Letters to the Editor

Sigmoid volvulus as a presentation of neuronal intestinal dysplasia type B in an adolescent

Key words: Neuronal intestinal dysplasia. Sigmoid volvulus. Hirschsprung’s disease.

Dear Editor,

We report a 17-year-old man admitted to emergency department due to abdominal pain and distension. He referred a history of 18 months with constipation. Computerized tomography showed dilatation of the colon and coprostasis. The patient improved after disimpaction with enemas. Colonoscopy and anorectal manometry were reported normal. A full thickness rectal biopsy revealed normal ganglion cells.

The patient was readmitted with a sigmoid volvulus. Sigmoidectomy and a termino-terminal anastomosis (descending colon-rectum) were performed (Figs. 1A and B). Histologically, neuronal intestinal dysplasia type B (NID-B) (NID) was reported (Fig. 1 C). Dehiscence and anastomatic leakage were found along with dilatation of the descending and transverse colon. Subtotal colectomy with a colostomy of the ascending colon and Hartmann’s pouch were performed. The resected segments showed NID-B.

Discussion

NID-B is a hyperplasia of the intestinal submucosal plexus and hyperganglionosis with increased acetylcholinesterase activity in the lamina propria of the mucosa, and the adventitia of the submucosal arteries (1-3). Incidence varies from 0.3 to 60 % depending on the population studied and the diagnostic criteria used (1,2). NID-B presents in children from 6 months to 6 years and rarely occurs in adolescents and adults (1-3). Clinically it is characterized by constipation, ileus, and symptoms that resemble Hirschsprung’s disease (HD) (2,3).

The differential diagnosis includes HD, hypoganglionosis of the myenteric plexus and atrophic intestinal desmosis (4). The diagnosis is histological and quantitative (1,4). In more than half of the cases medical treatment is given with the use of laxatives, enemas, or colonic irrigation. Surgery is used in case of complications or in patients who do not respond to medical treatment (1,3,4). Initial surgical treatment failed because NID-B diffusely affects the intestine requiring a wider resection.
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References