Treatment of Adult Hirschsprung's Disease by Botulinum Toxin A through Anorectal Injection

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To the Editor: The patient was a 16-year-old adolescent boy who was diagnosed with congenital megacolon. He had difficulty with defecating for more than 10 years. His bowel movements stopped more than 1 week ago. He was admitted to the hospital with incomplete intestinal obstruction. The patient showed signs of moderate nutrition, poor mental health, and full abdominal bulging and had an abdominal circumference of 104 cm. Rectal examination showed blasting exhaust and defecation, and the anal pressure measurement value was 89 mmHg. The total abdominal augmentation computed tomography and total colorectal sputum angiography showed that the upper and middle rectum, descending colon, transverse colon, and ascending colon were dilated, the maximum diameter of the intestine was 23 cm, and the intestine was filled with feces, suggesting that the lower rectum and sigmoid colon were narrow [Figure 1a and 1b]. Endoscopic ultrasonography of the colon revealed intestinal stenosis at 4 cm above the anal verge, and ultrasonography revealed a thickened muscular layer of 12.3 mm [Figure 1c].

The patient was placed in a supine position, and a total of 100 U botulinum toxin A (BoTox A) was injected into the muscularis propria at 3, 6, and 9 o'clock from the rectal stenosis. The number of bowel movements increased 2 weeks after the injection, the symptoms of bloating were relieved, and the abdominal circumference was reduced to 88 cm. Two months after the injection, a colorectal sputum angiography showed that the lower rectum and sigmoid colon were filled, the intestinal lumen dilation was significantly reduced, and the intestinal peristaltic function was restored [Figure 1d]. Anal manometry showed that the pressure was 53 mmHg. Nine months after the injection, the symptoms of intestinal obstruction recurred. Total colorectal sputum angiography showed that the lower rectum was narrower than the anterior stenosis and that the sigmoid colon was more dilated than before, suggesting disease recurrence [Figure 1e]. Anal manometry revealed that the pressure was 66 mmHg, hence supporting the recurrence diagnosis. Thus, under the guidance of ultrasound colonoscopy, a longitudinal incision of the submucosal muscle of the narrow intestine was performed [Figure 1f]. At present, 2 months after the operation, the follow-up showed that the patient recovered well after surgery, with no postoperative complications.

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Hirschsprung's disease (HD) is generally believed that the intestinal nervous system develops disorders during the embryonic stage that lead to the development without any ganglia of the distal intestinal mucosa and the muscular layer.[1] A recent study has suggested that this may be associated with specific chromosomal abnormalities and syndromes.[2] In 1948, Swenson *et al*. first proposed rectal resection, colonic anesthesia, and anal anastomosis for HD. However, the scope of the surgery was wide and the damage was extensive, thus leading to its gradual replacement. In 1998, Torre *et al*. first reported transanal megacolon surgery. Resection of the diseased stenosis or incision of the sphincter is a traditional surgical procedure for the treatment of HD.

BoTox A has been successfully used to treat skeletal muscle spasm and high-tension diseases in adults and children, such as esophageal achalasia.[3] Langer and Birnbaum first described transabdominal ultrasound localization and anal sphincter injection of BoTox A in children with congenital megacolon causing intestinal obstruction.[4] However, this treatment is technically challenging. The safety of BoTox A depends on the accuracy of the injection site. We used ultrasound colonoscopy to measure the thickness of the sphincter, and under the direct guidance of ultrasound colonoscopy, we adjusted the drug dose according to the thickness of the muscle layer. After the injection, the anal sphincter pressure was reduced by >30% before injection, all of which showed good clinical results. The transient direct effects provided by BoTox A can only last for 3–4 months, but fortunately, the duration of the symptom relief in this patient extended to 9 months. The efficacy of the BoTox A treatment, along with the use of laxatives, antianxiety treatment, behavioral changes, and patient education[5] after the injection, also played a significant role in the improvement of the patient's condition. However, the time of action of the BoTox A drug is limited. As new presynaptic membranes are formed,

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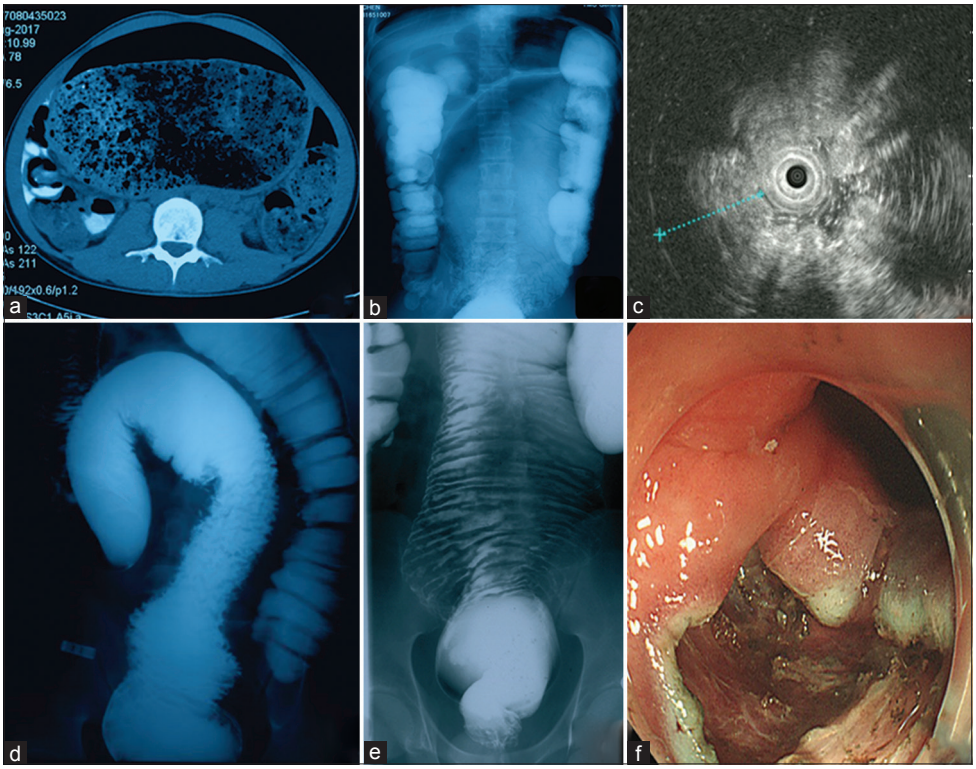

Figure 1: (a) Computed tomography showed dilatation of the intestine, which was filled with feces and had a maximum diameter of 23 cm. (b) Lower rectal stenosis; the upper middle segment is dilated, suggesting that there is stenosis in the rectum. (c) Colonic endoscopic ultrasonography showed local rectal stenosis and thickening of the muscular layer. (d) After 2 months: The lower rectum and sigmoid colon were filled. (e) After 9 months: The colon was once again dilated. (f) Longitudinal incision of the submucosal muscle layer at the site of the stenotic intestine under colonoscopy.

the effects of the treatment will gradually decrease, so clinical treatment often requires repeated injections. Langer has pointed out that BoTox A injections are expected to be effective for muscle incision in the intestinal lesions, probably because both of these methods relieve the high tension of the stenotic myenteric muscles that cause intestinal obstruction. Therefore, in patients taking effective drugs, especially young patients, longitudinal incision of the stenotic submucosal muscle layer under colonoscopy may be indicated. Early surgical intervention is conducive to the recovery of colonic function and may be a better solution for the treatment of this disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient/patient's guardian has given his consent for his images and other clinical information to be reported in the article. The patient/patient's guardian understands that their name and initial will not be published and due efforts will be made to conceal the identity of the patient, although anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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