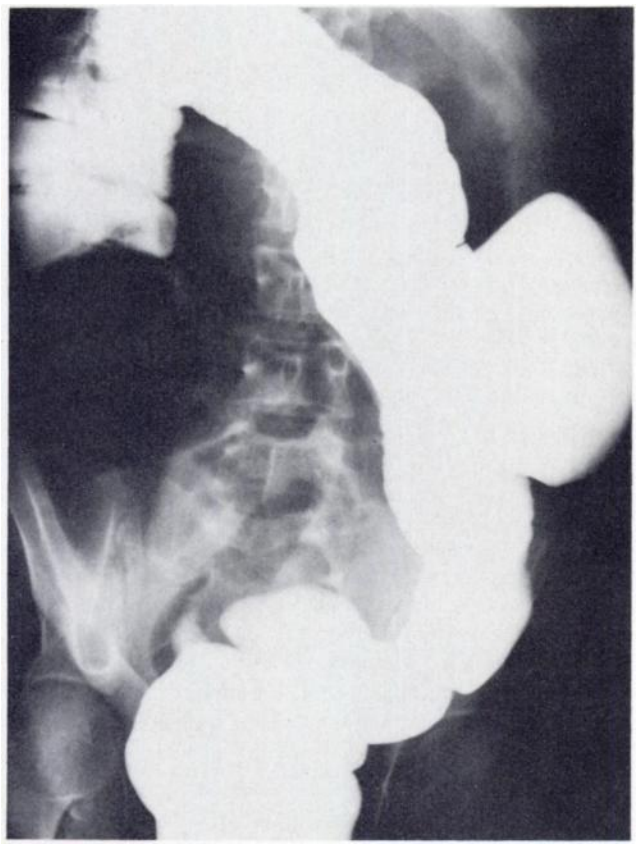

Fig. 2.—Megacolon; no pseudodiverticula are recognized.

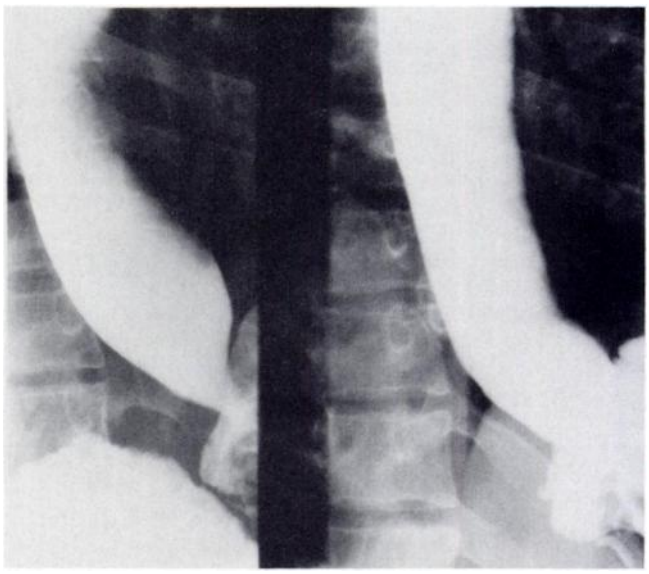

Fig. 3.—Barium study showing massive dilatation of esophagus without stricture formation. Free reflux was noted during fluoroscopic examination.

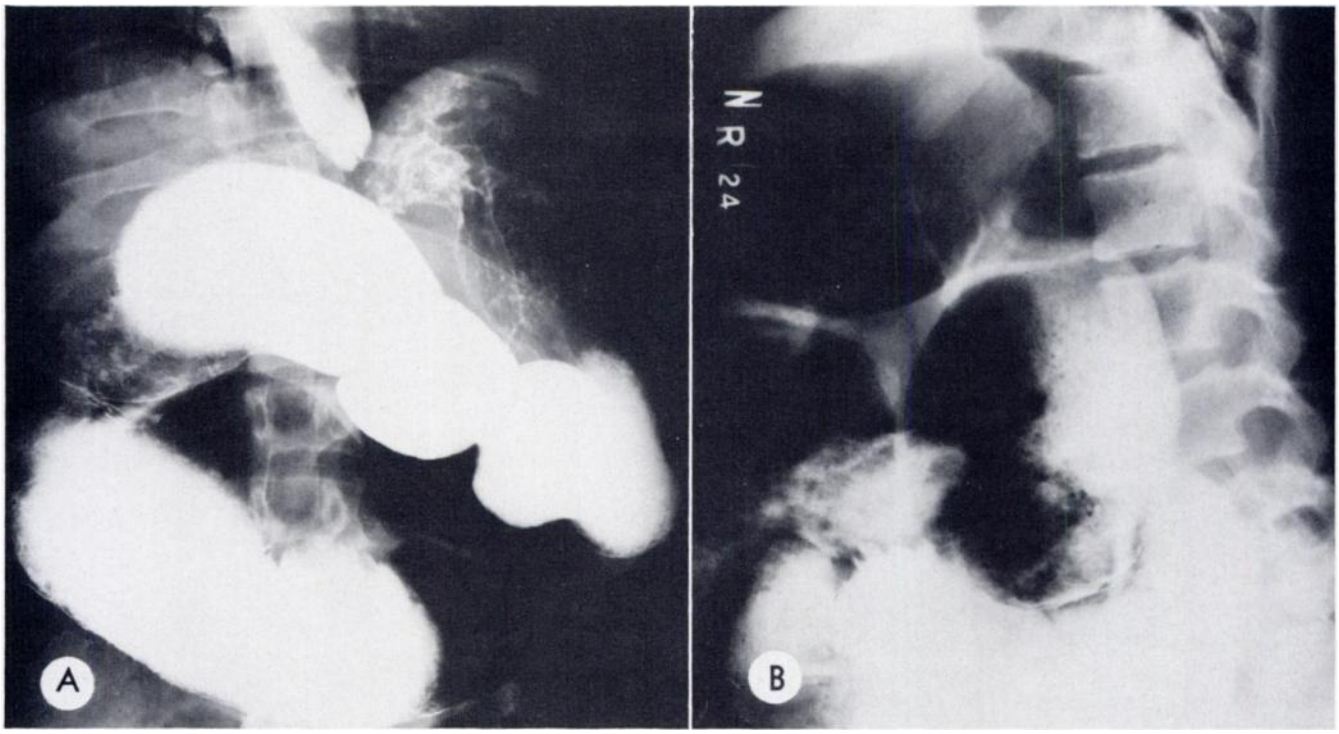

Fig. 4.—*A*, Barium study showing stomach dilated, occupying entire upper abdomen. Duodenum shows more striking widening, measuring 10 cm in diameter, and loss of normal mucosal pattern. *B*, 24 hr follow-up showing barium in bowel. Impossible to differentiate small from large bowel. Note again massive dilatation of duodenum.

findings of scleroderma in the bowel clearly differentiate the two diseases [16, 17].

Chagas' disease and myotonia dystrophica [16] are other entities which can cause megaesophagus and megacolon, but the stomach and small bowel are not affected. Numerous other diseases can cause pseudo-obstruction of the small bowel: pneumonia, renal failure, pancreatitis, myxedema, sprue, amyloidosis, lesions of the myenteric plexus [18, 19], and idiopathic steatorrhea [20].

When dilatation and hypomotility of the entire gastrointestinal tract are present in a patient with a long history of malabsorption and intestinal obstruction, ceroid deposition should be suspected. This can be proven by small bowel biopsy. Major surgery in pseudoobstructed patients may thus be avoided.

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