Pedi-Gyn Derm

What’s Your Diagnosis? Painful Nodules on the Perineum with Scarring and Sinus Tract Formation

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Introduction

Hidradenitis suppurativa (HS) is an androgen dependent acneiform eruption primarily affecting areas with apocrine hair follicles, such as the axilla, under the breasts, and the perianal and perigenital regions (Figs. 1–3). It is a chronic condition that is very difficult to treat. Sequelae in the most severe cases include painful and disfiguring scarring and an increased risk of skin cancer in the affected areas.

Questions for the Clinician

What’s your diagnosis? Would you recommend a biopsy or a culture in this case? What treatments would you consider? (1) Medical (2) Surgical

Differential Diagnosis of Painful Vulvar Nodules

The differential diagnosis in this case is somewhat limited. It includes hidradenitis suppurativa and cutaneous Crohn’s Disease. Interestingly, there have been several case reports describing an association between Crohn’s Disease and hidradenitis suppurativa.¹

Evaluation of a Girl with Painful Perineal Nodules

History

Key questions include:

How long have the lesions been present? Hidradenitis occurs most frequently in females between the ages of 11–20 years. It is thought to be an androgen-dependent condition, and only 2% of cases occur before age 11, although only some patients show elevated levels of androgens. The importance of finding out how long the patient has had the lesions is to find out if they appeared before or after adrenarche and menarche. It would be usual for hidradenitis to appear before these two development stages. The earliest case of HS reported was in an infant with congenital adrenal hyperplasia.²

Are there other areas of involvement? Although hidradenitis can occur only on the perineal area, a history of lesions also occurring in the axillary vaults or under the breasts can help to confirm the diagnosis.

What treatments have been tried previously, both medical and surgical? Many individuals with hidradenitis will have sought therapy from other doctors prior to seeing you. It is obviously important to know which treatments they have tried and which if any have worked. If the patient has seen their primary care provider, they probably received short courses of oral antibiotics such as cephalaxin or amoxicillin. If they were seen by a surgeon, they may have had several lesions excised. Lastly, if they have ever had significant pain

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associated with their lesions, they may have been seen in a local ER for incision and drainage.

Physical Examination
Hidradenitis is considered to be one third of the follicular occlusion triad. The other two diseases are Acne Conglobata, which is a very severe scarring form of inflammatory nodulocystic acne, which can affect the face and trunk, and perifolliculitits capitis, also known as dissecting cellulitis or folliculitis of the scalp. This later condition often leads to scarring scalp alopecia. Any patient with HS should be evaluated for additional evidence of the follicular occlusion triad. Hidradenitis can certainly occur independent of the other two conditions, and in those cases, the extent of the HS must be determined before a treatment plan is finalized. Hidradenitis commonly affects the axillary vaults, the areas under the breasts of developed females, and both the perianal and perigenital areas. Any combination of these areas can be affected. The clinical exam can show areas of erythema and or induration studded with follicular pustules and or abscesses, as well as sinus tracts and hypertrophic or keloid scarring. If only one area is involved, the patient may be a good candidate for surgical therapy, while more widespread diseases should probably be treated with medical therapy.

Treatment

Medical
The first medical treatment option pursued is often oral antibiotic therapy. Although short-term oral antibiotic therapy is often tried initially, these patients generally need several-month courses of antibiotics, similar to the courses used to treat acne. A study done on the microbiology of HS showed an average of 2.5 bacterial isolates per specimen. This finding suggests that either antibiotics with a broad spectrum of action, or a combination of oral antibiotics should be used.

Oral retinoid therapy has also been used to treat the more severe forms of HS. Although there are several oral retinoids currently on the market, based on the issues of teratogenicity and drug half life, isotretinoin is the treatment of choice in women of child bearing potential. One study showed significant benefit by combining a short course of oral steroid with long term oral isotretinoin therapy.

Finasteride, which is an oral antiandrogen, has been reported to be successful in treating one case of HS in a 55-year-old female. Lastly, infliximab, which is a monoclonal antibody against tumor necrosis factor, has also been reported to be an effective treatment for severe forms of HS. Five of the six reported patients have been women, including one with co-existing Crohn’s disease.

Surgical
For hidradenitis that involves only one or two body sites, many believe that complete surgical excision of the involved area is the treatment of choice. In one of the largest case series reported, 106 patients, 61 of whom were women, underwent complete surgical excision. In this series the recurrence rate was only 2.5% and the infection rate was only 3.7%. Although the method of reconstruction varied in this case series based on the size and location of the area excised, in all cases, the extent of sinus tracts was determined.
intraoperatively with use of dye. Excision was considered complete when all dyed areas were removed. The other important surgical issue related to this is the development of Squamous Cell Carcinoma in the skin in areas of scarring. A large cancer incidence study done in Sweden showed an increased risk of genital non melanoma skin cancer and breast cancer in patients with hidradenitis.

The problem of squamous cell carcinoma of the skin arising in an area of chronic scarring is not limited to hidradenitis. This phenomenon has been seen in burnt out lesions of discoid lupus and in old burn scars, where the name Majorlin’s ulcer has been used cursorily.

Two other procedurally oriented therapies have also been used to treat hidradenitis. In a small study of 10 patients with persistent painful nodules of hidradenitis, cryotherapy was effective in 8 out of 10, and 7 out of 8 voted it as more effective than oral antibiotics. However, the procedure was considered to be more painful in the short term, and healing on average took greater than 3 weeks. Lastly, intralosomal steroid injections, similar to those given for acne cysts, have also been used.

**Conclusion**

Hidradenitis suppurativa is a chronic scarring condition primarily affecting areas with apocrine hair follicles, such as the axillary vaults, under the breasts, and both the perianal and perigenital areas. The scarring can be both painful and disfiguring. Early aggressive treatment can spare some patients from these poor outcomes. Treatments range from oral antibiotics through to radical surgery. The decision as to which treatment to pursue must take into account the areas involved, and the overall extent of that involvement, as well as the comfort level and skill of the provider with each of the different therapies.
Fig. 3. Chronic scarring and sinus tract formation in the axilla.

References