Hidradenitis Suppurativa

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A 14-year-old girl presented with a history of recurrent bilateral axillary abscesses that had been fluctuating in severity for the past 2 years. The nodules were warm to the touch, with active drainage of malodorous pus and areas of scarring.

History

On 3 previous occasions, she had undergone incision and drainage of the abscesses. She recently received 2 courses of oral rifampicin and clindamycin with partial remission. Her developmental milestones, physical development, and mental development were within normal limits. A review of systems was noncontributory.

She had no history of acne, autoimmune disease, arthritis, or diabetes. She was not taking any medications. Her family history revealed nonconsanguineous and healthy parents and no family history of skin disorders, recurrent abscesses, immunodeficiency, or autoimmune disease.

Physical examination

The patient was overweight with a body mass index (BMI) of 26.4 kg/m2. She was afebrile. Multiple tender, firm papules and nodules were noted in the axilla bilaterally **(Figures 1 and 2)**. The nodules were warm to the touch, with active drainage of malodorous pus and areas of scarring. No nodules were found in the inguinal area, inner thigh, buttock, or inframammary folds. There were no other skin, nail, or hair changes.

The purulent discharge from the axillae was cultured and grew *Staphylococcus aureus* sensitive to rifampicin.

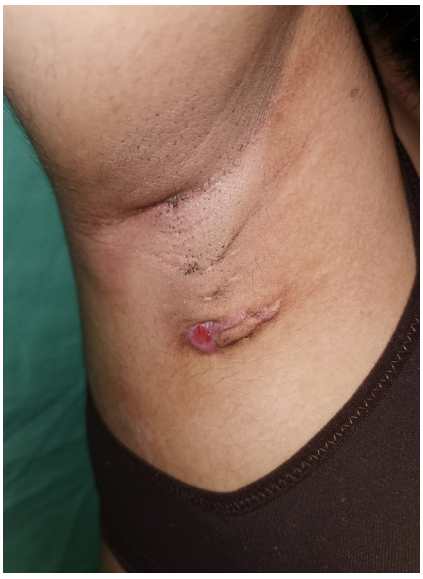

Figure 1. A tender nodule and horizontal scar were noted over the right axillae.

A diagnosis of Hurley Stage II hidradenitis suppurativa (also referred to as acne inversa or, historically, as Verneuil disease) was made based on the triad of characteristic lesions, typical distribution, and recurrence of lesions. An uncomplicated staphylococcal skin infection was unlikely, as the lesions recurred despite appropriate antibiotics.

Treatment and management

Her acute axillary abscesses were treated with incision and drainage. She was prescribed 3 months of oral rifampicin and clindamycin in addition to a daily topical triclosan wash and topical fusidic acid. She was counselled on weight loss, with a referral to a nutritionist, and was advised to wear loose-fitting clothing around the axilla.

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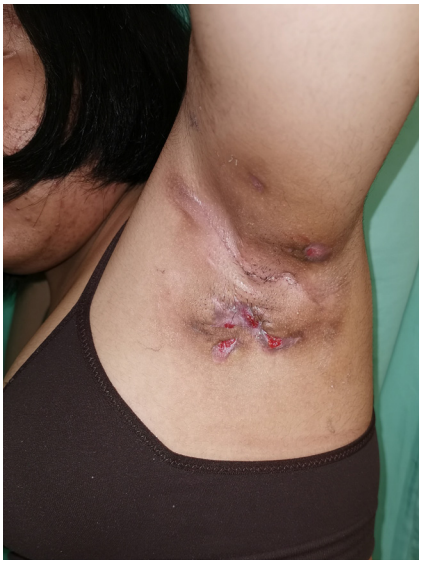

Figure 2. Multiple tender nodules with active drainage of malodorous pus were noted over the left axillae.

Discussion

Hidradenitis suppurativa (HS) is a chronic, painful inflammatory skin condition, whose name originates from the Greek words "hydros," meaning water, and "adeno," meaning gland.[1-3] Local inflammation leads to destruction of the hair follicles and results in tunneling or scarring over the affected area.[2,3] The diagnosis of HS is made clinically based on a triad of (1) characteristic lesions (deep-seated inflamed nodules, abscesses, open comedones, fibrotic scars, and/or sinus tracts), (2) typical distribution over the skin flexures (intertriginous areas: axillae [most common site], groin, inframammary, and buttocks), and (3) a chronic and relapsing course (> 2 times in 6 months).[2-5] HS is associated with significant psychosocial distress because of the pain, appearance, and malodorous discharge of lesions.[6]

HS occurs in approximately 1% to 4% of the population and is often diagnosed between puberty and age 40 years.[1,3] In the pediatric population, the peak age of onset is at 12 years, and the female-to-male ratio is approximately 3:1.[3] HS is underrecognized, and approximately half of pediatric patients already have scarring on initial presentation.[7] The exact etiology is unknown but believed to be linked to genetic and environmental factors.[2,3] Some studies suggest abnormal Notch signaling in hair follicles leads to nodule development and skin inflammation.[4] HS affects individuals of all ethnic and racial groups, but approximately 35% of affected adults and 56% of affected children have a first-degree relative with HS.[2,3] Individuals with HS often have overweight or obesity.[3,8,9] Mechanical stress (friction, pressure) on intertriginous skin may also play a role.

The differential diagnosis includes skin infections, cutaneous Crohn disease, and acne.[2] Skin infections such as furuncles, carbuncles, and abscesses should resolve with appropriate antibiotics and can be excluded.[2] Cutaneous Crohn disease is often accompanied by gastrointestinal Crohn disease and has "knife-cut" ulcers with no comedones.[2] Acne characteristically affects the face and upper trunk, whereas HS affects the skin folds.[2]

The Hurley staging system, which ranges from I (mild) to III (severe), is commonly used to measure the severity of HS based on presence of sinus tracts and scars and to provide management guidelines.[4,5] Other systems include the Severity Assessment of Hidradenitis Suppurativa Score, sartorius hidradenitis suppurativa score, the hidradenitis suppurativa physician global assessment, and the hidradenitis suppurativa clinical response.[1,10]

Pediatric and adult patients with HS often have increased BMI and comorbid metabolic syndrome.[3,8] Additionally, higher BMI is thought to be related to more-severe disease in individuals with HS.[8] Obesity increases surface area, friction, and sweat production within skin folds.[3,8] Obesity can also lead to excess androgen production, and children with HS are more likely to have a hormonal imbalance.[4] Thus, it is crucial to offer children and adolescents with obesity weight management strategies.[8] Children with HS may also warrant referral to an endocrinologist for workup of comorbid hormonal conditions (ie, adrenal hyperplasia, premature adrenarche, metabolic syndrome).[4]

Treatment is based on disease severity with the aim of minimizing pain, inflammation, and scarring; avoiding the need for surgery; and aggressive prevention of disease progression.[4,11] Conservative therapies include weight loss, smoking cessation, and reduced friction in sensitive areas (eg, avoiding mechanical or chemical irritation, wearing loose clothing).[4,8] Options for topical therapies include daily antibiotic lotions, azelaic acid, or aseptic washes (eg, hibiclens, benzoyl peroxide wash, zinc pyrithione).[4,11,12] For moderate HS, intralesional corticosteroids or long courses of oral antibiotics or retinoids may be used.[4,13] For severe or recalcitrant HS, surgery or biologic therapies may be considered. In 2018, adalimumab was approved in Canada and the United States for treatment of HS in children aged 12 years or older.[13-15] Biologics have successfully been used in children as young as 6 years with Crohn disease and 2 years with juvenile idiopathic arthritis, but data is limited.[13,14] Finally, light-based therapies like photodynamic therapy with or without a topical or intralesional photosensitizer and intense pulsed light are reasonable to consider despite a paucity of evidence and guidelines.[16]

Conclusions

It is imperative that HS is recognized early and patients are provided lifelong management and continuous psychosocial support, as HS has a marked effect on quality of life.[4,6] In adolescents, feelings of anger and frustration toward their disease and appearance of skin most profoundly diminishes quality of life.6 Moreover, social embarrassment or physical pain can limit exercise and participation in social activities or school and work.[4,11] Screening for depression should also be considered.[6]

Affected patients typically have a normal life span, but comorbidities are common. Lastly, complications of HS include infection, anemia, arthritis, lymph-

edema, strictures, contractures, and for lesions located in the groin, fistulae to the urethra, bladder, and/or rectum.[4]

References

1. Seyed Jafari SM, Hunger RE, Schlapbach C. Hidradenitis suppurativa: current understanding of pathogenic mechanisms and suggestion for treatment algorithm. *Front Med (Lausanne).* 2020;7:68. https://doi.org/10.3389/fmed.2020.00068
2. Saunte DML, Jemec GBE. Hidradenitis suppurativa: advances in diagnosis and treatment. *JAMA.* 2017;318(20):2019-2032. https://doi.org/10.1001/jama.2017.16691
3. Ballard K, Shuman VL. Hidradenitis Suppurativa. In: *StatPearls.* StatPearls Publishing; August 11, 2021. http://www.ncbi.nlm.nih.gov/books/nbk534867/
4. Liy-Wong C, Pope E, Lara-Corrales I. Hidradenitis suppurativa in the pediatric population. *J Am Acad Dermatol.* 2015;73(5 Suppl 1):S36-S41. https://doi.org/10.1016/j.jaad.2015.07.051
5. Lee EY, Alhusayen R, Lansang P, Shear N, Yeung J. What is hidradenitis suppurativa? *Can Fam Physician.* 2017;63(2):114-120. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5395382/
6. McAndrew R, Lopes FCPS, Sebastian K, Diaz LZ. Quality of life in hidradenitis suppurativa: A cross-sectional study of a pediatric population. *J Am Acad Dermatol.* 2021;84(3):829-830. https://doi.org/10.1016/j.jaad.2020.09.072
7. Liy-Wong C, Kim M, Kirkorian AY, et al. Hidradenitis suppurativa in the pediatric population: an international, multicenter, retrospective, cross-sectional study of 481 pediatric patients. *JAMA Dermatol.* 2021;157(4):385-391. https://doi.org/10.1001/jamadermatol.2020.5435
8. Reichert B, Fernandez Faith E, Harfmann K. Weight counseling in pediatric hidradenitis suppurativa patients. *Pediatr Dermatol.* 2020;37(3):480-483. https://doi.org/10.1111/pde.14131
9. Buimer MG, Wobbes T, Klinkenbijl JH. Hidradenitis suppurativa. *Br J Surg.* 2009;96(4):350-360. https://doi.org/10.1002/bjs.6569
10. Hessam S, Scholl L, Sand M, Schmitz L, Reitenbach S, Bechara FG. A novel severity assessment scoring system for hidradenitis suppurativa. *JAMA Dermatol.* 2018;154(3):330-335. https://doi.org/10.1001/jamadermatol.2017.5890
11. Choi E, Ooi XT, Chandran NS. Hidradenitis suppurativa in pediatric patients. *J Am Acad Dermatol.* 2020;S0190-9622(20)32436-1. https://doi.org/10.1016/j.jaad.2020.08.045
12. Alikhan A, Sayed C, Alavi A, et al. North American clinical management guidelines for hidradenitis suppurativa: A publication from the United States and Canadian Hidradenitis Suppurativa Foundations: Part I: Diagnosis, evaluation, and the use of complementary and procedural management. *J Am Acad Dermatol.* 2019;81(1):76-90. https://doi.org/10.1016/j.jaad.2019.02.067
13. Rundle CW, Price KN, Hogeling M, Hsiao JL, Shi VY. Recent advances in hidradenitis suppurativa in pediatrics. *Dermatol Online J.* 2020;26(12):13030/qt5gt6m9n3. https://doi.org/10.5070/D32612051346
14. Bi Y, Liu J, Wang J, et al. Model-informed drug development approach supporting approval of adalimumab (HUMIRA) in adolescent patients with hidradenitis suppurativa: a regulatory perspective. *AAPS J.* 2019;21(5):91. https://doi.org/10.1208/s12248-019-0363-5
15. Humira (adalimumab). Prescribing information. Abbott Laboratories; 2011. Accessed November 8, 2021. http://www.accessdata.fda.gov/drugsatfda_docs/label/2011/125057s0276lbl.pdf
16. Gracia Cazaña T, Berdel Díaz LV, Martín Sánchez JI, Querol Nasarre I, Gilaberte Y. Systematic review of light-based treatments for hidradenitis suppurativa. *Actas Dermosifiliogr (Engl Ed).* 2020;111(2):89-106. https://doi.org/10.1016/j.ad.2019.04.008