Long per-rectal endoscopic myotomy for a case of Hirchsprung's disease

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CLINICAL CASE

A 19-year-old woman presented to our hospital with chronic refractory constipation. She was passing hard stools (Bristol type 1 or 2) since childhood with no improvement despite multiple laxatives and required daily enemas to evacuate. A high-resolution anorectal manometry (HR-ARM) (Fig. 1) revealed an absent rectoanal inhibitory reflex (RAIR), suggestive of Hirschprung disease (HD). A barium enema and colonoscopy showed a nondilated sigmoid colon and rectum with a diffusely dilated proximal colon (Fig. 2A and B; Video, available online at www.videogie.org). The transition zone was seen at 23 cm from the squamo-columnar junction. Colonoscopic biopsy specimens were obtained at 3- to 5-cm intervals along the anterior wall of the rectum and sigmoid colon, starting at 2 cm proximal to the squamo-columnar junction. Aganglionosis on histopathology was seen up to 23 cm, confirming the endoscopic transition zone (Fig. 2C and D). A per-rectal endoscopic myotomy (PREM) was planned.

PROCEDURE

Before the procedure, the patient received bowel preparation with polyethylene glycol for 2 days and tap water enemas on the day of the procedure. She received a dose of intravenous ceftriaxone (1 g). The procedure (Video) was performed using a gastroscope (GIF-HQ 190; Olympus, Tokyo, Japan) with a distal cap (Olympus). A submucosal injection of methylene blue and normal saline was given just proximal to the anorectal junction. A transverse mucosal incision was made using a triangular tip knife (Olympus). Submucosal vessels were coagulated with monopolar coagulation forceps (Coagrasper; Olympus). Gentle submucosal dissection was performed with the triangular tip knife using a spray coagulation current (EndoCut-Q; ERBE, Tubingen, Germany, 275 W) delivered through the cautery (VIO 3; ERBE, Tubingen, Germany). Tunneling was done up to 25 cm and the extent was confirmed on the luminal side. Full-thickness myotomy was performed in the oral-to-anal direction with the end of the myotomy just at the mucosal incision site. Mucosal apposition was not possible with standard clips. A clip-loop technique to close the mucosal entry led to invagination of the clips in the tunnel; thus, the clips had to be removed. Tunnel entry was closed by placing clips between the proximal mucosa and the underlying circular muscle.

The patient had no signs of air leak on the third post-procedure day. Given concerns of inadequate tunnel closure, after discussion with the surgery team, previously placed clips were removed and Prolene sutures (Ethicon Inc, Raritan, NJ, USA) were placed. She was discharged the following day.

She presented 2 weeks later with abdominal pain and rebound tenderness. A CT scan showed pneumo-peritoneum. A proctoscopy revealed mucosal dehiscence 2 cm proximal

Abbreviations: HD, Hirschprung disease; HR-ARM, high-resolution anorectal manometry; PREM, per-rectal endoscopic myotomy; RAIR, rectoanal inhibitory reflex.

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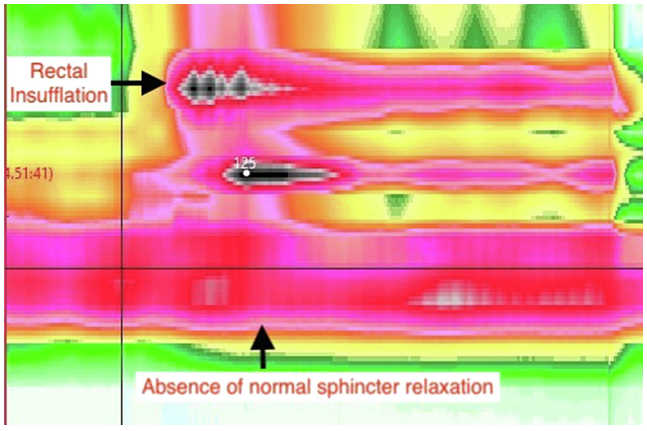

Figure 1. Baseline high-resolution anorectal manometry showing absent rectoanal inhibitory reflex. The balloon inflation led to pressure rise in the rectum (*black arrow*), but it did not lead to relaxation of the anal sphincter (*white arrow*), as would happen in a normal anorectal manometry.

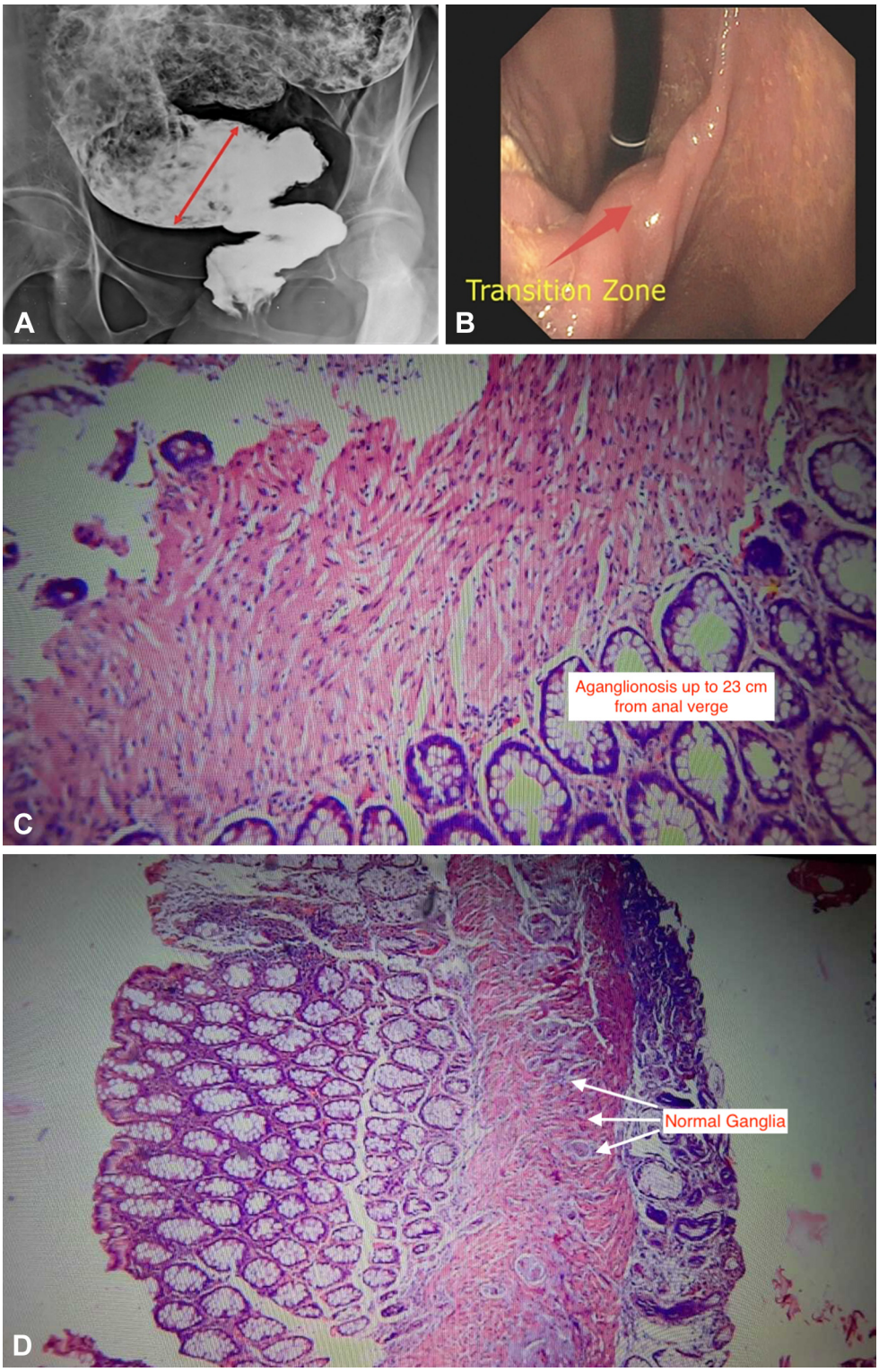

Figure 2. A, A barium enema showed a dilated proximal colon and a narrow distal colon and rectum. **B,** A colonoscopy showed the transition zone at 23 cm. The colonoscopic biopsies confirmed presence of aganglionosis **(C)** to be present up to 23 cm, beyond which ganglia were present **(D)** (H&E, orig. mag., ×100).

to tunnel entry. This may have been caused by the tension from surgical sutures because of a lack of muscular support underneath. The mucosal dehiscence was closed using hemoclips (EZ; Olympus).

OUTCOME AND DISCUSSION

The patient recovered uneventfully and was discharged in 2 days. Nine months later, she was doing well with 1

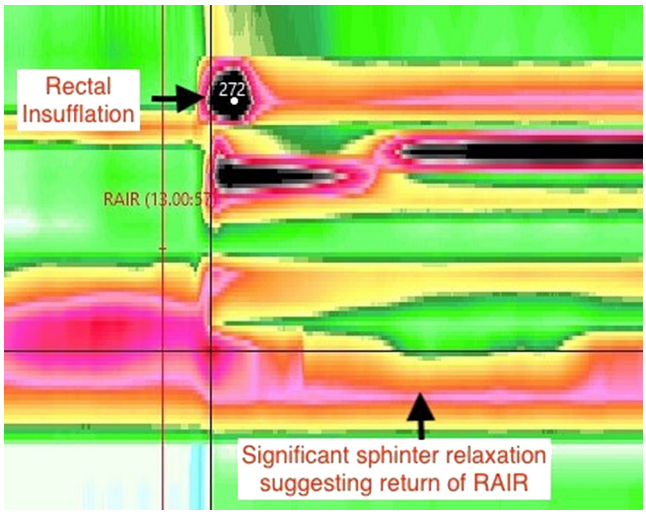

Figure 3. A, The postprocedure barium showed normal caliber of the distal colon and rectum, with disappearance of the transition zone (seen in Fig. 2A). **B,** The anorectal manometry in this visit showed partial return of the rectoanal inhibitory reflex.

bowel motion daily on low-dose laxatives. A barium enema revealed a dilated proximal colon, and an HR-ARM revealed significant improvement in relaxation during straining (Fig. 3). The RAIR was present.

PREM is a new addition to the therapeutics for short-segment HD.[1] The first series of 9 patients was published by another Indian center.[2] The same group also published a case in which they performed PREM for pediatric HD.[3] In these videos, we present the longest successful myotomy completed for HD. The case is also unique, given the delayed mucosal dehiscence, which was successfully managed with clips. With more gastroenterologists choosing PREM for HD, new facets of knowledge are expected to appear about periprocedure care and adverse events.

DISCLOSURE

The authors disclosed no financial relationships.

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