Hidradenitis suppurativa complicated with secondary lymphedema and lymphangioma of the scrotum

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Abstract

Hidradenitis suppurativa is a chronic relapsing inflammatory skin condition affecting the apocrine sweat glands. Among the complications of this rare condition are local lymphedema and lymphangioma. A case of scrotal lymphedema and lymphangioma following chronic hidradenitis suppurativa in the genital area is described. Excisional skin biopsy was compatible with secondary lymphangioma. A resection of the scrotal mass was performed with reconstruction of the scrotum, penis and perineum.

Key words

Hidradenitis suppurativa, acne inversa.

Introduction

Hidradenitis suppurativa is a chronic relapsing inflammation of the apocrine sweat glands, affecting an estimated 1-4% of the world population. The most common locations are the axillae, groins, buttocks and under the breasts. The disease is caused primarily by follicular occlusion with secondary involvement of the apocrine glands. The main features of hidradenitis suppurativa include painful, recurrent deep seated nodules, papules, pustules, abscesses, scarring, and sinus tracts. The disease may occur as solitary or multiple lesions in one area or in many areas. Hidradenitis suppurativa may be complicated by lymphedema and lymphangioma.1 Lymphangioma (also called lymphangiectasis) is either primary (present at birth or develop in early childhood, or secondary (induced by impairment of fluid flow). Lymphangiomas are found most commonly on proximal extremities, trunk, axillae and oral cavity but also may occur on the penis, scrotum and in the vulva.2,3

Case Report

A 30-year-old male presented to dermatology clinic in 2009 with a one-year history of painful skin lesions with a pus discharge that were recurrent at the beginning and then became persistent in both axillae. He was diagnosed as hidradenitis suppurativa based on clinical and histopathological features. He was then treated with systemic and topical antibiotics on multiple occasions with negligible improvement. Afterwards systemic isotretinoin, adalimumab and surgical intervention led to complete clearance of the axillary lesions in 2011. One
Figure 1 Multiple skin colored subcutaneous nodules with pus discharge on the scrotum and suprapubic area.

Figure 2 (a) Low power H&E slide shows cystic spaces that are lined by flattened cells and filled with lymph; (b) CD31 stain is strongly positive in the lining endothelial cells; (c) CD34 stain is strongly positive in the lining endothelial cells; (d) CK-Pan stain were focally positive.

Year later (2012), he developed similar skin lesions in inguinal regions. Again he was given the same treatment that he received for the axillary lesions. But this time, it could not be effective and with time he developed huge scrotal swelling. The patient was referred to urologist. A massive resection of the scrotal mass was performed and the mass was sent for histopathology. The histological findings were in keeping with lymphangioma.
Discussion

Chronic lymphedema affects patients with severe and long lasting hidradenitis suppurativa. It is due to chronic inflammation that leads to blockade or destruction of local lymph drainage routes. Subsequently lymphangioma develops in long standing lymphedema due to proliferation of lymphatic vessels. In the management of hidradenitis suppurativa complicated by lymphedema and lymphangioma, there is no standard treatment of choice for lymphangiomas. Medical treatment such as frusemide is often ineffective. Ablative therapies, such as sclerotherapy, CO2 laser, liquid nitrogen and electrocoagulation have been described with varying success. Surgical treatment is reported to be curative for hidradenitis suppurativa, lymphedema and lymphangiomas. Wide excision therefore should be considered the most effective treatment option. Two patients with penoscrotal lymphedema were successfully treated with wide radical excision of the lymphedematous tissue and reconstruction with good cosmetic and functional results 8 and 15 years afterwards and no recurrence. In another case report of a 22-year-old patient with penoscrotal lymphedema was subjected to surgical treatment after unsuccessful medical treatment. A total excision of the penile skin and subcutis to Buck's fascia was performed and split-thickness skin grafts were used to cover the defects with excellent cosmetic and functional results.

Conclusion

We believe that lymphangioma is often undiagnosed or misdiagnosed, with many patients experiencing substantial delay until a correct diagnosis is established. These issues compound the high morbidity and disabling nature of the disease. The challenges of timely diagnosis, quality research, community and patient education, and improvement in patients care are all elements to be considered.

References