Dear Editor,

Pyoderma gangrenosum (PG) is an ulcerative cutaneous condition of unknown etiology (1). It was first described in 1930 (2) and it is associated with systemic diseases as inflammatory bowel disease (IBD). We present the case of a girl with a perineal PG who after failure of infliximab was successfully treated with adalimumab.

The most common extraintestinal disorders related with inflammatory bowel disease include ophthalmologic, musculoskeletal, dermatologic and hepatobiliary diseases. These extraintestinal disorders can considerably contribute to morbidity and thus damage the overall life quality of the patient, especially in case of a girl.

Case report

This is the case of a girl who is 16 years old at present. She was diagnosed with Crohn’s disease at 9 years of age following onset with fever and abdominal pain. The perineum was very severely affected with loss of dermis and a deep fibrinous ulceration, which had strongly erythematous margins involving from the anus to the vulva (Fig. 1), diagnosed by a dermatologist as a PG, preventing her from leading a normal life, attending classes or mixing with other children of her age. She was treatment with azathioprine without results so she was derivated to our hospital at age of 13 years.

Colonoscopy could not be performed because the perineum and anus were so severely affected that even their examination was very difficult. A contrast-enhanced abdominal computer tomography revealed severe thickening of the ileocecal junction with filiform contrast flow. Scintigraphy with 99m-Tc HMPAO labeled leucocytes was also performed, which showed pathological deposits in the ascending colon and ileocecal valve, descending colon and rectosigmoid. It was decided to start treatment with infliximab in doses of 5 mg/kg at 0, 2 and 6 weeks, with good tolerance and no adverse effects. Subsequent treatment consisted of similar doses every 8 weeks until completing a total of 9 doses. However, during this time and despite a partial response of her disease with a reduction in the number of stools and absence of fever, no improvement was seen in the PG. After the ninth dose, she began to have diarrhea and fever again, and it was decided to change her treatment to adalimumab. Treatment with adalimumab was started with an induction dose of 80 mg sc at week 0, followed by 40 mg sc at 2 weeks and a maintenance dose of 40 mg sc every week. From the onset of treatment, the perianal lesion began to improve and had completely healed in barely 2 months (Fig. 1), thus allowing visualization of the anus and perineum which were previously not visible. There was a marked improvement in the general status of the patient, with normalization of stools and disappearance of abdominal pain and fever. Her nutritional status was recovered and she had menarche after 3 months of treatment. She achieved a very important improvement in her quality of life and was able to attend school.

Discussion

We present the case of a girl with Crohn’s disease and severe perineal PG, with a magnificent response to adalimumab.
Although the incidence of IBD is lower in children than in adults, it is estimated that in up to 25% of cases, the disease begins in childhood (3). In addition, it has been reported that mucocutaneous lesions in children with IBD (4,5); nevertheless, atypical localization as a perineal PG is not really frequent. In Graham et al. review (6), 74% of the children with PG had a systemic illness, most commonly, ulcerative colitis. Our case is a Crohn’s disease. Its review suggests that PG in children has a similar clinical appearance and distribution to that in adults, but PG of the head and face appears to be more common in children.

Our patient has a severe disease whose pathological onset occurs in childhood, requiring continuous corticosteroid therapy and unresponsive to azathioprine. Because of her poor course, it was decided to start biologic therapy, and although she failed to respond to infliximab, with adalimumab the remission was achieved, resulting in successful healing of the extensive perineal lesion suffered by the patient and restoring to her a quality of life that she had lost years ago. The patient was unable to attend to the school and often could not sit down correctly, and only after this treatment was able to do so.

Clearly, the advent of biological therapy has revolutionized the treatment of CD since it has allowed a significant group of patients to be rescued for treatment. Biologic agents have also demonstrated their usefulness in the childhood age group. The REACH study (7) was the first clinical trial conducted in children with biologic therapy, specifically infliximab. Adalimumab, the other approved biologic agent, has demonstrated its efficacy in Crohn’s disease. In addition, it has demonstrated efficacy in extraintestinal manifestations as PG (8,9) and in children. In a recently published case, a child of only 5 years of age achieved remission of his severe perianal disease after treatment with adalimumab (10). Also, biologic therapy improves quality of life in patients, and in our patient case, an improvement in both nutritional parameters and growth was immediate and rapid after the initiation of treatment with adalimumab.

Therefore, and as a conclusion, we report an interesting pediatric case with an atypical PG successfully treated with adalimumab. With this treatment not only the PG achieved remission, but a clear improvement in quality of life was observed, who was able to integrate socially after years of isolation due to her disease.
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