Dermatological Quiz

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Lesions at the perivulval area

This 45-year-old woman complained of these mildly itchy and painful skin lesions at her vulval and perianal areas for six months. The skin lesions increased in number and size progressively and bled frequently. She was treated as genital warts with topical treatment by a private doctor with no improvement. An incisional biopsy at the vulval lesion showed a vascular proliferation in the dermis with overlying epidermal hyperplasia. The dermal collection of blood vessels was lined by cobblestone-like epithelioid endothelial cells with abundant dense eosinophilic cytoplasm which contained vacuoles, bulging into the lumen of larger sized vessels. Abundant eosinophils are readily found in the dermis.

Questions:

1. What is your diagnosis or possible differential diagnoses?
2. Name one blood test which may help to support your clinical diagnosis?
3. What is the treatment for her skin condition?

Answer to Dermatological Quiz

1. There are multiple small 2 to 3mm erythematous vascular papules over the right labium majora, perivulval and perianal areas asymmetrically. Some lesions showed central erosions and umbilications. The clinical diagnosis may include molluscum contagiosum, haemangioma, pyogenic granuloma and angiokeratoma. The relatively smooth vascular surface of most of the papules make the diagnoses of viral wart, bowenoid papulosis and prurigo nodularis less likely. Coupled with the histological features of haemangiomatous dermal vascular proliferation with the typical cobblestone or hobnail epithelioid endothelial cell and tissue eosinophilia, a rare diagnosis of angiolymphoid hyperplasia with eosinophilia (ALHE) affecting the vulva and perineum can be made.

   ALHE is a rare condition affecting young to middle-aged adults, with a female preponderance. The typical lesion usually presents as red to purplish haemangiomatous dermal or subcutaneous papules, plaques or nodules of 0.1 to 2cm affecting the head and neck, especially in the auricular and peri-auricular areas. The disease less commonly affects the trunk, limbs and mucosal area and only rarely occurs in the genitalia as in this patient.

2. Peripheral eosinophilia in blood is found in about 20% of ALHE patients which might aid the clinical diagnosis of this rare disease.

3. Surgical removal by excision or cryotherapy is usually required although spontaneous regression has rarely been reported. Carbon dioxide laser therapy and electrocauterisation can be useful especially when intraoperative bleeding can be problematic. About 30% of lesions recur after excision. Other treatment options with anecdotal benefits include intralesional steroids, oral isotretinoin, oral pentoxifylline, oral indomethacin farnesil, intralesional interferon alfa-2b, intravenous anti-interleukin-5 antibody. Most of these treatments probably act through mechanisms of anti-angiogenesis, anti-inflammation or inhibition of eosinophils.

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