Granulosis rubra nasi: a rare condition treated successfully with topical tacrolimus

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Abstract

A 20 years-old girl presented with multiple asymptomatic reddish vesicles on face for four years. It used to get worse in summer and was associated with localized hyperhidrosis. The lesions were notable for disappearance on diascopy. Histopathology from the vesicle showed mononuclear cell infiltration in the upper dermis, especially around eccrine sweat apparatus, along with dilatation of superficial capillaries and lymphatics. Based on clinical presentation and histopathology, diagnosis of Granulosis rubra nasi (GRN) was made. GRN usually resolves at puberty; however, rarely it may persist in adulthood. We here report a case of GRN having lesions persisting in adulthood. Moreover, she showed excellent response to topical tacrolimus, a finding not observed in literature.

Introduction

Granulosis rubra nasi (GRN) is an inflammatory condition involving eccrine sweat glands of central face and clinically presents as erythema, hyperhidrosis, papules, pustules and vesicles over central face.1 It usually presents in childhood with peak age of presentation 7-12 years.2 It usually presents in childhood between 6 months and 15 years of age.2 Peak incidence is from age 7-12 years.2 Pinkus has described a case in man aged 59 years.5 There is no known racial or sexual predilection.2 It is widely believed that it was first described in 1901 by Jadassohn.1,2

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GRN is believed to be an inherited condition.2,3 Familial cases are known and inheritance is believed to be autosomal dominant.2,3 However, etiology and pathogenesis of this disorder have not been elucidated.2,3 It is believed that persistent localized hyperhidrosis of central face is responsible for this condition.3 At times, hyperhidrosis of palm and sole too has been noted.2,3 Pinkus and Lebet have separately reported an association with hidrocystoma, another eccrine gland disorder.2,3 Summer aggravation of lesions is occasionally found.

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Excessive sweating may precede other changes by several years. It is seen over tip of nose and sometimes, cheeks. With persistent hyperhidrosis, diffuse erythema develops on nose, cheeks, and chin. This erythema may be studded with sweat droplets, giving damp glinting appearance.1,2 After this, erythematous macule, papule, or vesicle may form at sweat duct orifices.2,3 These lesions disappear on diascopy, and reappear on relieving pressure.1 The condition is largely asymptomatic; however, itching or tingling sensation may be appreciated.1 The course of the disease is extremely chronic. It usually resolves at puberty without any sequela. However, in some cases it may persist indefinitely.3

Discussion

Key words: Granulosis rubra nasi , GRN, eccrine sweat glands, face, hyperhidrosis.

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persist in adulthood. Residual telangiectasia and occasional small cysts dominate the clinical picture in such cases.

The common differential diagnoses include miliaria crystallina, hidrocystoma, rosacea, periorificial dermatitis, acne vulgaris and milia. Perioral dermatitis presents with monomorphous small papules and pustules, erythema, and scaling with a distribution primarily around the mouth. Rosacea can be differentiated by accentuation of the erythema by vaso-motor instability, which is not a feature of GRN. Moreover, hyperhidrosis seen in GRN is not a feature of rosacea. Hidrocystoma is characterized by cystic papules of about 1-3 mm in diameter usually appearing in the periorbital area of middle-aged or elderly women. Histopathology can differentiate between these two conditions.

The diagnosis is usually made clinically. Histologically, dilation of dermal blood, and lymphatic vessels with perivascular lymphocytic infiltration and dilation of sweat ducts (at times simulating a hidrocystoma) are seen. The epidermis, connective tissue and pilosebaceous apparatus are otherwise normal and no heterotopic apocrine glands are found.

No preventive measures or complications are reported and the disease has an excellent prognosis with self resolution at puberty in most cases. Hence reassurance is what is needed. Treatment with Botulinum Toxin A is under trial.

Our case presented late with vesicular lesions. At this stage, hyperhidrosis, the characteristic feature of GRN, was absent/ minimal. The authors believe that inflammation around sweat ducts are responsible for decreased sweating and hence, vesicle formation- findings in the late stage of GRN. Owing to scarcity of literature, authors could not ascertain if other authors have had similar observation of decreased sweating in late stages. Further case reports and case series can establish/contradict our observation. Persistence of inflammation around sweat apparatus prompted us to undertake therapy with topical tacrolimus, considering poor response to topical steroids in past. We are not in a position to comment on the response of tacrolimus in earlier stages of disease process, when hyperhidrosis is prominent as we got the patient in a very late stage. We hope this case report will prompt other authors to use and document the response of tacrolimus in various stages of GRN.

References