Management and operative strategy for Currarino syndrome associated with thickening of the internal anal sphincter, megarectum and presacral tumor: A case report and literature reviews

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ABSTRACT

Currarino syndrome is characterized by anorectal malformation, a presacral tumor and sacral malformation. A funnel-shaped anal stenosis causes chronic constipation resulting in the development of megarectum and requires surgical intervention. We present a three-year-old girl with Currarino syndrome consisting of a presacral tumor, anal stenosis and megarectum associated with the thickening of the internal anal sphincter. After transverse loop colostomy, excision of the presacral tumor was performed via a posterior sagittal approach and posterior anoplasty with sphincterotomy was done for the anal stenosis. After discharge, anastomotic stenosis in the anal ring remained and anal dilatation, along with closure of colostomy, was performed with an extended skin graft method. Five months postoperatively, there had not been any recurrence of her tumor and she felt the desire to defecate and smoothly defecated with a laxative.

In Currarino syndrome, the typical patient has a combination of sacral, anorectal, intraspinal and presacral anomalies. In most cases of Currarino syndrome, presentation occurs in infancy or childhood. Symptoms such as intractable constipation and bowel obstruction in infancy are frequently associated with this condition [1]. In its classic form, the anorectal anomaly consists of a very characteristic anal stenosis which is funnel-shaped up to the dentate line, resulting in the development of megarectum. The presacral anomaly is described as a mass typically consisting of a teratoma, a meningocele, an enteric cyst or any combination of these [2]. We present a three-year-old girl with Currarino syndrome consisting of a presacral tumor, anal stenosis and megarectum associated with the thickening of the internal anal sphincter, and describe the management and operative strategy.

1. Case report

A girl was born at 39 weeks of gestation, weighing 3010 g with a congenital upper extremity deformity. After birth, she developed abdominal distension and chronic constipation such as once or twice bowel movement a week, but anatomical abnormalities had not been described in her before. Upon admission to another hospital for operations on the congenital upper extremity deformity, she was first recognized to have a bowel disorder. At the age of 3 years old, she was transferred to our hospital for constipation. At the outpatient clinic, physical examination showed the anal orifice was funnel-shaped and stenotic, and was located slightly anterior to the center of the perineum, suggestive of Currarino syndrome (Fig. 1a). Barium enema (Fig. 1b), computed tomography (Fig. 1c and d), and magnetic resonance imaging (Fig. 1e and f) showed megarectum, anal stenosis, a mildly misplaced anus, thickening of the internal anal sphincter, a presacral tumor and sacral malformation. A misplaced anus means that it is centered in the anal sphincter, but there is a shortened perineal body. The stenotic part of the anal canal was recognized to be easily dilated by pulling out an indwelling urinary balloon catheter inserted into the megarectum, but a short stenotic part was identified, which means that the cause of the stenosis is not attributed to organic factors such as fibrotic lesion or scarring (Fig. 1g). From these finding, she was diagnosed with Currarino syndrome consisting of anorectal malformation, a presacral tumor, and sacral malformation associated with the thickening of the internal anal sphincter. She underwent anal...
Fig. 1. a. Physical examination shows a funnel-shaped, stenotic, and anteriorly displaced anus. b. Barium enema shows megarectum, anal stenosis. c, d. Computed tomography reveals sacral malformation consistent with scimitar sacrum, megarectum with a huge fecal mass and a presacral tumor. e. Magnetic resonance imaging shows thickening of the internal anal sphincter and presacral tumor. Arrow indicates the internal anal sphincter. f. The stenotic part of the anal canal is recognized to be easily dilated by pulling out an indwelling urinary balloon catheter inserted into the megarectum, but a short stenotic part is identified, which means that the cause of the stenosis is not attributed to organic factors such as fibrotic lesion or scarring.
bougienage, irrigation of megarectum and then transverse loop colostomy. Two months later, the presacral tumor was resected via a posterior sagittal approach at the jack knife position (Fig. 2a–d). For the anal stenosis, posterior anoplasty with sphincterotomy as the internal sphincter muscles was done (Fig. 2e–h) [3,4]. Pathological review of the resected tumor and sphincter muscle revealed mature teratoma and striated muscle. The resected muscles were identified as the external sphincter muscles. The patient was discharged on the 14th postoperative day. After discharge, she was treated with anal bougienage at the outpatient clinic, but anastomotic stenosis of the anal ring remained (Fig. 3a). Six months later, anal dilatation along with closure of the colostomy was done. By anal bougienage, the mucosa in the posterior wall tore and the scar tissues in the posterior wall were incised longitudinally (Fig. 3b). At that time, the dentate line was exactly identified and sphincterotomy at the internal anal sphincter was re-done. Pathological review of the resected sphincter muscle revealed smooth muscle. In first operation, the resection of the external sphincter was partially performed, but not wholly. Therefore she may be not incontinent, as she would be able to clench her anal sphincter. Furthermore, to complete the anal dilatation procedure, an extended skin graft was performed [5]. Five months postoperatively, there had not been any recurrence of her tumor and she felt the desire to defecate and smoothly defecated using small doses of sodium picosulfate hydrate as a laxative.

2. Discussion

For the treatment of childhood constipation, conservative therapy and surgical methods have been described [6]. In chronic idiopathic constipation and obstructed megarectum, thickening of the internal anal sphincter has been observed [7,8]. Compared with controls, patients with megarectum have significant thickening of the muscularis mucosae, circular muscle, and longitudinal muscle. This thickening is relatively greater in the longitudinal than in the circular muscle [9]. The common treatment of internal sphincter anal achalasia is internal sphincter myectomy [6,10]. An anteriorly displaced anus and congenital funnel anus causes chronic constipation, resulting in development of megarectum requiring surgical intervention. Patients with an anteriorly displaced anus show a prominent posterior shelf and an anteriorly located anal opening on barium enema examination [4,11]. A posterior anoplasty with a posterior rectal myomectomy (internal anal sphincterotomy) is the surgical treatment for this cause of constipation [11,12]. Congenital funnel anus presents as anorectal junction stenosis, in which the perianal skin extends as a funnel up to a stenotic ring, consisting of a fibrotic internal sphincter. In congenital funnel anus, resection of megacolon is mostly performed for constipation [13]. On the other hand, several surgical approaches have been described for the treatment of megarectum with chronic constipation. Resection of megarectum or megacolon is commonly performed. Appendicostomy, coloanal pull-through for Hirschsprung disease, and posterior sagittal anorectoplasty have been frequently performed [13–16].

For surgical treatment of typical Currarino syndrome, defunctioning colostomy appears a reasonable option for megarectum with a huge fecal mass and deferring surgery to excise the presacral mass and repair the anorectal anomaly. The presacral mass and anorectal malformation may be approached through a posterior sagittal approach. In the present case, thickening of the internal anal sphincter with anal stenosis and a presacral tumor were observed. First, the presacral tumor was removed via a posterior sagittal approach. Next, posterior anoplasty was done, but sphincterotomy was performed in error as the external anal sphincter was mistaken for the internal anal sphincter. Finally, because of

Fig. 2. Resection of presacral tumor via a posterior sagittal approach and posterior anoplasty with sphincterotomy. a: Posterior sagittal approach; the incision starts at the inferior border of the coccyx and continues down through the midline, ending at the outer border of the external sphincter of the anus. The posterior parasagittal fibers and the levator ani muscles are divided into equal sides symmetrically. b–d: Resection of presacral tumor; The presacral mass is identified, bordered by the rectum anteriorly, and does not extend up into the spinal cord. The mass is excised completely. Arrow indicates tumor. e: Posterior anoplasty (Cutback Anoplasty); A semicircular incision of the posterior half is made at the mucocutaneous junction of the anus and then incised posteriorly in the midline near the posterior sagittal incision at the resection of the presacral tumor. A posterior mucosal flap is elevated for a distance of 2–3 cm f and g: Sphincterotomy; The muscle layers are partially incised posteriorly in the midline for 1 × 2 cm strip (wide × length) under the mucocutaneous junction of the anus. The resected muscles are judged as the internal anal sphincter. Arrow indicates sphincter muscle of anus. h: The mucosal flap is advanced posteriorly and sutured to the cut edge of the skin.
Fig. 3. Anal dilatation with extended skin graft. a: Postoperatively, anal stenosis remains. b: By anal bougienage, the mucosa in the posterior wall tears. The dentate line is exactly identified. Arrow indicates dentate line. c–h: Extended skin graft; The scar tissues in the posterior wall are incised longitudinally. The muscle layers under the dentate line are definitely recognized as the internal sphincter muscles. The sphincterotomy is done in a 1 × 2-cm strip above the dentate line (c and d). The incision distal to the dentate line never can result in the postoperative incontinence. The edges of the incised mucosa and the incised skin are sutured transversely with interrupted suture (e). The radial skin incision is added from both sides of the advanced mobilized skin (f). Free skin flap formation is done between both radial incisions (g). Mesh-like skin incisions are done appropriately on the free skin flap (h). This technique places the mucocutaneous suture line in a high position by the effect of mesh-like skin incisions and free skin flap within the anal canal postoperatively.
inadequate internal sphincter myectomy and postoperative scarred anastomotic stenosis, an extended skin graft method with internal sphincter myectomy was performed for anal dilatation along with closure of colostomy. She must undergo frequent general anesthesia and operations on the repair of the congenital upper extremity deformity and she was treated with anal bougienage at the outpatient clinic many times, but anastomotic stenosis of the anal ring remained and developed refractory to treatment, and then the closure of the colostomy was simultaneously performed at the time of the second repair of the postoperative anastomotic stenosis. In the case that the anus is in the center of the muscle complex and the thickening of the internal anal sphincter is pointed out, an internal anal sphincterotomy is the treatment of choice for children with an anteriorly placed anus. As posterior anoplasty for postoperative anastomotic stenosis, the extended skin graft (ESG) method was performed. ESG method has been performed for chronic anal fissure with severe anal stenosis. The basis of the operative approach is excision of stenosis and fissure, and internal sphincterectomy. ESG results in the reduction of postoperative pain and shorter healing period by alternative skin graft such as mesh-like skin incisions and free skin flap. She was discharged uneventfully without complications. It is important to determine appropriate surgical methods after due consideration in complicated cases.

3. Conclusion

On Currarino syndrome associated with the thickening of the internal anal sphincter with anal stenosis, internal sphincter myectomy with extended skin graft method was effective to perform anal dilatation.

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Conflicts of interest

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