

Case Report

Anorectal Strictures and Genital Crohn Disease: An Unusual Clinical Association

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Perianal disease is a well-recognized complication of Crohn disease and is often present when diagnosis is made (1,2). The reported incidence of perianal Crohn disease in the pediatric population varies between 13% and 62% (3–6). Anorectal stricture is a less common manifestation, and rarely are symptoms noted unless complicated by perianal sepsis or fistulae (7,8). Genital involvement is rare in adults and children.

Crohn disease already poses a major medical and psychosocial impact on young people. The additional problems of chronic genital and rectal involvement, poor response to treatment, and need for repeat treatments presents an additional burden.

We report a series of nine young patients with Crohn disease and anorectal stricture, five of whom had associated genital disease. All patients were actively undergoing treatment at the Royal Children's Hospital, Melbourne at the time of reporting, and information was collected retrospectively and prospectively.

CASE REPORTS

Nine patients (5 boys, 4 girls) with Crohn disease and anorectal stricture are described. The mean age at diagnosis was 9.5 years (range, 5–15 years). Five patients had genital involvement. Clinical features are summarized in Table 1.

Anorectal stricture was present when diagnosis was made in four patients; the others developed strictures within the subsequent 4.5 years despite ongoing maximal medical therapy. Strictures were first detected by digital examination in the office and subsequently assessed by examination during anesthesia. In most patients, the strictures were sufficiently tight to prevent the full insertion of the examining finger or the colonoscope. The

characteristics of the strictures are described in Table 2. Severe strictures were defined as those which were tight (limiting digital examination and insertion of a colonoscope), friable with easy bleeding during dilatation, and requiring repeated dilatation in addition to maximum medical therapy.

All dilatations were performed by one surgeon (K.B.S.) using rectal dilators up to sizes 17 to 23, depending on severity.

One patient (patient 3) also had an upper esophageal stricture that remained active requiring repeated dilatation despite the fact his rectal stricture responded well to treatment. Another patient (patient 2) also had a stricture in the terminal ileum requiring surgical treatment. No other patients had additional strictures.

One patient (patient 9) had a rectal perforation. A de-functioning sigmoid colostomy was performed but has had no beneficial effect on the stricture.

Seven patients are still receiving regular dilatation every 2 to 6 months in addition to medical treatment. Six of the patients still have tight strictures.

Five patients have had genital involvement that was present at or before the diagnosis of stricture. Patient 5 had the most florid perianal and genital involvement of all our patients (Fig. 1). She was examined for large fleshy skin tags and bilateral swelling of her labia majora and clitoris 1 year before the eventual diagnosis of Crohn disease and stricture. This improved with medical treatment and regular dilatation during a 3-year period. Currently, she has minimal genital disease but requires regular stricture dilatation.

Patient 6 (Fig. 2) presented 2 years after the diagnosis of Crohn disease with inflammation of the skin of his penis and scrotum, which initially improved with medical therapy. The inflammation was confined to the skin and did not involve the penile shaft. Subsequently, he had four recurrences associated with worsening of the stricture but has improved with regular dilatation, and his genital disease has become quiescent.

Patient 7 developed swelling of the left side of her vulva several months after her initial diagnosis of Crohn

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TABLE 1. *Clinical features in 9 patients with Crohn Disease and anorectal strictures*

Patient	Age at presentation (years)	Sex	Disease localization	Perianal disease	Genital disease	Surgical procedure	Infliximab	Outcome of stricture
1	12	M	Ileum Colon	Anal Fissure, perianal fistula, perineal ulcer	None	Fistula excision Perineal ulcer debridement Stricture dilatation	Yes	Active & persistent
2	15	F	Oesophagus Stomach Ileum, Colon	Anal Fissures, Anal tags, Perianal Abscess	None	Ileo-caecal resection, drainage of abscess, diversion colostomy, Stricture dilatation	Yes	Inactive
3	9	M	Oesophagus Ileum, Colon	None	None	Oesophageal and rectal stricture dilatation	Yes	Improved rectal stricture Active oesophageal stricture
4	9	F	Total GIT	Anal fissures Perianal fistula	None	Subtotal colectomy Stricture dilatation	Yes	Active & persistent
5	8.5	F	Stomach Ileum Colon	Florid skin tags Rectocutaneous fistula to introtitus	Bilateral labial swelling	Stricture dilatation	Yes	Active & persistent
6	9	M	Stomach Ileum, Colon	Anal Tags	Inflammation of skin of penis and scrotum	Stricture dilatation	Yes	Active & persistent
7	11.5	F	Ileum Colon	Anal tags, anal fissures	Left labial swelling	None	No	Inactive & minimal
8	5	M	Oral Rectum	Anal fissure	Scrotal inflammation	Stricture dilatation	No	Active & persistent
9	6	F	Colon	Anal tags and fissures Perianal fistula	Bilateral labial inflammation	Stricture dilatation Colostomy (perforation)	Yes	Active & persistent

disease. With medical treatment, her genital disease improved significantly, and there was no progression of her rectal stricture. She has not required dilatation.

All nine patients have received various combinations of some or all of the local and systemic corticosteroids, 5-aminosalicylic acid preparations, azathioprine, metronidazole, and ciprofloxacin. Those who developed strictures later were already receiving maximal therapy at the time. None had had treatment with infliximab at that stage.

Eight of the nine patients (except patient 7) have required stricture dilatation during general anesthesia, and seven patients continue to require regular dilatation every 2 to 6 months.

More recently, seven patients have been treated with infliximab (5 mg/kg, 1–3 doses). Preliminary results have been disappointing regarding established strictures, although all patients have had a good systemic response. In relation to strictures, three (patients 2, 5, 9) of the seven patients have improved, although one (patient 2) has also had surgery (partial colectomy and ileostomy). One patient (patient 6) has required only minimal dilatation at 3- to 4-month intervals, and his penile disease has been quiescent. Another patient (patient 5) has had the interval between dilatations doubled. Four patients (patients 1, 3, 4, 9) taking infliximab have shown no improvement in strictures to date. Patient 3 initially had a rectal stricture that resolved before the use of inflix-

TABLE 2. *Characteristics of anorectal strictures in 9 patients with Crohn Disease*

Patient	Onset of stricture from diagnosis (months)	Severity #	Average frequency of dilatation (months)	Duration of dilatation (months)	Associated strictures
1	At presentation	Severe	3.2	48	None
2	At presentation	Severe	5.5	24*	Distal ileum
3	At presentation	Severe	2	7	Oesophagus
4	54	Severe	2	2*	None
5	44	Severe	3.2	48*	None
6	28	Severe	2.7	48*	None
7	39	Mild	None	None	None
8	At presentation	Severe	2	15*	None
9	16	Severe	1.7	36*	None

Severe stricture defined as very tight (limiting digital examination and insertion of a colonoscope), and friable with easy bleeding during dilatation. *Ongoing dilatation.



FIG. 1. Severe vulval inflammation and florid perianal skin tags seen in patient 5.

imab, but three doses have had no effect on his esophageal stricture.

DISCUSSION

In this article, we report a series of nine young patients with Crohn disease and anorectal stricture, and the association with genital disease in five patients. All patients with genital disease were examined for or eventually developed an anorectal stricture. In our experience, genital disease was always associated with anorectal stricture, which should be actively sought by clinical assessment or examination with anesthesia.

Anorectal strictures may develop in patients during any stage of Crohn disease, particularly in those with

active perianal disease. Patients may not volunteer any information and may be surprisingly asymptomatic despite tight strictures. Crohn disease should also be considered a possible cause of otherwise unexplained genital disease.

Anorectal strictures have been reported in adults with Crohn disease usually in association with severe perianal disease (8,9). Treatment options include excision and dilatation. Linares et al. (9) reported 44 adults with anorectal stricture. Six patients were treated by rectal excision, 33 by dilatation, and 5 patients did not require treatment. A single dilatation was effective in only one third of patients; most required two or more dilatations. Greenstein (4) described 12 patients with anorectal stricture; 80% of those patients also had associated perianal disease and strictures at other sites in the gastrointestinal



FIG. 2. Penile and scrotal involvement seen in patient 6.

tract. Three patients required repeated dilatation, and two needed surgical diversion because of severe narrowing.

Tolia (6) described 18 pediatric patients (aged 9–18 years) with perianal Crohn disease, of whom 4 had anal canal strictures and needed dilatations (2 uncomplicated, 1 wedge resection with some improvement, and 1 recurrent dilatation along with injection of local steroid).

In our series, all patients but one have required regular dilatations every 2 to 6 months to control the disease. One patient had a diverting colostomy to control perianal sepsis; another required a colostomy after a rectal perforation.

Genital involvement has been described in adults and children (10–18) and may occasionally be the initial manifestation of Crohn disease (12). Different presentations include unilateral or bilateral labial and clitoral swelling and ulceration of the vulva in female patients (10,13). In male patients, scrotal swelling (12) or metastatic involvement of the penile skin and soft tissue (14,15) has been reported. None of our patients was considered to have Behcet disease, which is characterized by discrete vulval or periurethral ulcers.

Our patients with genital Crohn disease demonstrate a unique association, not previously reported, between genital involvement and rectal stricture. The pathogenesis of this association is not well understood. Direct extension of the disease from the perianal region is one possible mechanism. Two of our patients had evidence of rectocutaneous fistulae, but in only one was there an obvious connection to the vulva. Genital involvement may result from an effect on local or pelvic lymphatics or from direct connection through microfistulae.

At present, our preferred treatment for these patients is regular dilatation of strictures and maximum medical therapy. However, it is recognized that this approach carries with it significant actual and potential morbidity with repeated anesthesia, possible hazards of sphincter damage, perforation, and local sepsis. The efficacy of newer medical treatments, such as anti-TNF α , has yet to be fully assessed. Although infliximab has been used effectively to treat patients with generalized and perianal disease, its use in the management of anorectal strictures has not previously been reported. Our preliminary results with established strictures have been disappointing but are subject to ongoing evaluation. However, three of seven patients appear to have had some benefit. It may be that aggressive medical therapy with the newer immunomodulatory drugs may be able to prevent stricture formation and severe perianal disease, if not reverse established strictures.

The psychosocial impact of Crohn disease in young people entering or going through puberty and adolescence is already great. Involvement of the anogenital

region carries with it additional concerns about body image and sexual development. Considerable support is required.

CONCLUSION

Anorectal stricture and genital involvement are rare complications of Crohn disease in children and adolescents. In all patients with genital disease, evidence for anorectal stricture should be sought not only at the time of presentation but also at subsequent evaluations. Regular dilatation and maximal medical therapy is currently the best available treatment option.

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