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Case Report

A rare case of giant rhinophyma

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Abstract Rhinophyma is a rare skin disease characterized by a disfiguring enlargement of the nose due to proliferation of sebaceous glands and surrounding soft tissue. It is generally considered to be a final stage of untreated rosacea. This report describes a 79-year-old man manifested with a giant rhinophyma measuring 7cm in diameter. It grew for many years but the patient had not sought any dermatologic intervention. The lesion was completely removed by using sharp scalpel incision. Although rhinophyma is not a life-threatening condition, it can sometimes reach a huge size, with a negative effect on quality of life. Such cases are usually a consequence of patient's delay in seeking medical advice. In more pronounced lesions, a simple surgical removal under tumescent anestesia is an optimal treatment modality.

Key words
Rhinophyma, rhinophyma severity index, rosacea.

Introduction

Rosacea is a frequent skin disease manifesting with erythema, papules, pustules, and teleangiectasias localized in the central face.[1,2] It usually begins with transitory flushing as the initial sign and if untreated, may terminate in the phymas (*phyma*, Greek=swelling, bulb).[2] Fortunately, a progression of disease is often incomplete, a number of intermediate stages occur and thus, many individuals who develop a facial erythema never develop phyma. Phymas may occur in various areas of the face, but nasal involvement (rhinophyma) is the most common.[2] Rhinophyma (potato or whisky nose) is characterized by a disfiguring enlargement of the nose due to proliferation of sebaceous glands and underlying connective tissue.[2,3] It typically affects Caucasian males between their 5th and 7th decades while it is very rare among African and Asian populations[2]. Histopathology of rhinophyma is nonspecific and similar to those found in the various subtypes of rosacea.[4] There is a broad spectrum of clinical appearances as well. In 2013, a team of German dermatologists[5] proposed the Rhinophyma Severity Index (RHISI). It numerically scales the disease based on degree of skin thickening, presence of lobules and fissures, and evidence of marked asymmetry, cysts, or dilated vessels **(Table 1)**. Giant rhinophyma represents the rarest and most serious form of this disease. Herein, I describe one such case from my own dermatopathologic practice.

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Case report

A 79-year-old man presented with a painless bulbous growth arising on the lower one-third of the nose. The patient claimed that the lesion had been manifested for many years and had gradually increased to the current size. His medical history did not reveal an alcohol abuse. He previously worked as a bus driver and he often attracted the attention of passengers due to the appearance of his face. Nevertheless, he had not sought any dermatologic intervention until

Table 1 Rhinophyma Severity Index (RHISI).

Score	*Description*
0	no evidence of rhinophyma
1	mild skin thickening
2	moderate skin thickening
3	strong skin thickening, small lobules
4	lobules with fissures
6	giant rhinophyma
maximum one extra point	presence of strong asymetry, multiple cysts or strong vessels
maximum score: 6 points	

Figure 1 Biopsy sample after surgical removal.

Figure 2 Section through resected tissue mass.

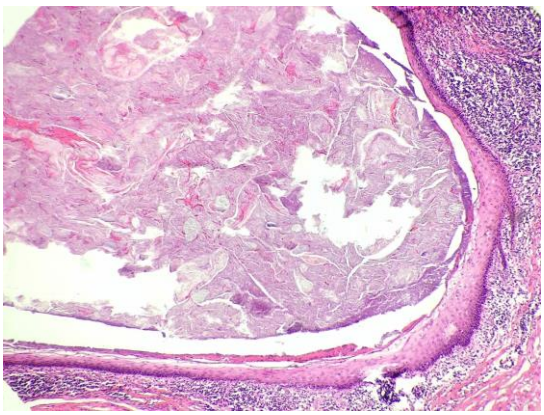

Figure 3 Follicular cyst filled with keratin debris. (hematoxylin & eosin, 20x).

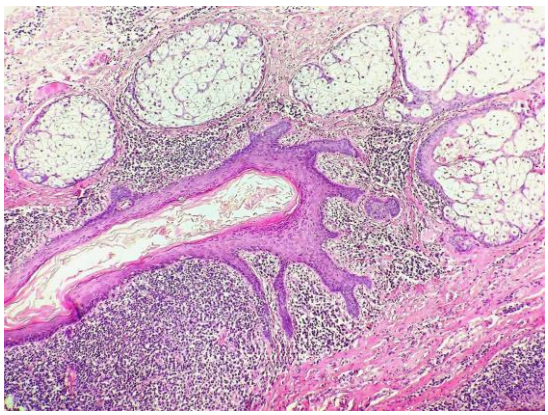

Figure 4 Enlarged hair follicle with massive perifollicular chronic inflammation. (hematoxylin & eosin, 20x).

that time. Grossly, there was a huge, pedunculated, cauliflower-like mass measuring 7cm in largest diameter, occupying the tip, dorsum and both alae of the nose. It partially obstructed the nostrils, causing breathing difficulties. The clinical appearance led to a diagnosis of a giant rhinophyma comprising 6 points according to the RHISI. The lesion was completely removed by using sharp scalpel incision without damaging alar cartilages.

The resected biopsy samples after fixation in formalin are illustrated in **Figures 1** and **2**. Histology revealed an edematous fibrotic stroma (with numerous Factor XIIIa-positive fibroblasts) containing ectatic vessels and multiple enlarged/cystic follicular infundibula. The cysts were filled with keratin debris **(Figure 3)** admixed with basophilic colonies of yeast-like microorganisms. Fragments of *Demodex*

folliculorum mites were not observed. Some infundibular cysts were packed with pus, the equivalent of a pustule. There was an intense perifollicular lymphocytic infiltrate (predominantly CD4+ helper T-cells) with plasma cells and disperse mastocytes **(Figure 4)**. Hyperplasia of sebaceous glands was only mild and focal. Spectrum of histopathologic findings was consistent with presumptive diagnosis of rhinophyma. The postoperative course was uneventful and the wound healed in with satisfactory cosmetic and functional results. Further clinical course can not be assessed, because this paper was written shortly after operation.

Discussion

Rhinophyma is a slowly progressive disorder of unknown etiology.[2,3] It is considered to be multifactorial with a primary etiology of dysregulated superficial vasodilatation.[3] The prolonged extravasation causes chronic interstitial edema with a sequela of local inflammation and connective tissue hypertrophy.[3] It has been assumed that in advanced forms of rhinophyma, chronic inflammation leads to fibroplasia due to overexpression of Factor XIIIa in dermal fibroblasts and dendritic cells.[4] In the past, rhinophyma was incorrectly linked with alcoholic beverages abuse because ethanol can cause local vasodilation, which typically worsens the symptoms of the disease. This alleged association, which has often led to social stigma, has already been declined.[2,3] Although rhinophyma is generally considered to be a final stage of rosacea,[2,3] only a few rosacea patients develop this condition. In an earlier study of 108 individuals with rosacea,[6] only 14% were suffering from rhinophyma. However, rhinophyma may result in marked enlargement and deformity of the nose, with a negative effect on quality of life. Such cases are usually a consequence of patient's delay in seeking medical advice. The treatment strategy of the disease can be very challenging but in larger lesions, the primary mode of therapy is a total surgical extirpation of phytomatous tissue with preservation of the nasal estetic subunits.[2] Resected tissue should always be microscopically assessed to rule out a malignant neoplasia, especially in older persons. Some malignancies may be occult within existing rhinophyma or may mimic it clinically. For example, Iranian authors[7] described an unusual case of sebaceous carcinoma arising from nasolabial region in an 81-year-old man that grossly resembled a rosaceous rhinophyma. From a prognostic point of view, no histopathologic feature has been found to predict the clinical course of this disease after surgery. In a study of Schüürmann et al,[4] a positive association was only identified between the pre-operative RHISI score and the recurrence rate, with a high score being a risk determinant for relapse.

Conclusion

Although rhinophyma is not a life-threatening disease, it can sometimes attain a huge size, cause significant psychosocial morbidity and pose a difficult management challenge. In more pronounced lesions, a simple surgical removal under tumescent anestesia is considered as an optimal treatment modality. These patients require a careful follow-up due to a high risk of local recurrence.

Consent. The examination of the patient was conducted according to the Declaration of Helsinki principles.

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