

Rhinophyma and Gnathophyma Concomitantly in a 54-Year-Old Female a Clinical-Histopathology Correlation and Review of the Literature

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Abstract

Rhinophyma (Greek “nose growth”) is a benign dermatologic disease of the nose [1]. It is characterized by a slowly progressive enlargement of the nasal skin, with irregular thickening and nodular deformation leading to an erythematous appearing nose. Phyma is the last stage of rosacea and is due to chronic inflammation and edema. Although the great majority of phymatous lesions occur on the nose (rhinophyma), they may also occur at other sites. It affects mostly men. Even though the exact cause of rhinophyma is unknown, it is believed to be multifactorial in origin with a principal etiology of unregulated superficial vasodilation.

The main reasons that urge the patients to seek help are functional and cosmetic impairments, such as nasal obstruction or disfigurement. Surgical removal of the hyperplastic tumor mass is the treatment of choice for rhinophyma.

The aim of the article is to present an update on the pathophysiology, clinical features and treatment of rhinophyma and gnathophyma in a 54-year-old female patient.

Keywords: Rhinophyma, Gnathophyma, Sebaceous Hyperplasia, Nasal Tumor, Rosacea

Introduction

Rhinophyma is a less-common subtype of rosacea that presents as thickened skin with enlarged sebaceous glands that may progress to large bulbous growths with tumoral aspect over the nose. It can also cause upper airways obstruction and difficulty in eating. It can affect nose (rhinophyma), chin (gnathophyma), forehead (metophyma), ears (otophyma) and eyelids (blepharophyma). The tumour-like phymas progressively enlarge in size causing deformity. It affects mostly men and is characterized by fibrosis, sebaceous glands hyperplasia and lymphedema.

Historically, rhinophyma was erroneously considered to be linked with alcohol consumption because substances such as alcohol and caffeine can cause local vasodilation, which worsens the symptoms [2]. This pretended association with alcohol has caused social stigma and loss of self-esteem in patients suffering from the disease, with several nicknames for the condition such as “whiskey nose” and “rum nose” [3, 4]. However alcoholism does not cause rhinophyma.

Although the exact pathogenesis of rosacea and rhinophyma is not known, it is thought to be a combination of multiple factors leading to vascular changes and a trigger of the innate immune system. Numerous vascular growth factors and receptors have been shown to be increased in affected skin leading to an overall state of abnormal

vascular reactivity. The extravasation leads to chronic edema of the dermal interstitial with a sequel of local inflammation, fibrosis, and dermal and sebaceous gland hyperplasia [2-4]. Over time, this leads to the characteristic bright red to purplish telangiectasia’s and irregular, lobulated thickening of the skin of the nose [5-10]. Other risk factors include: UV light exposure, the excess of steroid hormones (androgens), trauma, and the presence of a parasite (Demodex mites), and vitamin deficiency [11-15]. Additionally, smokers have been found to have a higher risk of developing rosacea [16-18].

The diagnosis of rhinophyma is clinical and can be identified by the nose’s bulbous shape, skin pitting/scarring, and telangiectasias. Rhinophyma can be complicated by unnoticed cutaneous malignancies. Occult basal cell carcinoma is estimated to occur in 3% to 10% of rhinophyma cases, while other types of skin cancers and systemic malignancies have been found to mimic rhinophyma [19-21].

Case presentation

A 54-year-old female with no significant past medical history, was presented to the hospital because of a 1-year history of growing masses, located on the nasal alae and the chin area. The patient reported they started as small papules and spread bilaterally over the alae and the chin, over the course of a year. She noticed most of the growth during the last 5 months up to presentation.



Figure 1

Physical examination revealed an enlarged nose by several skin-colored nodules of different sizes with overlying prominent dilated pores encompassing the entire surface of the nasal alae. Also 2 smaller nodules with the same features as shown above in the chin area (Figure 1).

On examination of the remainder of her face, her bilateral cheeks and nose showed sebaceous skin with multiple scattered dilated pores and a few small telangiectasias. There were no facial pustules. There was not any palpable lymphadenopathy. The remainder of her skin on her body was normal.

The patient underwent electrosurgical excision of the lesions.

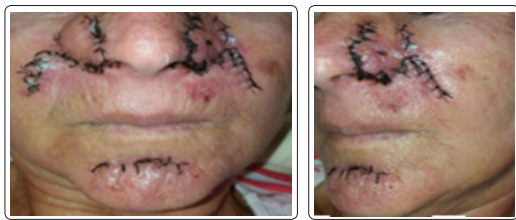


Figure 2 **Figure 3**

Figure 2, 3: The results after removing the lesions. The aesthetic result after surgical intervention was very good; there were no postoperative complications or recurrences. The site healed successfully with secondary intention and a restored normal nasal alar contour.

Microscopic examination on H&E stain

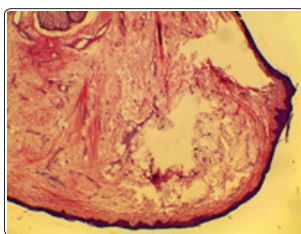


Figure 4: H&E stain reveals papillomatous growth, lined by squamous epithelium accompanied by dermal edema.

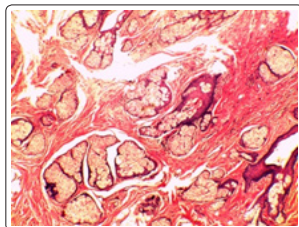


Figure 5: Low magnification of the sebaceous glands hyperplasia.

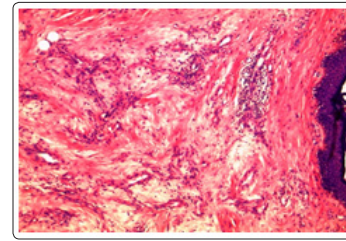


Figure 6: Low magnification revealing angiogenesis, telangiectasia and perivascular inflammatory infiltrates.

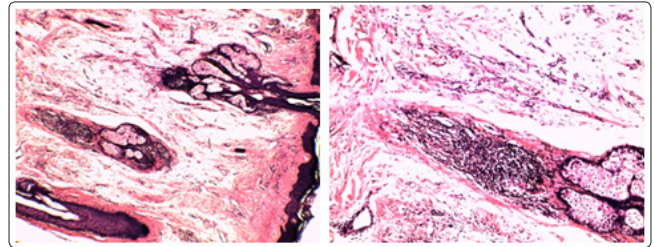


Figure 7, 8: Perivascular and peripilar inflammatory infiltrates (Lymphocytes and few plasma cells), involving the superficial and mid-dermis.

Discussion

Rosacea is a difficult entity to classify, not only because its pathogenesis is poorly understood, but also because of the broad spectrum of histopathological changes found. They can differ considerably from case to case, reflecting the clinical aspect on presentation. Four variants of rhinophyma can be recognized: glandular, fibrous, fibroangiomatic, and actinic. In glandular form, the nose is enlarged mainly because of sebaceous gland hyperplasia. In the fibrous form diffuse hyperplasia of the connective exists. In the fibroangiomatic form fibrosis, telangiectasia's, and inflammatory lesions are present. In the actinic form, nodular masses of elastic tissue distort the nose. The differential diagnosis is made with: squamous cell carcinoma, basocellular carcinoma, adnexal tumors, and skin metastasis especially in pulmonary neoplasms, eosinophilia facial granuloma, hemangioma and nasal keloid scar. Granulomatous processes such as sarcoidosis and infectious diseases such as rhinoscleroma (Klebsiella) or leishmania should also be considered in the appropriate clinical setting.

Multiple treatment modalities exist for rhinophyma depending on the size of the lesion and experience of the surgeon. Surgical management is divided into 3 approaches: full-thickness excision followed by split-thickness skin graft, full-thickness excision followed by full-thickness skin graft, and decortication or partial excision. Use of electro surgery to excise the rhinophyma can reduce bleeding [19]. Preservation of the sebaceous glands allows for adequate re-epithelialization, and the excision should not extend deeply enough to expose cartilage [19]. Skin grafts or local flaps can be used to cover the defect, or the wound can be allowed to re-epithelialize spontaneously within 2 weeks [22-26].

Conclusion

Rhinophyma is a rare, disfiguring disease characterized by a progressive hypertrophy of the soft-tissues of the nose associated to an end-stage of severe acne rosacea. It can lead to serious cosmetic deformities, functional impairments following with psychosocial consequences and the possibility of developing occult malignancy.

Although topical antibiotics or retinoid are effective medical treatment options for rosacea, they have not been shown to improve rhinophyma. Surgical correction appears to be the treatment of choice for optimal treatment of rhinophyma, with multiple modalities available at the discretion of the surgeon.

Even though the diagnosis of rhinophyma and gnathophyma is made clinically, the histopathological examination is very useful for confirming the diagnosis, establishing the progression of the lesions also excluding other underlying malignancies.

Acknowledgement

Our patient has healed well after electrosurgical treatment and is pleased with her improvement.

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