Granulosis Rubra Nasi: Case Report

Abstract

A 10 years old Yemeni boy presented to the dermatology department with a 4 years history of localized hyperhidrosis, erythema, and telangiectasias over the tip, dorsum, and alae of the nose. On cutaneous examination erythema, covered with beads of sweat, telangiectasias, multiple 2-4 mm transparent cysts, and few erythematous papules were present on the lateral sides and alae of the nose.

Based on history and clinical symptoms a diagnosis of granulosis rubra nasi was made.

Introduction

Granulosis rubra nasi (GRN) was first described by the German dermatologist Josef Jadassohn in 1901. It is a rare autosomal dominant genodermatosis affecting the eccrine glands of the nose, cheek, and chin. It usually manifests during early childhood, but can manifest in adolescence, and adulthood. It usually starts with localized hyperhidrosis of the tip, dorsum, and alae of nose, years before other signs appeared. GRN usually disappears spontaneously during puberty. Its diagnosis is based on clinical findings, and biopsy is rarely needed.

Case Report

A 10 years old Yemeni boy presented to the dermatology department with a 4 years history of localized hyperhidrosis, erythema, and telangiectasias over the tip, dorsum, and alae of the nose. On cutaneous examination erythema, covered with beads of sweat, telangiectasias, multiple 2-4 mm transparent cysts, and few erythematous papules were present more on the right side, but also on both alae, and dorsum of the nose (Figures 1a-1c).

The patient was otherwise healthy, and all family members were free of this condition. Based on history and clinical symptoms a diagnosis of granulosis rubra nasi was made.

Discussion

Granulosis rubra nasi (GRN) is an inflammatory condition that involves the eccrine sweat glands of central face and manifests as erythema, localized hyperhidrosis, papules, pustules and vesicles over central face [1]. GRN was first described by the German dermatologist Josef Jadassohn in 1901 [2]. It is a rare autosomal dominant genodermatosis that usually manifests during early childhood, in patient aged 6 months to 10 years, but can manifest in adolescence, and adulthood. It usually starts with localized hyperhidrosis - seen as small beads of sweat-of the tip, dorsum, and alae of nose, years before other signs appeared. This localized hyperhidrosis seems to be responsible for the secondary changes like erythema and erythematous papules. GRN usually disappeared spontaneously during puberty, but erythema and telangiectasia may persist. Its diagnosis is based on clinical findings, and biopsy is rarely needed [3,4]. The histology shows dilatation of blood vessels, dilated

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sweat ducts with a discrete mononuclear cell infiltrate surrounding them.

The condition must be differentiated from other similar conditions like, acne, rosacea, perioral dermatosis, milia, miliaria, lupus pernio, and lupus erythematosus. In acne there are comedones, papules and pustules distributed symmetrically over the cheeks, forehead, and chin, but there is no localized hyperhidrosis, and no erythema, or telangiectasias. In rosacea there is erythema of the cheeks and nose along with telangiectasias but no hyperhidrosis, in addition to the elder age of rosacea patients. In the case of periorifical dermatitis there

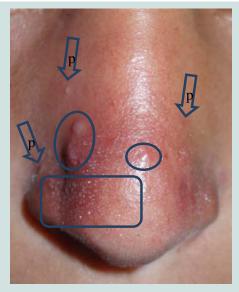


Figure 1a: Frontal view of the nose demonstrating telangiectasias and erythema covered with plenty of droplets of sweat (quadrant) on both sides along with few papules (arrow) and cysts (circle) over the dorsum and right

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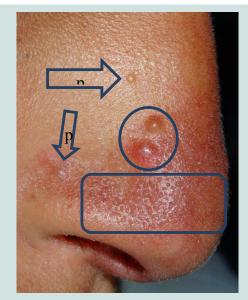


Figure 1b: Right sided lateral view showing telangiectasias, and erythema covered with beads of sweat (quadrant), 2 cysts (circle), and 2 papules (arrows).



Figure 1c: Left sided lateral view, depicting erythema covered by beads of sweat (quadrant).

are small monomorphic papules, pustules, erythema, and scaling involving the perioral and periocular area, leaving a characteristic small free zone around the mouth but no hyperhidrosis. In milia there are longstanding multiple white firm micropapules over both cheeks, but it lacks erythema, and hyperhidrosis. Miliaria crystallina may simulate the beads of sweat but the vesicles cannot be wiped, appear on areas occluded with clothes, and last few days only. Lupus pernio, or chilblain lupus, consists of red or dusky purple papules and plaques not only on the nose, but also on the toes, fingers, and sometimes, elbows, knees and lower legs. The lesions are exacerbated by cold, particularly moist cold climates. Lupus pernio patients have no hyperhidrosis. The bilateral malar erythema "butterfly rash" of

acute cutaneous lupus erythematosus (ACLE) is typical, and cannot be confused with GRN. This tends to be transient, follow sun exposure, and resolve without scarring (but sometimes with dyspigmentation). The condition is associated with anti-dsDNA antibodies, lacks hyperhidrosis, and is not confined to the nose. Treatment of granulosis rubra nasi has been described in the literature with topical indomethacin, oral corticosteroids, tetracycline, cryotherapy, and even X-rays [5]. However, reassurance is more important. Grazziotin et al. have recently reported a long-term remission in a patient with granulosis rubra nasi by using botulinum toxin A [6].

A few years ago Piyush Kumar et al. reported the successful use of tacrolimus cream [7].

To conclude GRN is not rare as it seems, but is usually overlooked. An increased awareness of the dermatologist about this condition will lead to early diagnosis. Treatment is not usually needed and reassurance of the patient is enough, as it is self-limited and disappears during puberty.

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