

Granulosis Rubra Nasi: Dermoscopic Characterization of an Uncommon Entity

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Abstract

Granulosis rubra nasi is a rare disease of the eccrine sweat glands whose etiopathogenesis is still obscure but is known to have been determined genetically with mode of inheritance being autosomal dominant. It presents clinically as increased sweating of mid-facial region with the tip of nose being most affected. Small red macules, papules and vesicles may form at sweat duct orifices. It commonly presents in children and usually resolves when child reaches pubertal age but may remain in adult life. Data on dermoscopy of granulosis rubra nasi is lacking in literature. We herein report a case of a female with adult-onset granulosa rubra nasi along with its dermoscopic features.

Keywords: Granulosis Rubra Nasi; Dermoscopy.

Introduction

Granulosis rubra nasi (GRN), also known as "Acne papulo-rosacea of the nose," is a disorder of the eccrine glands first described by German dermatologist Jadassohn in 1901.¹ This inherited condition is an autosomal dominant familial disease of childhood which involves the eccrine sweat glands of the nose, cheeks and chin. This condition is usually seen in children 7–12 years of age and symptoms disappear when child attains puberty without any sequelae but may remain till adult life.¹ The presenting features of this entity are hyperhidrosis of midface region along with erythema, papules, pustules and vesicles.² It is treated according to symptoms troubling the patient and their associated cosmetic concern. Here we report a case of a female patient with adult onset GRN along with its dermoscopic characterization.

Case Report

A 29-year-old female presented to the dermatology OPD with chief complaints of multiple, asymptomatic papular lesions over the nose associated with occasional sweating for 2 years. On examination, multiple, skin colored to erythematous papules, 2-4mm in size, were seen on the dorsum of the nose [Figure 1]. Rest of the mucocutaneous examination was normal. Hair and Nail examination revealed no abnormalities. Systemic examination was unremarkable.



Figure 1: Multiple, skin colored to erythematous papules, 2-4mm in size seen on the dorsum of the nose.

Dermoscopy of the lesions was done using Hiene Delta 20T dermatoscope, polarized contact mode at 10x magnification. The dermoscopic examination revealed a diffusely erythematous background with discrete oval to round pinkish red structureless areas. Some of the structureless areas showed overlying brown dots [Figure 2].

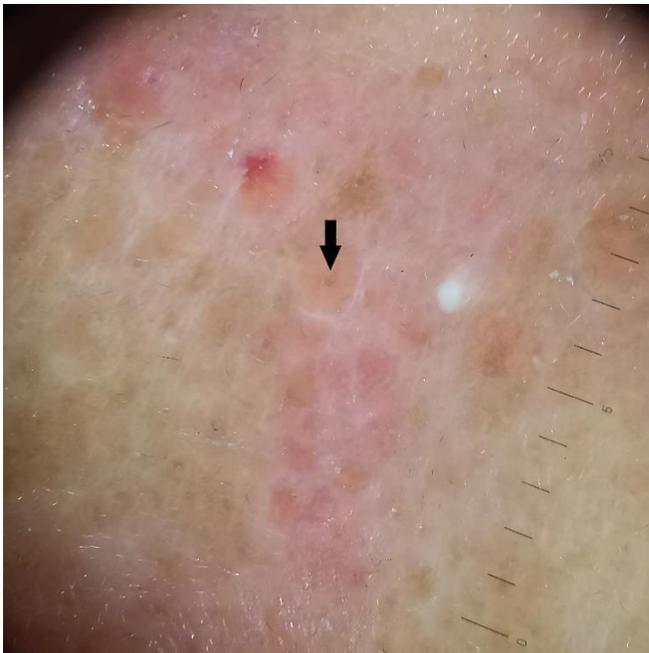


Figure 2: Dermoscopic examination of the lesions showing a diffusely erythematous background with discrete oval to round pinkish red structureless areas. Some of the structureless areas showed overlying brown dots (black arrow) (Hiene Delta 20T dermatoscope, polarized contact mode, 10 ×).

Discussion

GRN is a rare disorder of the eccrine sweat glands which is known to be inherited as autosomal dominant. Etiopathogenesis of this disease is still obscure. This disease usually presents between 6 months of age to 15 years with peak age of presentation being 7 to 12 years.³ Clinical presentation is characterized by excessive sweating of the middle region of the face which is most conspicuous on the tip of nose and later there is development of erythema and papules which disappear when pressure is applied. The tip of nose is generally red or violet, feels cold on touching and not infiltrated.⁴ Excessive sweating precedes development of macules, papules, vesicles, pustules, and telangiectasia by several months or even years. Hyperhidrosis is attributed to cause cystic dilatation of eccrine ducts which then leads to the development of erythema and papules.⁵ Symptoms of GRN usually improves with complete disappearance at puberty on its own but may persist in adult life in which case telangiectasia are a striking feature and small cysts can be seen.⁶

In some cases, affected patients may also have poor peripheral circulation along with excessive sweating of the palms and soles. Symptoms may aggravate in summers.⁷ It may be asymptomatic, mildly pruritic but main concern of patient is cosmetic.

Heid et al. reported a case of granulosa rubra nasi along with diffuse hyperhidrosis in a 19-year-old female who also had an underlying pheochromocytoma and nasal dermatosis regressed after the tumor was surgically removed.⁸

Histology helps in cases which are difficult to diagnose and shows a perivascular and periductal mononuclear cell infiltrate, cystic dilatation of eccrine sweat glands, and vascular proliferation. Histopathology of a papule shows atrophy of epidermis, vacuolar degeneration of basal cells, and nodular collection of lymphocytes along with few histiocytes; and multiple blood vessels with swollen endothelial cells.⁹

Dermoscopic features of GRN have been reported only once in literature till now. Under polarized contact dermoscopy, Palit et al. reported a diffuse erythematous background with discrete round-to-oval pink and red structureless areas, a few of them showing overlying scattered to grouped brown dots.⁹ The diffuse erythematous background seen is probably due to the increased vascularity as seen on histopathology and the discrete round-to-oval pinkish red areas correspond to the dermal nodular inflammatory collections visualized on histopathology.⁹

Possible differential diagnoses include rosacea, acne vulgaris, sarcoidosis and lupus erythematosus. In rosacea, there is transient or persistent erythema of the cheeks, nose and chin with or without telangiectasia and history of flushing with intake of hot liquids but no hyperhidrosis of the central face which is a salient feature of granulosa rubra nasi. Photosensitive dermatoses are usually easy to differentiate from granulosa rubra nasi as they are more extensive with history of photosensitivity but they do not show the prominent feature hyperhidrosis seen in GRN.^{6,10} In acne vulgaris, the skin is usually oily rather than hyperhidrotic and presence of comedones is characteristic feature of acne which is rarely seen in already rare GRN.

Various treatment options include topical indomethacin, tacrolimus, botulinum toxin A,⁵ cryotherapy, X-ray therapy, oral steroids and tetracyclines¹¹ but response to treatment is usually disappointing. Piotr and Katarzyna reported a 14-year-old patient in whom the nasal dermatosis responded well to low-dose isotretinoin¹¹ and Kumar et al. reported topical tacrolimus to be effective in treating the lesions of GRN in a 20-year-old patient.⁷

Patient should be counseled regarding benign nature of disease to alleviate associated anxiety. Cases persisting in adult life are, however, associated with significant psychological impact mainly because of cosmetic concern.

Conclusion

To conclude, GRN is an uncommon nasal dermatosis. Dermoscopy can serve as a useful non-invasive tool to differentiate it from clinically similar looking lesions. Owing to the benign nature of the disease, the patient needs to be counselled well and treatment is indicated mainly for cosmetic purposes.

Disclosure

The authors certify that they have obtained appropriate patient consent. No funding and conflict of interest is declared.

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