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A CASE REPORT ON GRANULOSIS RUBRA NASI

Dermatology

Dr. R. Sindhya* Junior Resident, Department of Dermatology, Venereology & leprosy, Chettinad Hospital And Research Institute, Kelambakkam, Chennai, Tamil Nadu. *Corresponding Author

Dr. Udaya Nagapurapu Junior Resident, Department of Dermatology, Venereology & leprosy, Chettinad Hospital And Research Institute, Kelambakkam, Chennai, Tamil Nadu.

Dr. Jayakar Thomas Head of Department, Department Of Dermatology, Venereology & leprosy, Chettinad Hospital And Research Institute, Kelambakkam, Chennai, Tamil Nadu.

ABSTRACT

Granulosis Rubra Nasi is an uncommon eccrine gland disorder with an autosomal dominant inheritance. It usually occurs over the face particularly on the tip of the nose and typically is thought to be initiated due to hyperhidrosis which then eventually manifests as diffuse erythema over the nose, chin, cheeks, and upper lip. Although frequently seen in childhood, it occasionally occurs in adults as well. Here we present a case report on a 22year old female patient who presented with complaints of uncontrolled sweating over the nose. On detailed examination of the nose, erythema and telangiectasias were noted. Biopsy findings supported the diagnosis of granulosis rubra nasi as well. This case has been reported due to its rare occurrence.

KEYWORDS

eccrine glands, granulosis rubra nasi, nose, hyperhidrosis, telangiectasia

INTRODUCTION

Granulosis rubra nasi was first described by Jadassohn in 1901 which is uncommon disorder of the eccrine glands. It is found to be common among the European population. It is also called as "Acne papulo-rosacea of the nose." [1] The disease usually appears in the childhood but adolescent or adult-onset cases are also reported.[3] This disease mostly resolves at the time of puberty but still persistent cases are also seen. The pathogenesis of GRN remains obscure. It is likely to represent a unique form of sweat retention with the occurrence of erythema, papules, and vesicles as secondary changes. Familial cases have been reported, and both autosomal dominant and recessive modes of inheritance have been suggested.

Case Report

A 22-year-old female presented with multiple reddish lesions over the nose and cheeks along with mild itching for 10 years. The lesions initially started with occurrence of excessive sweating over the tip of nose that extended to involve the central face, followed by the gradual appearance of diffuse redness over the nose and cheeks. There was no history suggestive of excessive sweating of palms and soles, photosensitivity, or poor peripheral circulation. Family history was not significant.

Physical examination showed hyperhidrosis and erythema of the nose covered by beads of sweat [Figure-1], and multiple telangiectatic vesicles over the Centro facial region along with multiple 2-4-mm-sized erythematous papules and vesicles over the dorsum, tip and alae of nose, adjoining areas of the cheeks, glabella, and philtrum [Figure - 2]. On performing a diascopy test on nose blanching effect was seen [Figure-3]. Other physical examination was normal. Skin biopsy showed epidermal thinning and vascular dilatation of upper and mid-dermal blood vessels and perivascular lymphohistiocytic infiltrate. The patient was counselled about the benign and self-limiting nature of the disease.

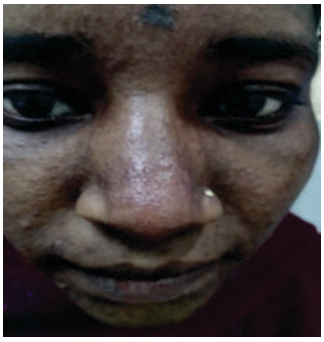

Figure-1: hyperhidrosis on an erythematous base over the nose

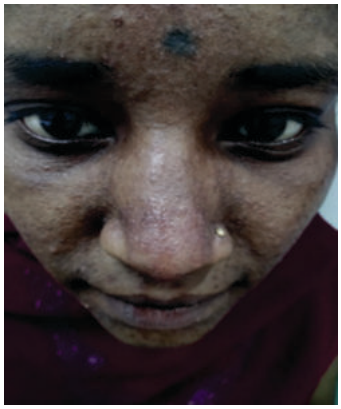

Figure-2: erythematous papules over the dorsum, tip and alae of nose, adjoining areas of the cheeks and glabella

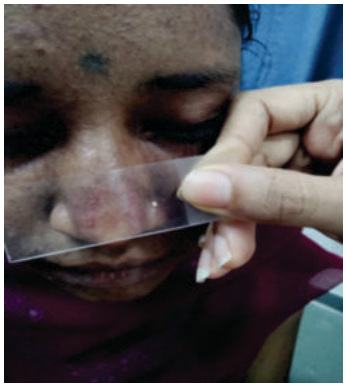

Figure-3: on performing a diascopy test blanching was seen over the nose which suggests of erythema

DISCUSSION

Our patient presented with granulosis rubra nasi which is an uncommon disorder of eccrine glands with autosomnal inheritance. Synonyms for granulosis rubra nasi are: perisyringitis chronica nasi ; dermatitis micropapulosa or granularis erythematosa hyperidrotica chronica nasi (infantum); false acne rosacea in children.Its onset is generally in childhood aged betwwen 6 months to 15 years although adult onset cases have been reported.The disease usually presents with hyperhydrosis of the central part of the face,commonly over the tip of the nose following which diffuse erythema over the cheeks,nose,upper

lip and chin.They may also present as erythematous macules,papules,vesicles or even pustules which is seen over the sweat duct orifices.[4]Telangiectasias can be seen over the areas like nose,cheeks and chin.[1,5] Comedo-like lesions may also be present.[1] The nose may be palpably cold and cystic lesions can develop.[3] It may sometimes be associated with hyperhidrosis of the palms and soles and acrocyanosis. The diagnosis is mostly clinical. Histopathology shows dilation of blood vessels and the lymphatic vessels with mild mononuclear cell infiltrate around the sweat ducts, blood vessels, and lymphatics. Pilosebaceous units are normal and no heterotopic apocrine glands are found[3] This disease usually remits after puberty, unlike the primary forms of localized hyperhidrosis but sometimes may continue into adulthood.[3,4] There is no systemic associations or abnormal laboratory findings.[3]

The pathogenesis of the disease is still unknown. Some authors have suggested that it is a disorder of vasomotor and secretory function of the nose. Goldsmith had described rhinorrhea as an associated feature.[6] Eddowes had suggested that adenoids could be involved which can provide a source of irritation to the tip of the nose.[7] Heid et al. reported a case of granulosis rubra nasi associated with phaeochromocytoma, surgical removal of which was followed by regression of the condition.[8]

The most common differential diagnosis includes rosacea ,hidrocystoma and perioral dermatitis. In rosacea there is erythema of the cheeks and nose along with telangiectasias but no hyperhidrosis of the central part of the face. 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. In the case of perioral dermatitis there are small monomorphic papules, pustules, erythema, and scaling involving the perioral area and no hyperhidrosis. Other differential diagnosis include acne vulgaris, lupus pernio, and lupus erythematosus.[3] miliaria crystallina, and milia. Treatment of granulosis rubra nasi has been described in the literature with topical indomethacin, oral corticosteroids, tetracycline, cryotherapy, [2] However, reassurance is more important. Drying lotions like calamine can be tried.[3] Grazziotin et al. have recently described a treatment using botulinum toxin A that induced long-term remission in a patient with granulosis rubra nasi.[9].Other options like atropine 1% cream and tacroclimus 0.03% topically have been used and are under trial.

CONCLUSION

Our case presented with granulosa rubra nasi with papular lesions and hyperhidrosis. An increased awareness of the dermatologist about this condition will lead to early diagnosis. Treatment is symptomatic and cosmetic. Counselling the patients about the self-limiting nature of the condition is of paramount importance. To the best of our knowledge, there are not many case reports of GRN reported among the indian population.

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