



Image Article

Teenage Male Patient Presents With Rash Above Eyebrows

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A 14-year-old male patient, accompanied by his mother, presented to his pediatrician with a primary complaint of swelling and discomfort in the knees. Upon examination, the patient's knees were enlarged, and subcutaneous nodularity of the knees was appreciated. The patient had a history of pulmonary valve stenosis, lower leg edema, and a rash above his eyebrows (Figures 1 and 2). The patient was diagnosed with pigmented villonodular synovitis and referred to our clinic for diagnosis and management of the rash. This rash was not painful or itchy and had been present and stable for several years. Biopsy revealed inflammatory keratotic pink papules near the eyebrows, which established a diagnosis of ulerythema ophryogenes. Treatment with topical emollients provided partial resolution; laser treatment was discussed, but the patient declined further therapy.



Figure 1: Hyperkeratotic follicular rash above the eyebrows.



Figure 2: Associated erythema and thinning of the eyebrows.

Ulerythema ophryogenes (UO), or keratosis pilaris atrophicans faciei, is a rare hereditary disease entity with features similar to keratosis pilaris that may occur in conjunction with several disorders including Noonan syndrome (as in our patient), Rubenstein-Taybi syndrome, Cornelia de Lange syndrome, and cardiofaciocutaneous syndrome [1]. It generally presents in childhood, often within a few months of birth, and may progress until puberty. Manifestations include hyperkeratotic follicular papules on erythematous skin around the lateral eyebrows and

cheeks. It may result in alopecia of the eyebrows, cheeks and, more rarely, the forehead. Histopathological examination of the skin should indicate mild chronic inflammation, follicular hyperkeratosis, and skin atrophy [2].

Differential diagnoses include keratosis follicularis spinulosa decalvans, atrophoderma vermiculatum, erythrosis interfollicularis colli, and pseudopelade. First-line treatments include sun protection along with keratolytics such as lactic acid, salicylic acid, and urea. Low potency topical steroids and pulsed dye laser have also been utilized with some success. Hair grafts may be considered for these patients; typically, alopecia and skin atrophy are permanent [1,3].

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