Seminars in Pediatric Surgery (2007) 16, 185-193

ELSEVIER

SEMINARS IN PEDIATRIC SURGERY

Perianal Crohn's disease

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KEYWORDS
Perianal Crohn's disease;
Anal stenosis;
Anal fissure;
Anal fistula;
Fistulotomy;
Rectal mucosal advancement flap

Perianal Crohn's disease in children is a potentially debilitating condition that can precede or follow the intestinal disease component. The perianal abnormalities are varied and can include lesions of the perianal skin or anal canal, abscesses or fistulas, and malignancies. The appropriate management of these problems is predicated on a thorough evaluation of the perineum and anus as well as the remainder of the alimentary tract. Therapy usually includes a combination of antibiotics, immunomodulators, and biologic agents as well as conservative operative procedures. The surgical options are intended to safely ameliorate disease-related symptoms without compromising function or continence.

Crohn's disease is a chronic, unremitting inflammatory disorder of uncertain etiology that can arise in persons of any age and involve all segments of the alimentary tract plus extraintestinal sites. The treatment of this disorder focuses on alleviating symptoms in the most safe and effective manner using a combination of medical and operative therapy, depending on the patient's age as well as the site and nature of the disease involvement.

Approximately 20% to 25% of patients with Crohn's disease are diagnosed when younger than 18 years of age, and the incidence of perianal involvement in patients with Crohn's disease ranges from 8% to 90%.[1] More specifically, population-based studies suggest that the worldwide incidence of pediatric Crohn's disease is region-dependent, and ranges from 0.2 to 8.5 per 100,00 children[2] with the incidence in the United States reported to be 4.56 per 100,000 children.[3] Moreover, 49% to 62% of these children will demonstrate manifestations of perianal disease.[4,5]

The perianal abnormalities of Crohn's disease can manifest themselves at any time in the disease course, but the intestinal symptoms usually antedate the perianal findings.[6,7] Although the perianal component can be completely asymptomatic in some fortunate children, in others it is the major source of disability.

Although much has been described about the diagnosis and treatment of perianal Crohn's disease in the adult population, relatively little has been published about the management of this disorder in children. Accordingly, many of the recommendations for the pediatric patient must be extrapolated from experience with adults.

Classification

A variety of perianal manifestations can complicate Crohn's disease, including perianal skin lesions, anal canal lesions, anoperineal abscesses or fistulas, anovaginal fistulas, and neoplasia. The skin lesions can be further described as skin tags or hemorrhoids, and the canal lesions can be categorized as fissures, ulcers, or strictures/stenoses. The abscesses and fistulas are typically labeled according to their anatomic location and relationship to the internal and external sphincters. The fissures and ulcers are considered primary disorders, whereas the others are secondary abnormalities.[8]

Skin tags and hemorrhoids

External skin tags are commonly observed as edematous and cyanotic swellings caused by lymphatic obstruction that

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1055-8586/$ -see front matter
doi:10.1053/j.sempedsurg.2007.04.007

occasionally suggests concomitant intestinal inflammation. Conversely, symptomatic internal hemorrhoids are rarely seen prolapsing out the anal canal.[9]

Fissures and ulcers

Anal canal fissures occur with considerable frequency,[10] appear broad-based with cyanotic overhanging edges, and cause no or minimal pain. These fissures are often multiple and eccentrically positioned around the circumference of the anal canal.[11] Cavitating ulcers, on the other hand, are much more uncommon and cause considerable pain as they erode the underlying anal sphincter, which leads to anoperineal abscesses and fistulas.

Strictures and stenoses

The strictures of perianal Crohn's disease can be short, web-like, intraluminal diaphragms positioned near the dentate line or long, indurated, extraluminal areas of stenosis located just above the anorectal ring and often associated with canal ulcers or proctitis.[12,13]

Abscesses and fistulas

Anorectal abscesses and anoperineal fistulas are common findings in patients with perianal Crohn's disease, and their incidence is increased with rectal involvement.[14] The abscesses are commonly painful and often present with coexisting fistulas.[15] The fistulas can either evolve from infection originating in the cryptoglandular area (Figures 1 and 2) or arise from an anal canal fissure or cavitating ulcer.

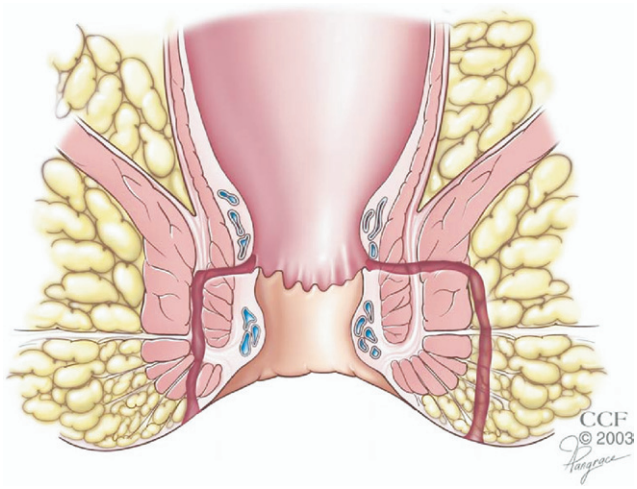

Figure 1 Intersphincteric (left) and trans-sphincteric (right) fistulas are the most common fistulas of cryptoglandular origin complicating Crohn's disease. (Color version of figure is available online.)

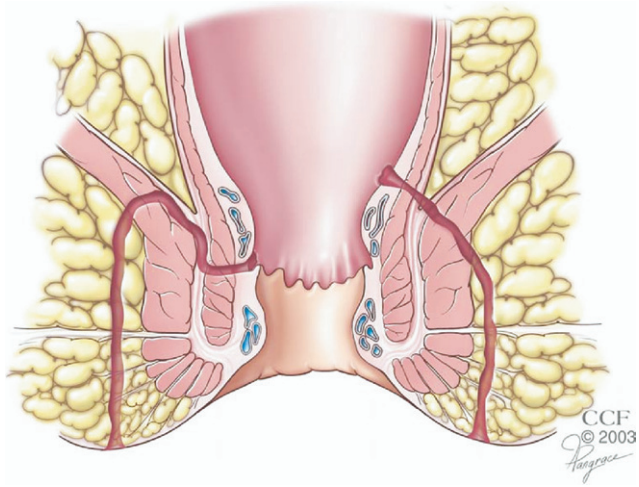

Figure 2 Supra-sphincteric (left) fistulas can originate from cryptoglandular disease or cavitating ulcers, whereas extra-sphincteric (right) fistulas are more commonly associated with significant proctitis or iatrogenic injury. (Color version of figure is available online.)

Anovaginal fistulas

Anovaginal and rectovaginal fistulas can target any level of the vagina and they vary in diameter ranging from <5 mm to >25 mm. The majority of these fistulas are trans-sphincteric in nature and originate from the anterior anal canal at the level of the dentate line.[16]

Neoplasia

Invasive cancer can affect areas of chronic inflammation associated with Crohn's disease of the perianum,[17,18] and the incidence is approximately 0.7% with adenocarcinoma and squamous cell carcinoma occurring in equal frequency.[19]

Classification and scoring schemata

The Cardiff classification schema was initially proposed[8] and later revised[13] to accurately describe the type and objectively score the degree of perianal involvement. The system records each of the major perianal manifestations, assesses the presence and severity of proximal intestinal disease, and grades the global activity of the anal disease. The classification scheme is accurate and comprehensive,[20,21] but it has not been prospectively validated.

A similar scoring system[22] has been more recently proposed, but also considers whether the disease is acute or chronic and primary or recurrent. This scheme appears to correlate with the short-term outcome of patients undergoing surgical management of their perianal Crohn's disease.[22]

An alternative scoring system was previously proposed by Markowitz and colleagues[23] that calculates a disease activity index based on points assigned for the presence and