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Case Report

Anal Canal Duplication in Adults: Report of Five Cases

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Anal canal duplications are very rare noncommunicating second anal orifices located posterior to the true anus. In this study, 5 adult cases of anal canal duplication are reported as extremely rare entities in the literature. The medical records of anal canal duplication patients treated from 2011 to 2014 were reviewed retrospectively. Five adult patients with symptoms of mucous discharge, anal pain, and or perianal fistula/abscess were admitted. Findings of physical examination and radiologic imaging (pelvic magnetic resonance, endoanal ultrasound, and or colonoscopy) suggested anal canal duplication. The mean age of patients was 40.4 ± 8.7 (range, 33–55), and the mean follow-up period was 18.4 ± 11.2 (range, 6–36) months. Histologic features of the removed samples confirmed anal canal duplication. All patients underwent complete surgical excision of the rudimentary anal canal. Anal canal duplication is a very rare congenital anomaly, and 5 additional adult cases are reported. Although this is a referral center, the recent accumulation of 5 adult cases of anal canal duplication suggests that this malformation might be more prevalent and frequently overlooked.

Key words: Anal canal – Anal canal duplication – Congenital malformation

Anal canal duplication (ACD) is an extremely rare congenital intestinal anomaly. It is defined as second anal orifices posterior to the true anus, ending blindly without connection to the rectum, and showing histologic features of a true anal canal, including squamous epithelium at the distal end and smooth muscle cells and anal glands in the wall of the canal.[1–3] This definition differentiates ACDs from rectal cyst duplications, which are cystic structures communicating with the rectum, and hindgut duplications, which include the histologic features of the rectum or colon.[2,4] Symptoms are often absent, but complaints of perianal abscess, discharge, fistula, and or pruritus ani tend to increase with age, as well as the possibility for malignant progression, all of which require surgical

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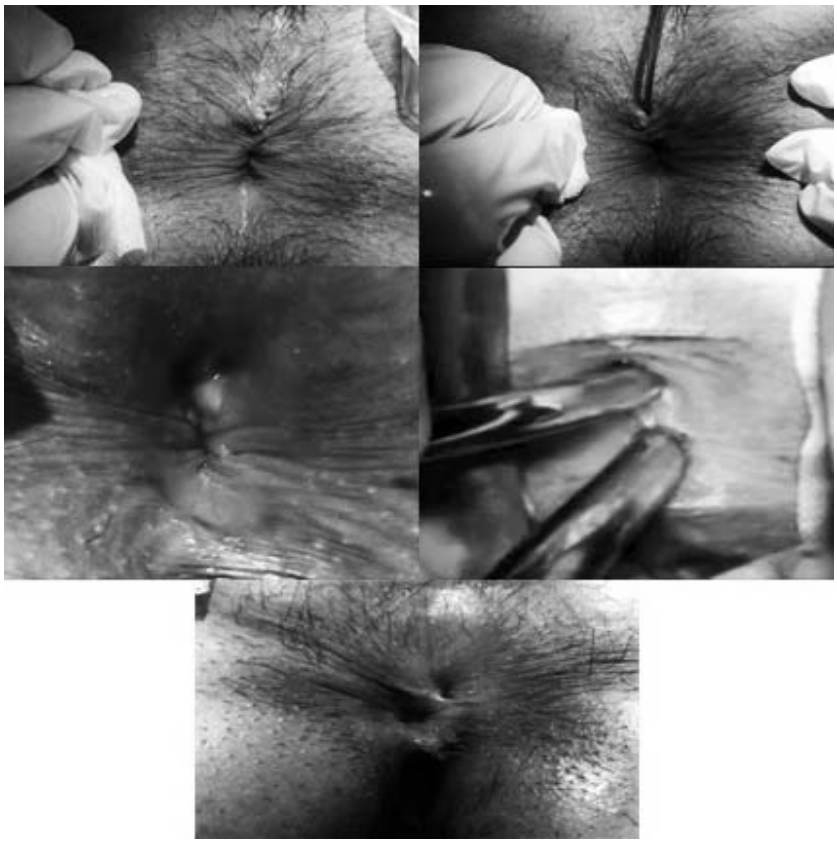

Fig. 1 Some cases were only small, epithelium-lined sinuses on the posterior midline, whereas some readily resembled ACD.

excision of the ACD.[5,6] Endoanal ultrasonography (EAUS), computed tomography (CT), fistulography, and magnetic resonance imaging (MRI) of the pelvis can be performed for differential diagnosis. Symptoms and findings may mimic several anorectal diseases or retrorectal tumors in adults, or malformations such as presacral masses, anorectal malformations, and urinary malformations in the newborn, infants, and children. Here, we report 5 cases of adult patients with the final diagnosis of ACD, with special emphasis on differential diagnosis.

Patients and Methods

The medical records of 5 cases of ACD treated from 2010 to 2014 were reviewed retrospectively to analyze clinical presentation, diagnosis, treatment, and outcome.

Results

Four patients (3 females) were referred from other centers/physicians with preliminary diagnoses of recurrent perianal abscess, fistula, or pruritus ani. Another male patient applied for the first time with symptoms of anal pain and purulent discharge. None had a previous diagnosis of ACD. The mean age of patients was 40.4 ± 8.7 (range, 33–55). On examination, all patients had an accessory opening/sinus appearing more or less as a secondary anus posterior to the true anus. Some cases were only small, epithelium-lined sinuses on the posterior midline, whereas some readily resembled anal canal duplication (Fig. 1). No additional anomalies were noted in this series.

MRI described the lesions in all patients as tubular lesions in 4 patients and cystic in 1 patient, but a definitive diagnosis of ACD was not suggested. EAUS findings were also different and nonspecific (Fig. 2). Interestingly, flexible endoscopy of the ACD was possible in a single case (Fig. 3).

In all cases, ACD was our initial possible diagnosis, and treatment was planned in this order. All patients underwent complete surgical excision. In 3 cases, we placed a small, soft vacuum drain deep into the defect created, whereas in 2 relatively small lesions, the wound was left open. Histology revealed squamous epithelium-lined tracts with anal gland remnants and the presence of smooth

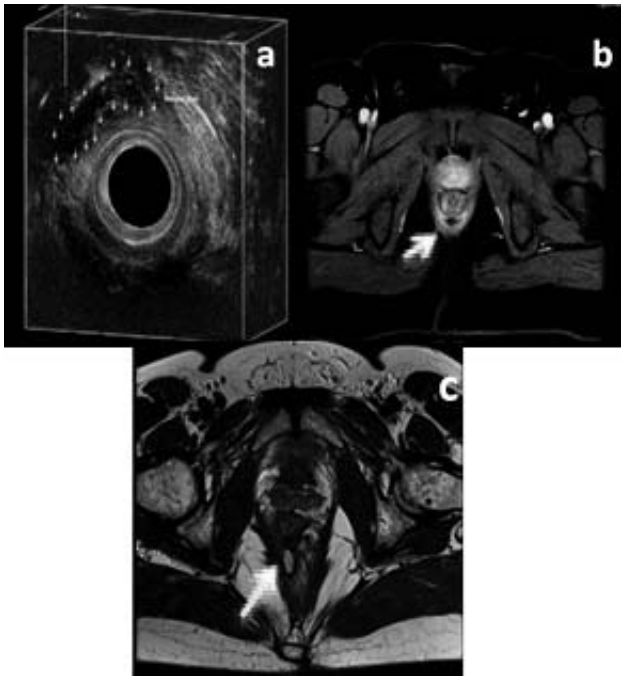

Fig. 2 EAUS (a) and MRI (b, c) views showing (arrows) the ACDs.

muscle within the wall, all consistent with ACD (Fig. 4).

The postoperative course was uneventful with no infectious or other complications. The postoperative recovery period, defined as "feeling safe and comfortable to return to daily activities", was 14.6 ± 5.1 days (range, 8–21), No recurrences were noted within the mean follow-up period of 18.4 ± 11.2 months (range, 6–36).

Discussion

ACD is the least frequent digestive duplication. Choi and Park postulate it as a consequence of recanalization of a cloacal membrane excess in late embryonic life while Hamada *et al* suggest a duplication of the dorsal cloaca in an early developmental stage.[1,2] Clinically, it presents itself as an extra perineal orifice (para-anal sinus) just behind the anus. Most anal canal duplications are tubular in nature (89%), although they occasionally occur with a cystic component (11%).[7] All reported cases have been found either at the level of, or posterior to, the anus, with most presenting along the posterior midline (83%).[7,8] Our series is consistent with these classical features of ACD, all cases being on the posterior midline and most being tubular in nature.

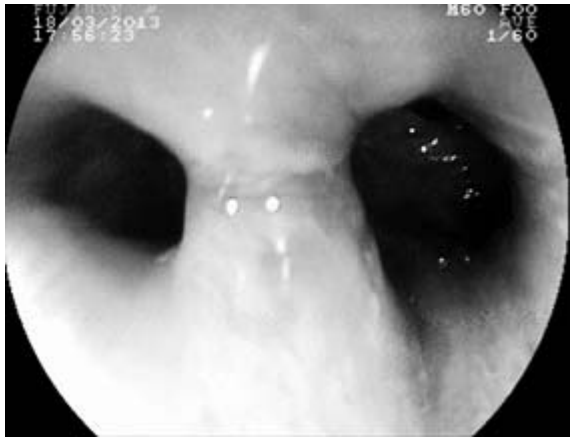

Fig. 3 Colonoscopic view of a prominent case of ACD.

To our belief, the most important clinical feature of ACD is that it's difficult to differentiate it from other incomparably common perianal disease entities, such as (peri)anal abscess, fistula, or pilonidal disease. Although this is a referral center, it's beyond imagination that an additional 5 cases have accumulated in our hands, considering that there have been only 4 adult cases of ACD reported in the literature. It's also inexplicable that we have seen all these cases during the last 3 years and never before. Apparently, this is a typical "*everyone looks but only the cognizant sees*" case. We probably failed to notice many previous cases and mistreated them, as other experts of this field did worldwide. The most obvious case was the first one in this series. This case probably inspired us, anchored ACD as a possible diagnosis in our minds, and it provided our recognition of further cases. The relatively common cases of ACD in infancy and childhood (65 patients reported) possibly stem from the facts that pediatricians are more wakeful about malformations and that other confounding anorectal disorders are rare in this age group.

No additional malformations were noted in any case in our series. ACD can be an isolated anorectal pathology or part of caudal twinning syndrome that is characterized by the presence of twinning of the hindgut derivatives and giving rise to doubling of its derivatives, namely colon, rectum, bladder, urethra, genital organs, and kidney.[9] That is the main reason for evaluation with pelvic MRI although pelvic MRI and EAUS findings are nonspecific for ACD, as exemplified in this study.

Symptoms are often absent but complaints of perianal abscess, discharge, and/or fistula tend to increase with age. The main complications docu-

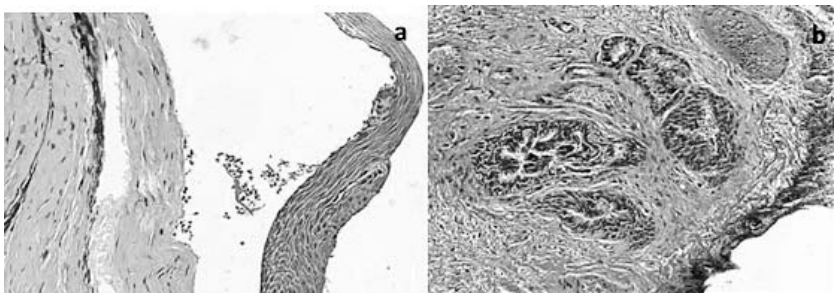

Fig. 4 Nonkeratinized stratified squamous epithelium and adjacent stroma (H&E) (a), and ductus lined with anal type transitional epithelium (H&E) (b), confirming the diagnosis of ACD.

mented in the literature include the risk of infection arising from the accessory anal glands and the possibility for malignant progression.[5,6] It's again possible that such complications further obscure the difficult diagnosis of ACD. The acute clinical picture of a perianal abscess is probably attributed to a cryptoglandular process, and it is urgently solved without further detailed investigation and any histologic evaluation. For the long term, Dukes and Galvin reported malignancy in 8 of 10 adult patients of what they believed to be ectopic tracks of congenital origin.[6] Early diagnosis of ACD is therefore important to reduce the likelihood of complications.

Different treatment strategies are suggested in the literature. The majority of patients received an ACD removal via perianal or posterior sagittal approach.[2,7,10] Mucosal stripping of the ACD is a new, less invasive approach most frequently used when the ACD is very close to the anal canal. Surgical repair is associated with good prognosis and minor surgical sequelae. Only 1 patient reported by Lisi *et al* developed a complication of sphincter insufficiency, which was later surgically repaired.[5] The dissection needs to be tedious, and we encountered no significant complications in our series.

Only histology gives diagnostic certainty describing the characteristics of ACD: squamous epithelium in the caudal end, transitional epithelium in the cranial end, and smooth-muscle cells in the wall of the canal.[3,11] Therefore, every resected piece of tissue needs to be evaluated carefully in proctology. This is again a generally neglected issue, especially when the initial diagnosis weighs in favor of an abscess or fistula.

In conclusion, our series strongly suggests that ACD may be a more prevalent disease entity in adults, and it might frequently be overlooked. Only clinical suspicion and histologic characteristics can lead to the tentative diagnosis of ACD. Imaging studies can only give extra information on the extent of the lesion and concomitant anomalies. Especially if there is a perianal sinus/orifice, posterior to the true anus and on the midline, the possibility of ACD should be kept in mind.

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