

CASE REPORT

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Cow's milk allergy presenting Hirschsprung's disease-mimicking symptoms

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Abstract The authors present a neonatal case of allergic colitis, which manifested the difficulty of spontaneous defecation and irregular narrowing of distal rectum in contrast enema. Rectal suction biopsy showed positive acetylcholinesterase activity. These clinical, radiological and histological findings were indistinguishable from Hirschsprung's disease. Gastrointestinal symptoms were improved by the cessation of cow's milk formula. The present findings may impact on the less recognizable gastrointestinal manifestation of allergic colitis.

Keywords Cow's milk allergy · Functional intestinal obstruction · Hirschsprung's disease · Allergic colitis

Introduction

Allergic colitis is one of the common gastrointestinal manifestations of cow's milk allergy [1, 2]. During the neonatal period, the most common presentation of allergic colitis is rectal bleeding [2], while Hirschsprung's disease-like symptoms are rarely reported [3–5]. We present a neonatal case of allergic colitis, which mimicked Hirschsprung's disease.

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Case report

A male patient was born at 40 weeks gestation weighing 3,280 g, and was breast fed. At 3 weeks, diarrhea and low-grade fever emerged, followed by abdominal distention. Because the abdominal distention was worsening and the frequency of emesis was increasing, he was transferred to our hospital with suspicion of Hirschsprung's disease at 4 weeks. On admission he was thriving, and showed a distended abdomen. He also presented frequent emesis, but no bloody stools. He required glycerin enema for the difficulty in spontaneous defecation. Blood examinations showed white blood cell count of 12,600 /mm³ with 7.1% eosinophils. An abdominal radiograph showed a dilated colon and intestine. Contrast enema demonstrated an irregular narrowing of the distal rectum with caliber change (Fig. 1). Suction biopsy specimens showed an increase of acetylcholinesterase (AChE) positive nerve fibers in the muscularis mucosae and lamina propria, and it also showed abundant infiltration of eosinophils in the lamina propria (Fig. 2a, b). Manometric studies demonstrated positive anorectal inhibitory reflex (Fig. 3). These findings were contradictory, and the diagnosis of Hirschsprung's disease was not established. Elemental formula and glycerin enemas were initiated, and the symptoms of abdominal distention and frequency of emesis attenuated thereafter. Contrast enema was undertaken at 82-days, and the previous findings seemed to have improved (Fig. 4). However, the patient had been taking elemental diet because a challenge test with regular formula evoked abdominal distention and impaired defecation. The repeated suction biopsy at day 48 demonstrated the same finding as the first study, or increased AChE activity in the muscularis mucosae and lamina propria, although significant infiltration of eosinophils had disappeared. Considering those findings of repeated rectal suction biopsies, radiological findings and symptoms including. Those in the chal-

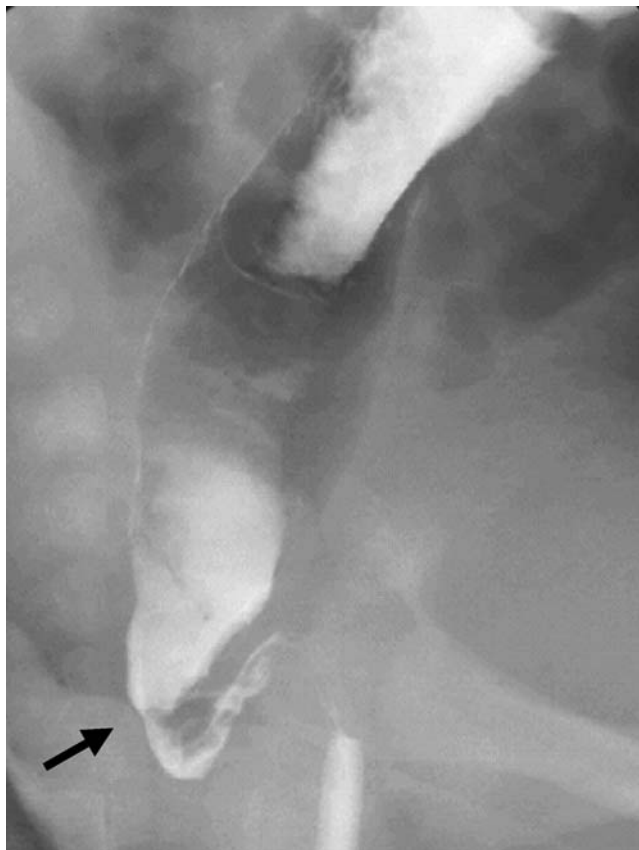


Fig. 1 Lateral view of the contrast enema at day 28. The irregular border and narrowing of the rectum and a transition to the dilated sigmoid colon (*arrow*) are shown. The finding is indistinguishable from that of Hirschsprung's disease

lenge test, Hirschsprung's disease was not completely excluded. Therefore, the Soave procedure was undertaken at 88 days. Histological study of the muscle layer obtained by anorectal myectomy revealed both Meissner plexus and Auerbach plexus, and the diagnosis of Hirschsprung's disease was completely denied. After the operation, The number of daily stool outputs increased and abdominal distention attenuated further. Seven hours after the challenge of cow's milk formula, skin rash and eczema appeared. Immunological tests, including serum immunoglobulin E of 43.8 IU/ml, radioallergosorbent test (RAST)-cow milk of 14.9 UA/ml, and lymphocyte stimulating test (LST) of cow milk of 1,476% confirmed the diagnosis of CMA. After he was fed with elemental formula, he showed no symptoms and thrived well.

Discussion

Allergic colitis due to food allergy is often overlooked because of the diversity and insidious nature of symptoms, and a definitive diagnosis is difficult [6].

Bloom et al. [3] reported four cases of allergic colitis, which presented Hirschsprung's disease-like symptoms. In all of them, contrast enema showed irregular narrowing of the rectum and a transitional zone, which required a differential diagnosis from Hirschsprung's disease. Richards et al. [4] also reported a patient with rectal bleeding and whose contrast enema showed similar findings that Bloom et al. reported. The contrast enema of the present case was quite similar to the findings of their reports. Swischuk et al. [5] reported the findings of contrast enema of their four patients who presented with bloody diarrhea, which showed irregular narrowing of the colon, and mentioned that these findings may have attributed to the segmental colonic spasms.

The increase of AchE-positive nerve fibers in the rectal suction biopsy usually suggests the absence of ganglion cells. The finding of AchE staining in the

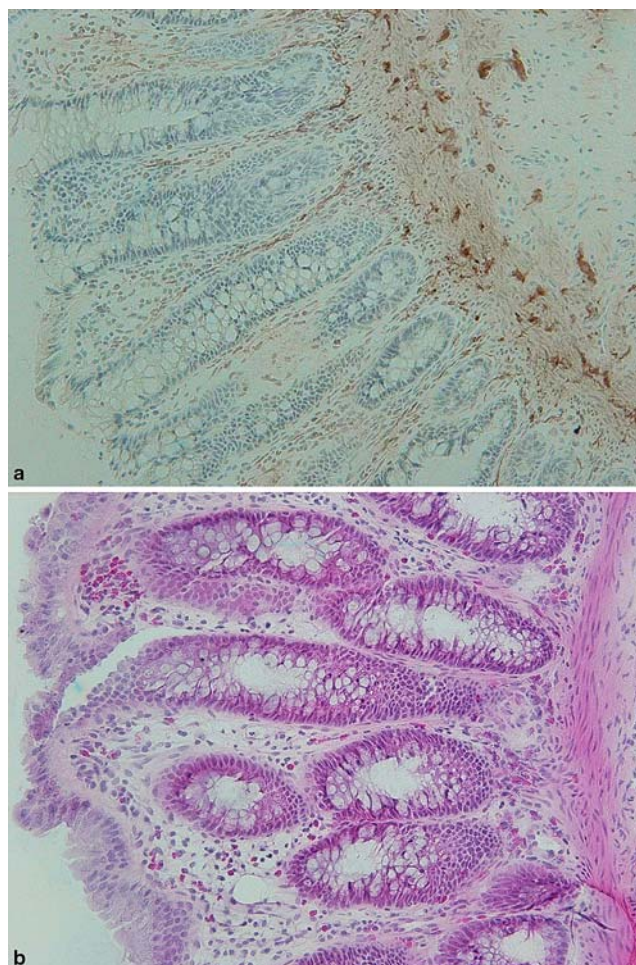


Fig. 2 a Acetylcholinesterase staining of rectal suction biopsy specimens. The obvious proliferation of AchE-positive nerve fibers is seen in the muscularis mucosa, but in the lamina propria the proliferation was limited to the area immediately adjacent to the muscularis mucosa. **b** Hematoxylin and Eosin staining of rectal biopsy specimens. Abundant infiltration of eosinophils is identified in the lamina propria

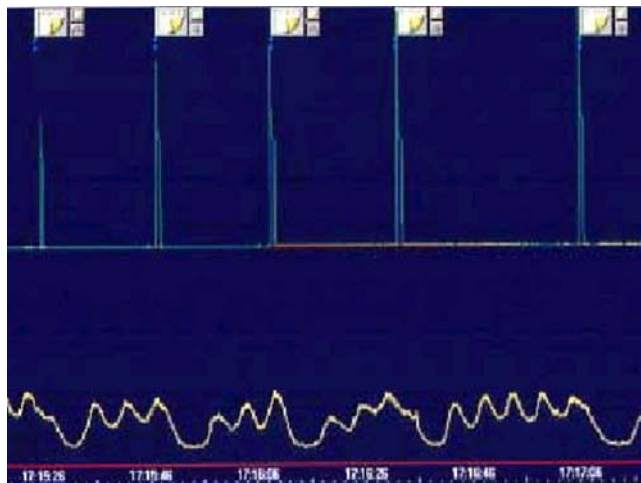


Fig. 3 Manometric study of rectoanal inhibitory reflex. The *upper row* indicates the pressure stimulation on the rectum. The *lower row* indicates the rectoanal pressure. The rectoanal pressure decreases immediately after the pressure stimulation, indicating the positive rectoanal inhibitory reflex

present case was compatible with Hirschsprung's disease, however, the findings of manometric study were against the diagnosis of Hirschsprung's disease. The operative specimens showed normal ganglion cells and the diagnosis of Hirschsprung's disease was denied, which suggested a false-positive AchE staining. Huntley et al [7], investigated pediatric patients with abdominal distention and constipation, and reported the patients who showed false-positive AchE staining. The pattern of AchE staining of these patients demonstrated that an increase of AchE-positive nerve fibers was limited in the muscularis mucosa and the areas of immediately adjacent lamina propria. The pattern of AchE staining of our patient was similar to this pattern. One possible reason for the false-positive increase of AchE-positive nerve fibers is that the dysmotility induced by the immunological inflammatory change of the rectum may have resulted in the increase of nerve fibers. However, there have been no studies that support this hypothesis.

Although the rationale of the emergence of Hirschsprung's disease-like symptoms in patients with allergic colitis has not been elucidated, an inflammatory change of the affected rectum may induce the spastic narrowing of the rectum. Dyschezia of the present case was relieved by the surgery for Hirschsprung's disease. One possible explanation for this is that the continuous colonic spasms might induce an irreversible narrowing or a hypertrophy of the rectum similar to the continuous



Fig. 4 Contrast enema at day 82 shows marked improvement of irregularity of rectum

spasms of the pylorus resulting in hypertrophic pyloric stenosis.

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