

Hidradenitis suppurativa in a prepubertal girl

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Abstract

Hidradenitis suppurativa (HS) is a chronic and recurrent inflammatory disease characterized by the presence of painful and deep inflammatory lesions usually located in intertriginous areas. It rarely occurs in children, especially in prepubertal age. Treatment for HS in this age group is challenging considering the scant data available and the risk of adverse effects in younger patients. We report the case of a 10-year-old girl with Hurley III HS, refractory to multiple topical and systemic therapies. After introducing adalimumab, there was significant improvement of the skin lesions and therefore in the child's quality of life.

Keywords: hidradenitis suppurativa, skin, prepubertal

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Introduction

Hidradenitis suppurativa (HS) is a chronic and recurrent inflammatory disease characterized by the presence of painful and deep inflammatory lesions usually located in intertriginous areas (1–3). It usually arises between the second and third decade of life, with less than 2% of patients experiencing it before age 11 (1, 4, 5).

Case report

A 10-year-old female was examined at our dermatology department due to perianal nodular lesions evolving over the previous year. She had no gastrointestinal complaints and no other relevant medical history. There was no family history of HS. Clinically, we observed inflammatory nodules of perianal location, Hurley stage II (Fig. 1).



Figure 1 | Perianal inflammatory nodules.

The patient had a body mass index within the normal range (18.45 kg/m²) and a female Tanner stage II. The hormonal study revealed no abnormalities. Pelvic magnetic resonance imaging showed fistulous submucosal pathways (Fig. 2), whereas the upper and lower digestive endoscopic study had no macro- or microscopic changes. Cutaneous biopsy excluded the presence of granulomas and was compatible with the diagnosis of HS (Fig. 3). Initially, she was medicated with topical clindamycin, without clinical improvement. Oral clindamycin and rifampicin were then initiated and maintained for 2 months, with a slight improvement in the inflammation. She also underwent two short courses of oral prednisolone 20 mg during this phase. There was contraindication for treatment with tetracycline due to incomplete dentition. We noted progressive attainment of the perianal area and simultaneous development of vulvar lesions, evolving to Hurley stage III. Menarche occurred in this period. Because there was no significant improvement with previous systemic therapies, adalimumab was initiated at 160 mg, followed by 40 mg weekly. Six months after the introduction of the therapy, there are only three non-inflammatory nodules in the suprapubic skin (Fig. 4), without interurrences related to the medication.



Figure 2 | Fistulous submucosal pathways in the perianal region (T1 C+ Fat Sat).

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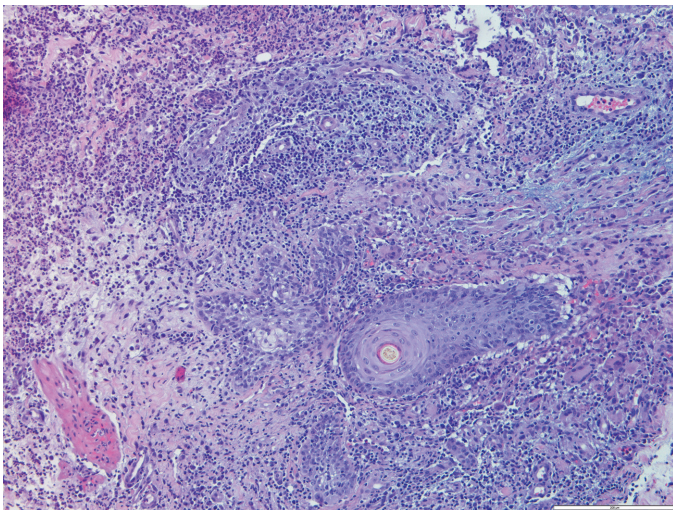


Figure 3 | Mixed inflammatory infiltrate around the apocrine gland with hyperplasia of follicular epithelium and areas of suppuration (H&E, 100×).



Figure 4 | Improvement of the inflammatory lesions after introducing adalimumab.

Discussion

The etiology of HS remains uncertain. Initially, it was linked to inflammation of the apocrine sweat glands (6). More recent studies show that these structures seem to be involved in more advanced stages of the disease, lacking evidence regarding the initial culprit event (6). On the other hand, several studies point to follicular involvement of the pilosebaceous unit as the primary event in the development of HS (6). There is formation of an intraductal plug of keratin, which leads to increased intraductal pressure, dilating the folliculopilosebaceous unit (6, 7). Ultimately, the structure ruptures and the leaked material causes a significant innate immune response (7). This inflammatory reaction clinically manifests with papules, nodules, swollen subcutaneous masses, and

sinuses draining to the surface (7).

HS is rare in pediatric age, especially in prepubertal patients, explaining the scant information in the literature (1, 7, 8). It is predominant in girls, which is in line with data reported in adulthood (2, 9, 10).

Early HS, before 16 years of age, is more frequent in female patients and in those with a family history of the disease (1, 10). Obesity seems to be another risk factor for early onset, whereas smoking shows no influence in these cases (1). Previous studies suggest that early-onset HS correlates with greater extent and severity of the disease (2, 10). On the other hand, there seem to be no differences in the affected areas of the body according to age group (1).

A diagnosis of HS in pediatric age may be a marker of precocious puberty (2, 9). According to previous reports, pediatric disease is more likely to be related to hormonal causes than in adults (2, 3, 8, 9).

The therapeutic approach in this age is complex, especially considering the shortage of studies available (1, 2, 10). Most data are based on small case series, expert opinions, and experience acquired in the treatment of adult HS (2, 8, 10). General measures include a low glycemic load diet and weight loss in obese patients (7, 8, 10). The treatment decision depends on the patient's age, severity and progression of the disease, and previous therapies (1). Whereas mild cases usually benefit from topical conservative treatment, moderate and severe cases require systemic options (2, 7–10). There are several systemic drugs for HS, including antibiotics such as clindamycin, a combination of clindamycin plus rifampicin, and tetracycline (2, 8, 10). Isotretinoin is ineffective in adult HS and therefore is not recommended in children (2, 8, 9, 11, 12). Curiously, there are some previous reports that show that isotretinoin could even contribute to flares in patients with HS (11, 12). The recent recognition of the absence of significant involvement of the sebaceous glands in the pathogenesis of HS may explain these clinical results (12). On the other hand, acitretin seems to be more effective than isotretinoin, although its use in pediatric age should be prudent due to its well-known side effects (8, 10). There are a few reports showing benefit with oral finasteride in recalcitrant cases (4, 5). Short courses of oral corticosteroids may be indicated in acute flares to provide rapid relief of inflammation (10). In moderate to severe cases, biological therapies such as infliximab, etanercept, adalimumab, ustekinumab, and anakinra are also pharmacological options with good response (7, 8, 10). In pediatric age, use of adalimumab is based on previously described cases and experience in adulthood. It is usually indicated in severe and refractory disease and is considered the most effective of the biological therapies (10). Starting with a loading dose of 160 mg and subsequent maintenance with 40 mg weekly, adalimumab does not require dose adjustment to weight (8). Surgical intervention should be considered as the only potentially curative option in HS (8). Ranging from simple de-roofing to extensive excisions, surgical techniques used in adults may also be an option in pediatric patients (8, 10).

HS is a complex disease with a significant impact on the patient's quality of life. Children are a particularly vulnerable group because treatment options may be conditioned by scant available data. Furthermore, HS is extremely rare in prepubertal age, as occurred in our patient. This distinguishes our case report and emphasizes the importance of introducing novel therapies in order to improve the effectiveness of the treatment in younger patients.

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