Erythematous Papules and Pustules on the Nose

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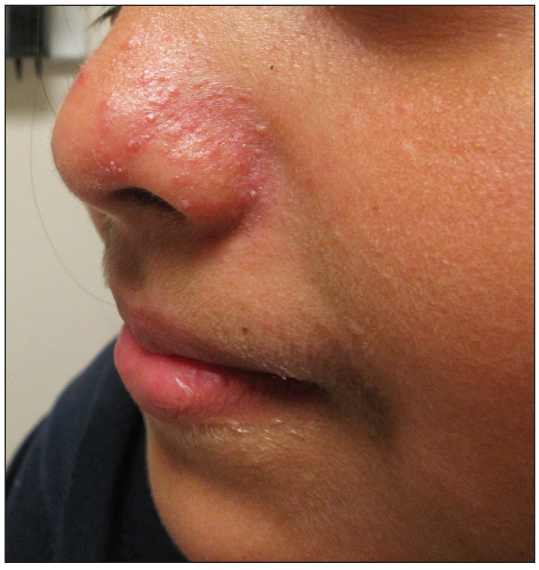

A healthy 9-year-old girl presented with a 2-year history of erythematous papules and pustules on the nose. There was no involvement of the rest of the face or body. At the time of presentation, she had been treated with several topical therapies including steroids, calcineurin inhibitors, antibiotics, and retinoids without improvement. A potassium hydroxide preparation from a pustule was performed and revealed only normal keratinocytes.

WHAT'S THE **DIAGNOSIS?**

a. comedonal acne
b. granulosis rubra nasi
c. nevus comedonicus
d. periorificial dermatitis
e. tinea faciei

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The authors report no conflict of interest.
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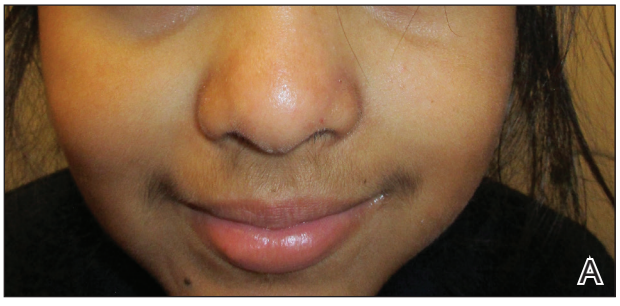
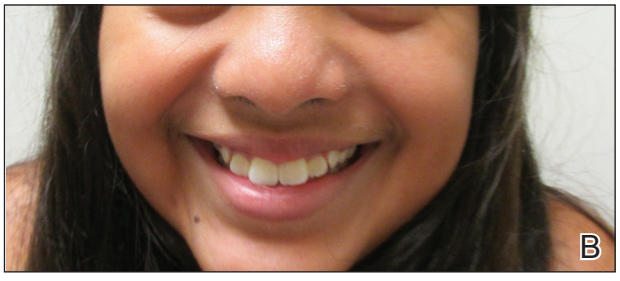
THE **DIAGNOSIS:** Granulosis Rubra Nasi

A history of prominent nasal sweating was later elicited and the patient was subsequently diagnosed with granulosis rubra nasi. She was instructed to continue daily use of topical pimecrolimus with the addition of topical atropine, resulting in complete resolution of the eruption at 6-week follow-up (Figure, A). She was then maintained on topical atropine monotherapy, only noting recurrence with cessation of the atropine (Figure, B).

Other successful treatment regimens of granulosis rubra nasi include injection of botulinum toxin into the nose,[1] monotherapy with topical tacrolimus,[2] topical indomethacin, steroids, and cryotherapy, among other modalities.[1] Topical atropine and pimecrolimus were selected as first-line agents for treating our pediatric patient due to tolerability and their anti-inflammatory and anticholinergic properties.





A, Complete clearance of granulosis rubra nasi at 6-week follow-up after using topical pimecrolimus and atropine once daily. B, Complete clearance 2 months later after using topical atropine monotherapy.

Granulosis rubra nasi is a form of focal hyperhidrosis that presents as erythematous papules, pustules, and vesicles of the midface, especially the nose.[3] It is a fairly rare condition that can mimic many other common clinical entities, including comedonal acne, nevus comedonicus, periorificial dermatitis, and tinea faciei, but is resistant to treatments aimed at these disorders. It was first described as a "peculiar disease of the skin of the nose in children" in a case report by Jadassohn[4] in 1901. It is most common in children aged 7 to 12 years and typically resolves at puberty; adults rarely are affected. Although the etiology has not yet been elucidated, autosomal-dominant transmission has been described, and the cutaneous changes are hypothesized to be secondary to hyperhidrosis.[5] This postulation is further corroborated by a case report of a pheochromocytoma-associated granulosis rubra nasi that resolved with surgical excision of the pheochromocytoma.[6] It is not uncommon for patients to have concomitant palmoplantar hyperhidrosis and acrocyanosis.[5] Histopathologic examination is not necessary for diagnosis, but when performed, it discloses a mononuclear cellular infiltrate surrounding eccrine sweat ducts, blood vessels, and lymphatics without other abnormalities of the epidermis or pilosebaceous unit.[1-3,7]

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