Fistulizing Crohn’s Disease Presenting After Surgery on a Perianal Lesion

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Perianal skin lesions, such as skin tags, can be an early presenting sign of Crohn’s disease. Surgical intervention on these lesions may increase the risk of fistula development and lead to worse outcomes. This case series examined 8 patients who underwent surgical intervention on what appeared to be benign perianal skin lesions, only to reveal fistulas leading to the diagnosis of Crohn’s disease. This patient population comprised 20% of all pediatric patients with Crohn’s disease who had perianal fistula present at diagnosis. The initial type of perianal lesion varied from case to case and included skin tags, hemorrhoids, and perianal abscesses. All of the patients had other presenting features that, in retrospect, may have been attributed to Crohn’s disease. None presented solely with a perianal lesion. Four patients had weight loss or growth failure. Most of the remainder had abnormal laboratory test results. These findings should raise the awareness of primary care providers that perianal lesions can be the first presenting sign of possible Crohn’s disease in otherwise healthy appearing children. Such children should undergo a thorough evaluation for Crohn’s disease before surgical intervention on perianal lesions because surgical procedures may be associated with worse outcomes.

Crohn’s disease can affect the entire gastrointestinal tract, including the perianal region. Multiple perianal lesions are associated with Crohn’s disease, including fissures, skin tags, hemorrhoids, abscesses, and fistulas. These lesions can develop at any time throughout the disease course, and they occur in 14% to 49% of pediatric patients with Crohn’s disease.1–5 Perianal lesions may be the only symptom present at the diagnosis of Crohn’s disease and can precede other symptoms by years.5–7 In addition, perianal lesions at presentation are more common in children than in adults.8

Due to their heterogeneous appearance, perianal lesions of Crohn’s disease can be mistaken for benign lesions. Nonpathologic perianal skin tags are common and are estimated to occur in 5% to 11% of healthy children.9–10 They are typically midline, small, nonerythematous, and are often caused by constipation. If inflamed, they are painful. These lesions may be operatively removed for comfort or hygiene purposes, although the frequency of such procedures is unreported in the pediatric population. In contrast, skin tags of Crohn’s disease are typically asymmetric, erythematous, can be large (Fig 1), and may be painful or painless.11,12 Hemorrhoids may be associated with Crohn’s disease, proctitis, or portal hypertension but are otherwise extremely rare in early childhood.13 Longstanding constipation can cause hemorrhoids in older adolescents. Although surgical conservatism has been recommended for perianal lesions of Crohn’s disease,9,14 these lesions may be operated on before

abstract

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the diagnosis is established or even considered. This action may lead to the discovery, or development, of perianal fistulas. Such cases are rarely reported.15–17 The present case series describes patients who underwent surgery for perianal lesions not previously recognized to be Crohn’s disease related but subsequently were found to be associated with perianal fistulizing Crohn’s disease.

METHODS

We conducted an institutional review board–approved retrospective study of pediatric patients with Crohn’s disease from January 2005 to February 2014 using the Electronic Medical Record Search Engine.18 Inclusion required presence of a perianal fistula and surgical procedure on a perianal lesion before diagnosis of Crohn’s disease. Patients with a history of inflammatory bowel disease, fistula development later in the disease course, or perianal lesion without fistula were excluded.

RESULTS

A total of 318 patients with Crohn’s disease were identified; 40 (13%) had a documented perianal fistula at diagnosis. Eight (20%) patients, ranging in age from 8 to 17 years, underwent surgical intervention on perianal lesions before the diagnosis of Crohn’s disease (Fig 2). The time from discovery of perianal lesion to diagnosis was 3 to 58 months. Four patients presented with a perianal abscess (Table 1), suggesting that a fistula was likely present before surgery. Three presented with only skin tags, and 1 with external hemorrhoids.

All patients had other findings that, in retrospect, may have been attributed to Crohn’s disease (Tables 1 and 2). Four had weight loss or growth failure, and 3 had abnormal laboratory test results at the time of presentation or surgery. Six had hematochezia. Three patients had constipation, to which their perianal lesions were attributed. However, 1 had abnormal growth, 1 did not have laboratory specimens obtained preprocedure and had abnormal laboratory test results afterward, and another had hemorrhoids (which are exceedingly rare at 11 years of age). In summary, every child had either abnormal weight, growth, or laboratory specimens or other abnormal findings (eg, abscess, hemorrhoid, labial swelling).
Incomplete evaluation was not uncommon. Two patients had no weight or height data recorded at initial evaluation, and 4 had no laboratory evaluations performed. The only patient with normal laboratory test results (patient 6) had no available growth data. Nonadherence was also documented. Two patients, both of whom had abnormal laboratory test results, were nonadherent with recommendations, possibly delaying diagnosis.

The patients underwent a range of surgical procedures before their diagnosis of Crohn’s disease (Table 2). Once diagnosed, 7 of 8 patients were treated with infliximab. The time from Crohn’s disease diagnosis to fistula healing was 8 to 34 months.

**Sample Case Reports**

**Patient 1**

A previously healthy 17-year-old girl presented to the emergency department after 2 weeks of diarrhea, nausea, epigastric pain, and blood per rectum with wiping. Her weight was 64.4 kg (79th percentile). No perianal examination was documented. The patient had microcytic anemia and was prescribed iron supplementation. She was instructed to see her primary care physician (PCP) if symptoms persisted, which she did 3 months later due to continued diarrhea, abdominal pain, and decreased appetite. The results of the patient’s physical examination were normal, although the results of the perianal examination were not documented. She received metronidazole for presumed infectious diarrhea and was referred to pediatric gastroenterology. She was given an urgent appointment, which the family deferred. After 10 days, new painful perianal “bumps” and worsening abdominal pain developed. The patient was seen again by her PCP, who described “2 external...”
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hemorrhoids with shallow ulcerated area between. Topical steroids and sitz baths were prescribed. The patient's perianal pain worsened the next day. Emergency department evaluation reported skin tags and a perianal abscess. When incised and drained, a fistula was identified, and she underwent fistulotomy. Results of the biopsies of the skin tags were nondiagnostic. One month postoperative, the patient had a persistent, open, draining fistula. She underwent fistulectomy and seton placement. Results of the sigmoidoscopy were normal, and biopsy results were nondiagnostic. A few days later, her skin tags were surgically removed. Her perianal pain and discharge persisted for 2 months. Repeat examination under anesthesia revealed nonhealing fistula, and the biopsy specimens revealed inflamed granulation tissue. She was again referred to pediatric gastroenterology.

### TABLE 2 - TREATMENT AND OUTCOMES

<table>
<thead>
<tr>
<th>Patient</th>
<th>Surgical Procedures Before Diagnosis of Crohn's Disease</th>
<th>Presentation to Procedure(s)</th>
<th>Presentation to Diagnosis of Crohn's Disease</th>
<th>Method of Fistula Identification</th>
<th>Initial Crohn's Disease Treatment</th>
<th>Time to Fistula Healing</th>
<th>Method of Assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>I&amp;D abscess, fistulotomy, fistulectomy, seton placement, skin tag removal</td>
<td>16 wk 4.7-kg loss 9 mo 7.7-kg loss</td>
<td>Physical examination/ endoscopy</td>
<td>Infliximab, ciprofloxacin</td>
<td></td>
<td>18 mo</td>
<td>MRE</td>
</tr>
<tr>
<td>2</td>
<td>Hemorrhoidectomy</td>
<td>16 wk 2.4-kg gain 5 mo 0.2-kg loss</td>
<td>MRI</td>
<td>Infliximab, metronidazole</td>
<td></td>
<td>15 mo</td>
<td>Examination</td>
</tr>
<tr>
<td>3</td>
<td>Skin tag removal</td>
<td>90 wk 1.5-kg gain 21 mo 1.2-kg gain</td>
<td>Physical examination/ endoscopy MRI</td>
<td>Infliximab, ciprofloxacin</td>
<td></td>
<td>17 mo</td>
<td>Examination</td>
</tr>
<tr>
<td>4</td>
<td>Skin tag removal x 3</td>
<td>5, 7, and 16 wk Unspecified weight gain 10 mo 2.3-kg gain</td>
<td>MRI</td>
<td>Infliximab, ciprofloxacin</td>
<td></td>
<td>9 mo</td>
<td>MRE</td>
</tr>
<tr>
<td>5</td>
<td>Skin tag biopsy</td>
<td>5 mo Minimal weight gain; exact value NA 6 mo 1.2-kg loss</td>
<td>MRI</td>
<td>Infliximab and methotrexate</td>
<td></td>
<td>8 mo</td>
<td>Examination</td>
</tr>
<tr>
<td>6</td>
<td>I&amp;D, fistulotomy, and seton placement</td>
<td>NA NA NA NA</td>
<td>Physical examination</td>
<td>Infliximab, ciprofloxacin and mesalamine</td>
<td></td>
<td>Not healed at last follow-up (16 mo)</td>
<td>MRE</td>
</tr>
<tr>
<td>7</td>
<td>Fistulotomy</td>
<td>7 wk 0.3-kg loss 58 mo 9.4-kg loss</td>
<td>MRI</td>
<td>Infliximab, metronidazole</td>
<td></td>
<td>34 mo</td>
<td>MRE</td>
</tr>
<tr>
<td>8</td>
<td>Fistulectomy</td>
<td>NA NA</td>
<td>Physical examination</td>
<td>Mesalamine, metronidazole</td>
<td></td>
<td>13 mo</td>
<td>Examination</td>
</tr>
</tbody>
</table>

### Patient 2

An 11-year-old with constipation presented to his PCP with anemia, and the biopsy specimens revealed inflamed granulation tissue. She was again referred to pediatric gastroenterology.

### Table Notes

- I&D, incise and drain; MRE, magnetic resonance enterography; NA, not available.
had resolved, and his pediatric gastroenterology appointment was canceled. After 2 months, the patient developed abdominal pain and a recurrence of hematochezia. At surgery revisit, external hemorrhoids were described as appearing “less swollen and inflamed” and were surgically excised 6 weeks later. During this procedure, the rectal examination produced contact bleeding. Anoscopy revealed friable mucosa, with biopsy specimens demonstrating chronic granulomatous inflammation, suggestive of Crohn’s disease. At follow-up 3 weeks later, the patient’s weight was 35.2 kg (22nd percentile), which was a 0.3-kg gain over 19 weeks. Due to poor weight gain and anoscopy findings, the patient was re-referred to pediatric gastroenterology.

Results of an esophagogastroduodenoscopy and colonoscopy were diagnostic of Crohn’s disease. Magnetic resonance enterography demonstrated a perianal fistula. The patient was started on infliximab and metronidazole. A repeat magnetic resonance enterography 9 months later revealed a persistent fistula. The fistula was closed on external examination 15 months after the diagnosis of Crohn’s disease.

DISCUSSION

Intestinal fistulas are abnormal connections from bowel to skin or other adjacent structures. Fistulas are the most severe Crohn’s disease–related perianal lesion and are associated with aggressive disease. Perianal fistulas can be a source of serious morbidity, causing fecal incontinence, infections, and pain. Severe cases require permanent diverting ostomy. Although prevention would be ideal, few predictors of fistula development exist. Therefore, prompt identification and treatment are critical to limiting morbidity.

Patients with Crohn’s disease often have poor wound healing and high rates of complications after perianal surgery. In addition, surgical intervention on perianal lesions may increase the risk of fistula development. Other than abscesses, most perianal lesions of Crohn’s disease respond to medical therapy without surgery. This finding suggests that surgery for Crohn’s disease–related perianal lesions other than abscesses may not be beneficial and could result in worse outcomes than medical therapy. A clear causal relationship has not been established linking surgical procedures to fistula development, and we could not establish causality in this study.

We identified patients who underwent surgery on perianal lesions and were subsequently found to have fistulating Crohn’s disease. This finding should serve as a reminder that perianal lesions may be a presenting feature, if not the only feature, of Crohn’s disease. Importantly, all the patients had other findings that should have alerted the clinician to the possibility of Crohn’s disease. Of these findings, weight loss and growth failure were the most frequent and objective discoveries suggesting underlying illness.

These patients presented with varying perianal lesions, demonstrating that no single lesion type is associated with Crohn’s disease. The finding of large, discolored, inflamed, or nonmidline lesions should prompt suspicion of Crohn’s disease and encourage review of growth and other symptoms. Painless, enlarged skin tags should also raise concern, as should concomitant genital lesions. All patients should have their weight and growth charts plotted for visual assessment, and all should have screening laboratory evaluations performed (consisting of complete blood cell counts, albumin, sedimentation rate, and C-reactive protein). Fecal calprotectin levels have also been shown to be useful and cost-effective in screening for potential inflammatory bowel disease. A prolonged delay in diagnosis increases the risk of developing more complex fistulas, abscesses, and other disease-related complications, highlighting the importance of having a high index of suspicion for Crohn’s disease; they should also prompt early pediatric gastroenterology referral. Due to concern for worse outcomes after surgical intervention, patients with perianal lesions and any suspicious symptoms should be evaluated by pediatric gastroenterology before surgery. Hemorrhoids in young children or unusual skin tags should be evaluated by pediatric gastroenterology even in the absence of other symptoms. Future investigation is needed to understand the role surgery may play in fistula development and healing.

CONCLUSIONS

This study illustrates that perianal lesions may be 1 of the presenting features of Crohn’s disease. The finding of abnormal perianal lesions should prompt suspicion of Crohn’s disease, especially if other symptoms such as growth failure or weight loss are present. Physicians should consider pediatric gastroenterology evaluation before surgical referral for perianal lesions. Earlier identification of Crohn’s disease may facilitate early medical therapy and avoid unnecessary surgical procedures. Furthermore, earlier identification and treatment of Crohn’s disease may help to prevent fistula formation, whether due to the disease or secondary to a surgical procedure.
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ABBREVIATION

PCP: primary care provider

REFERENCES

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