Hidradenitis suppurativa at the knees

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Abstract

Hidradenitis suppurativa is a poorly understood, destructive disease centered on pilosebaceous units and characterized by inflammatory nodules that progress to abscesses, sinus tracts, and scars. The typical patient is a young woman with involvement of the axillae, breasts, and groin. Presented is a 60-year-old man with poorly controlled diabetes, who initially developed pink-to-yellowish plaques with punched out ulcers on his knees that evolved to scars and draining sinus tracts. Draining sinus tracts were also noted under the left axilla. Histopathology revealed suppurative granulomatous inflammation centered on hair follicles as well as sinus tracts. Special stains for fungus and mycobacterial bacilli were negative. Cultures and PCR for mycobacterial tuberculosis were negative. The patient was diagnosed with hidradenitis suppurativa. A treatment trial of topical corticosteroids and antibiotics was given, but this regimen failed to improve his lesions. The patient was then started on adalimumab, which yielded marked improvement within three months. This case is reported because of the unusual clinical presentation and to highlight the spectrum of atypical hidradenitis suppurativa.

Keywords: hidradenitis suppurativa, knees, follicular HS, adalimumab

Introduction

Hidradenitis suppurativa (HS), an inflammatory condition clinically characterized by nodules that eventually progress to abscesses, sinus tracts, and scars, most commonly affects the axillae, breasts, and groin. This disease is believed to stem from follicular rupture causing release of keratin and bacteria into the surrounding dermis. Debris release sparks chemotactic response and subsequent formation of abscesses and sinus tracts. This disease generally affects young women and is exacerbated by smoking and obesity.

Canoui-Poitrine et al. divided HS into three phenotypes according to distribution of lesions: axillary-mammary, follicular, and gluteal [1]. The axillary-mammary phenotype, located predominantly in the underarm and breast regions and characterized by hypertrophic scarring, is found in roughly half of HS cases, predominantly in young women [1]. The follicular form often involves the ears, chest, legs, and back, in addition to the breast and axilla, occurring mainly in men. This form, as suggested by its name, also shows the highest probability for finding follicular lesions on histopathology among the three HS subtypes. The gluteal subtype presents with follicular papules and folliculitis in the gluteal areas. This HS phenotype is most associated with smoking and low body mass index [1].

An atypical case of HS, both in clinical presentation and demographic, is presented. The patient, a 60-year-old man with history of type II diabetes, presented with lesions on the knees evolving from plaques (with a pink-to-yellowish color) to draining sinus tracts.

Case Synopsis

A 60-year-old man, a non-smoker with a history of poorly controlled type II diabetes presented with a one-and-a-half-year history of lesions on his knees.

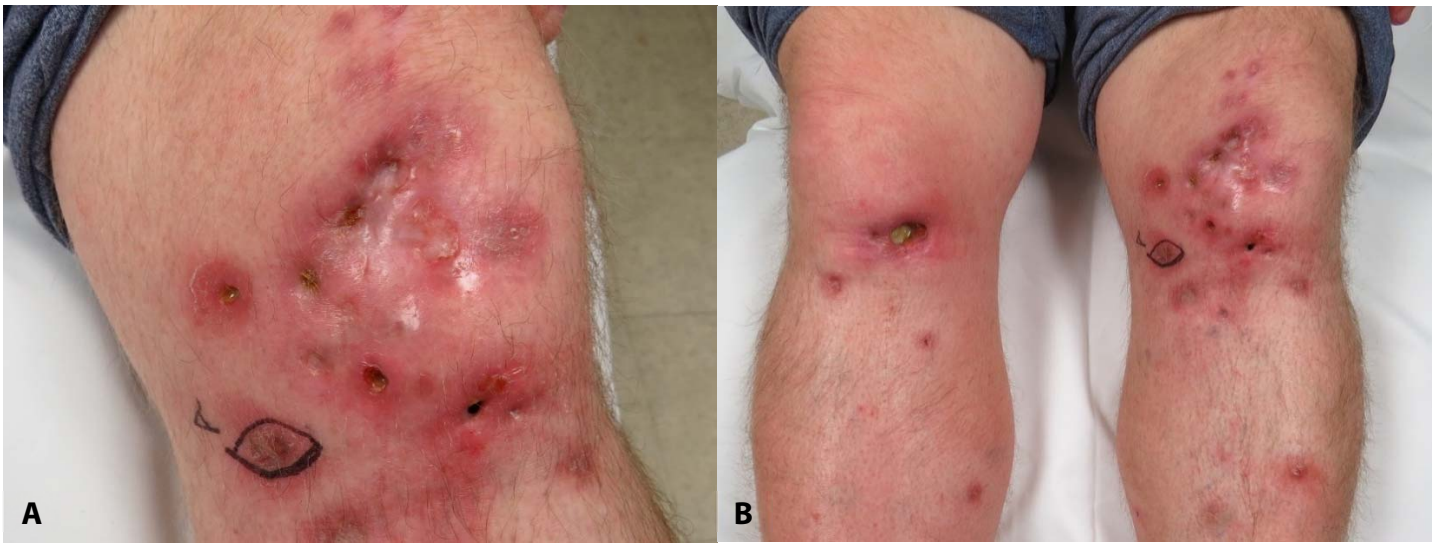

Figure 1. ***A)*** *Scarring and draining sinus tracts shown on the left knee as well as a marked region designated for excisional biopsy.* ***B)*** *Bilateral view of scarring and draining sinus tracts.*

Initially, his lesions were pink-to-yellowish plaques with punched out ulcers. However, over time, the patient's ulcerative plaques evolved into painful draining sinus tracts (**Figure 1**). He had no symptoms or family history to suggest cutaneous Crohn disease and a recent screening colonoscopy was unremarkable. Magnetic resonance imaging was obtained that ruled out osteomyelitis. Various cultures were obtained and were sterile.

Approximately two years after initial presentation, he began to develop painful draining sinus tracts within his left axillae. Large excisional biopsies were obtained from his lower legs. Histopathology demonstrated suppurative granulomatous inflammation centered on hair follicles and sinus tracts (**Figure 2**). The clinical and histopathologic findings supported a diagnosis of HS. Initially, a trial of topical corticosteroids and antibiotics was given, but this treatment failed to improve the patient's condition. Adalimumab was then initiated and the patient showed marked improvement within three months.

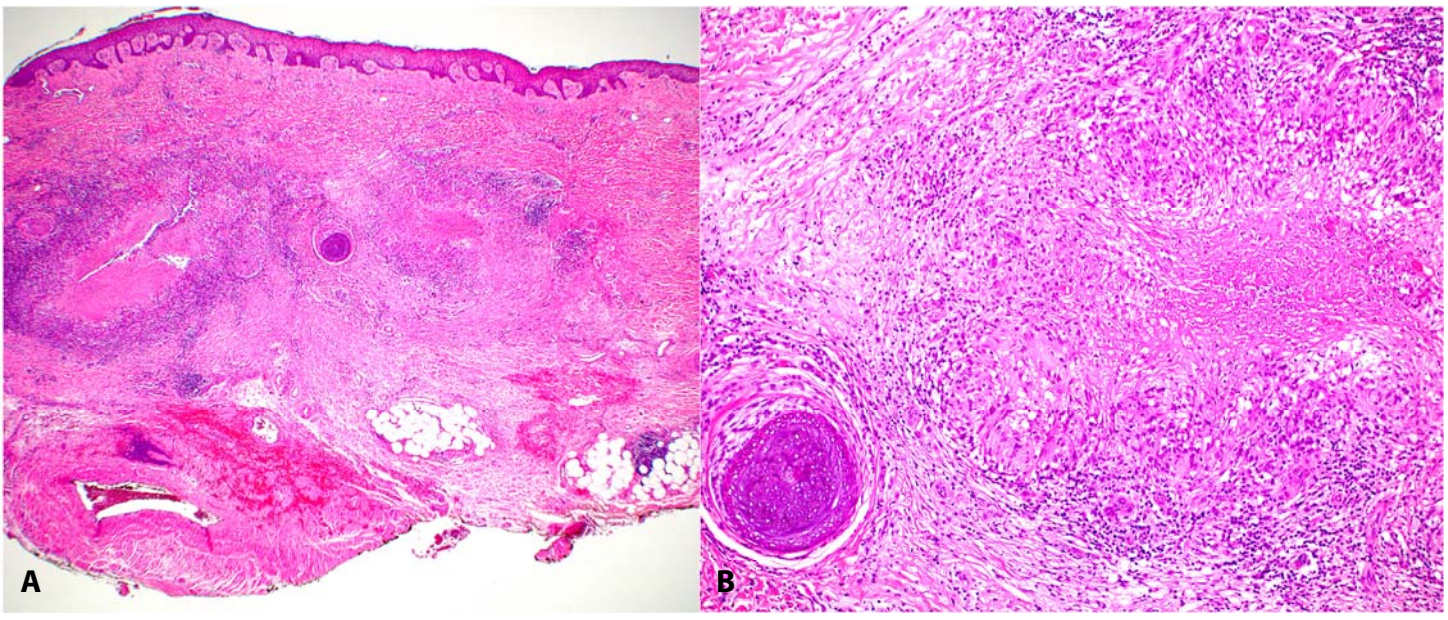

Figure 2. *H&E staining from left knee. Excision of suppurative granulomatous inflammation centered on hair follicles and forming sinus tracts.* ***A)*** *20×,* ***B)*** *100×.*

Case Discussion

A case of atypical HS on the knees of a 60-year-old man is presented. The distribution of his lesions and demographic make this case of HS particularly remarkable, as HS is usually characterized by scars and draining sinus tracts affecting apocrine-gland dense areas such as the axillae, breasts, and groin in young women. There have been several atypical cases of HS present on the chest, eyelids, scalp, thighs, and abdomen [2, 3]. This patient's presentation is most suggestive of the follicular HS given his demographic and lesion distribution on the knees as well as the left axilla.

The differential diagnosis includes cutaneous Crohn disease and cutaneous tuberculosis. Crohn disease can develop as a direct extension from the bowel to the skin and is usually seen in the perianal and orofacial areas. Clinically, the lesions may present as ulcerations, fistulae, fissures, or abscesses [4]. On biopsy, noncaseating granulomatous inflammation is usually found. Cutaneous Crohn disease can also arise in sites noncontiguous with the gastrointestinal tract secondary to "metastases." The most common sites involved with metastatic Crohn disease include intertriginous areas such as the extremities, face, and genitalia. These lesions are described as plaques or nodules with a red to purple hue, possibly having an ulcerative component [5]. Case reports have suggested the use of immunomodulators, such as anti-TNF biologics, or topical or systemic corticosteroids to treat metastatic Crohn disease [5]. The presented patient's clinical findings of pink-to-yellowish plaques with several punched out necrotic areas on physical examination could be seen in metastatic Crohn disease. However, the patient did not have a history of inflammatory bowel disease.

Although metastatic Crohn disease without gastrointestinal involvement has been reported, this diagnosis is unlikely in this particular case.

Cutaneous tuberculosis is a rare manifestation of hematogenous spread or direct extension from latent or active foci of *Mycobacterium tuberculosis* infection [6]. Scrofuloderma, a prevalent type of cutaneous tuberculosis, results from direct extension from an underlying infection, such as from an infected peripheral ganglion, bone, joint, or testicle [7]. The clinical picture of this cutaneous tuberculosis subtype is characterized by "subcutaneous, painless, slowly growing nodules that evolve to ulcers and fistulous tracts with drainage of serous, purulent, or caseous content" [6, 8]. The prognosis of scrofuloderma is insidious, resulting in persistent purulent discharge, chronic ulcers, atrophic sequelae, or spontaneous cure [7]. The presented patient had scars and draining sinus tracts that progressively worsened, which along with his history of diabetes, prompted consideration of cutaneous tuberculosis. However, ancillary studies, including culture and PCR were negative for tuberculosis. A QuantiFERON Gold study was also negative.

The atypical distribution of this patient's lesions, taken with many other published cases detailing HS in apocrine sparse regions, suggests a pathophysiological mechanism that is not reliant on the presence of apocrine glands. The patient's atypical or "ectopic" HS distribution at the knees has been accounted for, in part, by exposure to mechanical stress that could plausibly stimulate interfollicular hyperplasia, a phenomenon that could lead to follicular dilation and rupture, prompting an inflammatory response [9].

The most efficacious treatment for atypical HS also differs substantially from textbook cases. Mild cases of HS and/or cases with typical apocrine distribution often respond to a treatment regimen of antibiotics and/or topical steroids [10, 11]. The featured patient's failed response to antibiotics and topical steroids taken with the histological findings of suppurative granulomatous inflammation centered on hair follicles and sinus tracts further supported a diagnosis of atypical or "ectopic" HS. In moderate-to-severe HS cases and/or cases with atypical distribution, adalimumab along with other tumor necrosis factor inhibitors, has been shown to be significantly more effective than standard HS treatment [10, 11]. This case illustrates this concept well as the patient responded to adalimumab, showing marked improvement in three months.

Conclusion

An unusual presentation of HS in an atypical demographic for the disease is discussed. This case, along with other atypical HS cases published in the literature supports the notion that HS is not just

confined to the axillae, inframammary region, and pelvis. Analyzing diverse presentations can improve the understanding of the spectrum of disease and appropriate treatment options.

Potential conflicts of interest

The authors declare no conflicts of interest.

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