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Plasmacytoma Masquerading as Hidradenitis Suppurativa

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Abstract

Primary plasmacytoma of the skin without evidence of bone marrow plasmacytosis is a rare disorder belonging to the heterogenous spectrum of plasma cell neoplasms and hidradenitis suppurativa is a chronic disease characterized by painful recurrent abscesses, fistulas and scarring lesions in axilla, groin, perineum and rarely mass lesions at the affected site. Here we present a case of plasmacytoma masquerading as hidradenitis suppurativa that presented as a nodular growth in both the axilla in a 45 year old male.

Keywords: Hidradenitis suppurativa; Axillary mass; Plasmacytoma

Introduction

Hidradenitis suppurativa (HS) is an annoying chronic condition, first described by Velpeau [1] and is characterized by swollen, painful, inflamed lesions. The disease occurs in areas of the body that contain sweat glands including the armpits, groin, buttocks, scalp and under the female breast. HS is also known as "Verneuil's disease"2, "Pyoderma fistulans significa"2 and "Acne inversa" [2,3].

It is a common skin condition but is frequently misdiagnosed. HS does not occur before puberty and is widely reported between 20 to 40 years of age.4 More recent studies have indicated that hidradenitis suppurativa is caused by follicular occlusion first, which, in turn occludes the sweat glands and causes perifolliculitis [4,5]. Obesity and cigarette smoking may be triggering factors [4,5]. HS is diagnosed clinically based on its appearance. There are no lab tests or biopsies that establish the diagnosis. In the early stages, it can be misdiagnosed as an isolated boils. We report an unusual case of hidradenitis suppurativa which presented as a mass lesion and diagnosed as plasmacytoma based on the immunohistochemical, histological and urine examination findings.

Case Summary

A forty-five year old male presented in the skin outpatients department with a red, firm to hard, multiple nodular lesions with mild pain under both the arms since 4 months. An open area with spontaneous draining pus was noted. Laboratory tests revealed mild leucocytosis with negative blood culture. Culture of the exudate yielded saprophytic and pathogenic bacteria with a preponderance of staphylococci and streptococci. A provisional clinical diagnosis of hidradenitis suppurativa was made and the patient was subsequently treated with doxycycline (100 mg twice a day) and clindamycin (300 mg twice a day) for 3 months. With no remarkable improvement to therapy, wide local excision was performed.

Excisional biopsy of the nodular lesions in the axilla revealed hyperkeratotic epithelium with focal ulcerations and sheets of benign plasma cells in the dermis (Figure 1 & 2). Urine examination for Bence Jones protein was found to be positive. Immunohistochemically, plasma cells showed kappa and lambda light chain positivity (Figure 3 & 4). Underlying multiple myeloma was excluded by serum protein and immunoglobulin electrophoresis, roentgenographic skeletal survey, and bone marrow biopsy. A diagnosis of plasmacytoma was made based on the immunohistochemical, histological and urine examination findings. The optimal post adjuvant treatment of moderate-dose radiotherapy (40 Gy of Co-60) and cisplatinum based chemotherapy (50 mg x 6 cycles) was given to our patient and he is well after 2 years on follow up.

Discussion

In 1839, the first description of hidradenitis suppurativa was identified and published by Velpeau1. A surgeon from Paris, Velpeau described this unusual inflammatory process with formation of superficial axillary, sub-mammary and perianal abscesses. The problem is somewhat common, thought to occur in 1-2% of the population, but the precise incidence and prevalence are unknown [2].

The incidence of hidradenitis suppurativa is greater in females than in males [6], with female to male ratio of 4-5:1. Flare-ups have been associated with menses, with a higher incidence in females with shorter cycles [6,7]. Axilla is the most commonly involved site in men and women with incidence of 70.0% and 58.0% respectively [8,9].

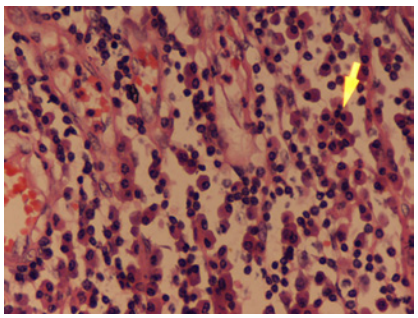

Figure 1: Photomicrograph showing sheets of benign plasma cells in the dermis with eccentrically placed coarse clumped nuclear chromatin and abundant cyanophilic cytoplasm. Haematoxylin and Eosin x 40.

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Post-pubescent individuals are more likely to exhibit HS [7]. A genetic predisposition to hidradenitis suppurativa likely exists, with one study noting that 38% of patients had a relative with hidradenitis [10]. Slade et al. [3] have stated the dermatohistological view of hidradenitis suppurativa as initial hyperkeratosis of the follicular infundibulum with bacterial super-infection, follicle rupture and granulomatous inflammatory reaction of the connective tissue.

Research is assessing possible relations of hidradenitis suppurativa with hashimoto's thyroiditis, crohn's disease, rheumatoid arthritis, and squamous cell carcinoma [11,12]. Squamous cell carcinoma has been found on rare occasions in chronic hidradenitis suppurativa of the anogenital region [13]. The mean time to the onset of this type of lesion is 10 years or more, and the tumors are usually highly aggressive [13].

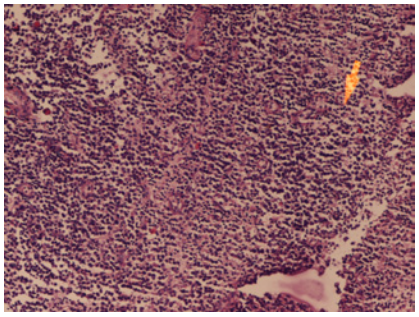

Figure 2: Photomicrograph showing sheets of benign plasma cells in the dermis with eccentrically placed coarse clumped nuclear chromatin and abundant cyanophilic cytoplasm. Haematoxylin and Eosin x 10.

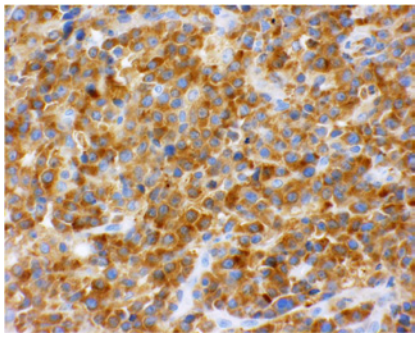

Figure 3: Photomicrograph showing plasma cells with kappa immunostain positivity. Immunostain kappa x40.

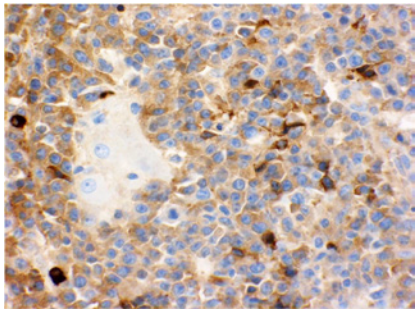

Figure 4: Photomicrograph showing plasma cells with lambda immunostain positivity. Immunostain lambda x40.

Plasmacytoma masquerading as hidradenitis suppurativa has not been documented in earlier literatures, though primary cutaneous plasmacytomas have been reported with an early significant mortality [14]. Primary cutaneous plasmacytoma is defined as monoclonal proliferation of plasma-cells that arises primarily in the skin without evidence of systemic disease. Chest, back and abdomen are the most frequently involved areas followed by face, scalp, neck and extremities [14]. It is an uncommon tumor and is mostly seen in the context of end-stage multiple myeloma. In a number of patients, however, the abnormal clone of plasma cells may arise in the skin and never progress to multiple myeloma involving the bone marrow [15]. Our case of plasmacytoma has been treated vigorously with chemotherapy and local radiotherapy and is doing well after 2 years of treatment.

Conclusion

All cases of chronic hidradenitis suppurativa should be closely supervised, as they are prone to be associated with cutaneous plasmacytomas, which have been reported with an early significant mortality.

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