Chronic Granulomatous Otitis Externa as an Initial Presentation of Cutaneous Crohn Disease

Eileen M. Raynor, MD

In the majority of patients with cutaneous Crohn disease, the gastrointestinal diagnosis is well established prior to the skin manifestation. These patients often present with noncaseating granulomatous lesions involving the groin or scrotal areas. Of the 14 reported cutaneous Crohn disease cases in children, only 1 involved the oral cavity (lip mucosa) and none had facial or auricular lesions.1 Patients who had lesions involving the facial skin and retroauricular area were adults with a long history of previously diagnosed inflammatory bowel disease (IBD).2 The diagnosis of IBD in pediatric patients is generally difficult because of vague symptomatology and lack of substantial gastrointestinal symptoms, leading to a mean delay in diagnosis of 3.5 months.3 In this particular study, none of the 31 patients studied had cutaneous manifestations of their disease.

The present case involves a 10-year-old patient presenting with a several-year history of chronic granulomatous otitis externa and multiple cutaneous lesions involving his scalp, extremities, trunk, and groin ultimately diagnosed as cutaneous Crohn disease. His condition improved when he started receiving tumor necrosis factor inhibitor therapy.

Report of a Case

A 10-year-old boy with a long history of psoriasis involving his scalp, face, and ears who was under the observation of a dermatology clinic presented to the pediatric otolaryngology clinic with a several-year history of recurrent bilateral painful draining ears, as well as skin breakdown and decreased hearing. He was initially treated with 6% salicylic acid for his psoriasis, cephalaxin for secondary bacterial skin infections, and ciprofloxacin and dexamethasone otic suspension for his otitis externa. Culture samples of his otorrhea grew *Corynebacterium*. There was no other pertinent medical history. He has no family history of autoimmune disease or IBD. He had no gastrointestinal complaints (ie, diarrhea, hematochezia, abdominal pain, nausea or vomiting) except for occasional constipation. He also denied tinnitus or vertigo but complained of severe bilateral otalgia, decreased hearing, and drainage.

On physical examination he had erythematous, edematous auricles with copious purulent otorrhea bilaterally (Figure 1 and Figure 2). Otomicroscopy revealed granulation and substantial stenosis of the external auditory canal such that the tympanic membranes could not be visualized. The preauricular creases exhibited severe skin breakdown and ulceration with a separation of the lobule from the facial skin. He did not have any oral or nasal mucosal lesions and had no other lesions on his face or neck. He was taken to the operating room, where he underwent debridement of the external auditory canals and tissue biopsies, as well as culture of the purulent drainage. Findings included severe stenosis of both external auditory canals with granulation tissue and normal tympanic membranes. There was no middle ear effusion or inflammation. There was separation of the helix and lobule from the facial skin due to fissuring and ulceration. Dexamethasone sodium phosphate–soaked wicks were placed into the canals.

IMPORTANCE In the limited number of Crohn disease cases involving the head and neck, there is a predilection for mucosal surfaces and rare reports of involvement in the postauricular region. To our knowledge, in all previously reported cases involving the head and neck, the patients had a known diagnosis of Crohn disease.

OBSERVATIONS This case describes a 10-year-old boy with a history of psoriasis and psoriasiform dermatitis who presented with bilateral chronic granulomatous otitis externa, obliteration of the external auditory canal, and fissuring, resulting in separation of the lobule from the preauricular skin.

CONCLUSIONS AND RELEVANCE Pathologic examination results were consistent with granulomatous dermatitis concerning for cutaneous Crohn disease, and a subsequent gastroenterologic workup confirmed the diagnosis of Crohn disease. This is a report of chronic granulomatous otitis as the initial presentation of cutaneous Crohn disease in a child.

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bilaterally and the helix treated with silver sulfadiazine ointment and MeroGel dressing.

Pathological analysis showed granulomatous dermatitis with suppurative infiltrate, plasma cells, and giant cells. The biopsy also demonstrated overlying spongiosis and neutrophilic infiltrate without foreign-body material. He also underwent biopsy of his groin skin by dermatology clinic staff at that time with similar pathologic findings. These findings were indicative of pyoderma gangrenosum secondary to cutaneous Crohn disease.

After confirmation of the Crohn disease diagnosis with a gastrointestinal evaluation, he was prescribed infliximab and continued receiving methotrexate, as well as cephalexin and tacrolimus topical ointment, with substantial improvement in the cutaneous manifestations. He continues to have moderate stenosis of the external auditory canals but no longer has otorrhea and ulceration. His hearing loss resolved with the resolution of the granulation tissue and otorrhea.

Discussion

Cutaneous manifestation of Crohn disease was first described in 1965 and mainly affects adults with a previous diagnosis of IBD. The majority of the cutaneous lesions involve the groin and anal areas, causing fissuring and scarring. Only 14 cases have been reported in children 18 years or younger. To date, there have been 80 reported cases of cutaneous Crohn disease; however, only 17 preceded the diagnosis of gastrointestinal disease. In the head and neck area, oral mucosal lesions account for 5% to 20% of these manifestations, with up to 90% containing granulomas on pathologic examination. The retroauricular crease, submammary folds, and extremities make up the remaining extragenital regions. Cutaneous manifestations range in severity and pathologic findings, including a psoriatic presentation. It is hypothesized that patients have an immunologic reaction to colonic bacterial–based protein fragments, leading to the cutaneous granulomatous findings.

The prevalence of cutaneous manifestations is essentially the same for Crohn disease and ulcerative colitis (9%-23% vs 9%-19%) despite the difference in genetic loci (IBD1 for Crohn disease and HLA-B27 for ulcerative colitis). There are also differences in the cutaneous lesions, in that Crohn disease lesions often involve mucous membranes and that the severity of the cutaneous lesions does not reflect the gastrointestinal disease state. These lesions range in severity. Erythema nodosum is the most common cutaneous manifestation and involves the extensor surfaces of the lower extremities but may also involve the upper extremities or face. Pyoderma gangrenosum is seen with immune dysregulation, and in up to 50% of patients it is associated with underlying systemic disease including IBD. These lesions present as enlarging ulcers with granulating borders and are commonly misdiagnosed as infectious or foreign-body reactions. Pyostomatitis vegetans affects the lips and buccal mucosa of patients with IBD and is
also believed to be a result of immune dysregulation. Cutaneous polyarteritis nodosa is a recurring vasculitis of the small arteries in the reticular dermis and is distinguished from other vasculitides histologically by panarteritis and perivascular inflammation in the lesion and no activity in surrounding tissue. In cases in which the skin is involved, the severity of the skin lesion is often not correlative with the degree of gastrointestinal involvement.6

Treatment of cutaneous manifestations ranges from treatment of the underlying IBD for erythema nodosum to topical corticosteroids, tacrolimus, infliximab, systemic steroids, methotrexate, and possibly colectomy for recalcitrant pyoderma gangrenosum. Tumor necrosis factor inhibitors (infliximab) are considered first-line treatment for refractory pyoderma gangrenosum. Cutaneous polyarteritis nodosa may be treated with nonsteroidal anti-inflammatory drugs or corticosteroids, whereas pyostomatitis vegetans would be treated more like pyoderma gangrenosum and is only identified in association with IBD. Other associated cutaneous conditions include psoriasis, vitiligo, arthritis, eczema, clubbing of the nails, and acrodermatitis, thereby making the diagnosis more difficult in the absence of gastrointestinal symptoms.5

Conclusions

This case highlights an unusual otologic presentation of Crohn disease in a child with presumed psoriasis and eczematoid dermatitis and a previously unknown diagnosis of IBD whose condition did ultimately respond to tumor necrosis factor inhibitor therapy after the diagnosis of IBD was established. He does continue to have scarring and moderate stenosis of both external auditory canals but no longer has drainage, granulation, or fissurization of the preauricular skin.

REFERENCES