Gnatophyma - A rare form of rosacea *

Gnatofima - Uma forma rara de rosácea

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Abstract: Phyma is the last stage of rosacea and is due to chronic inflammation and edema. It can affect nose (rhinophyma), chin (gnatophyma), forehead (metophyma), ears (otophyma) and eyelids (blepharophyma). Rhinophyma is the most frequent location and there are few reports about gnatophyma. We report the case of a female patient, 41 years old, who had an infiltrated, erythematous, edematous plaque around the chin and lower lip for two years. Histopathology showed perivascular lymphocytic infiltrate, hypertrophied follicles and sebaceous glands, dilated vessels and fibrosis. She was treated with oral tetracycline, oral ivermectin and metronidazole cream with a satisfactory response. The clinical, histopathological and therapeutic response correlation confirmed the diagnosis of gnatophyma, a rare variant of phyma.
Keywords: Chin; Ectoparasitic infestations; Female; Parasitology; Rosaceae

Resumo: Fima é o estágio final da rosácea e ocorre devido ao edema e inflamação crônica. Pode acometer nariz (rinofima), mento (gnatofima), fronte (metofima), orelhas (otofima) e pálpebras (blefarofima). Rinofima é a localização mais encontrada e há raros relatos de gnatofima. Relataremos paciente feminina, 41 anos, que apresentava placa infiltrada, eritêmato-edematosa, em todo o mento e lábio inferior há dois anos. Histopatológico com infiltrado linfocitário perianexial e perivascular, folículos e glândulas sebáceas hipertrofiadas, vasos ectasiados e fibrose perianexial. Foi instituído tratamento com tetraciclina via oral, ivermectina via oral e metronidazol creme com resposta satisfatória. Através da correlação clínica, histopatológica e resposta terapêutica confirmou-se o diagnóstico da variante rara de fima, gnatofima.
Palavras-chave: Ectoparasitoses; Feminino; Parasitologia; Queixo; Rosácea

INTRODUCTION

Rosacea is a chronic skin condition, characterized by redness, telangiectasia, papules, pustules, skin thickening and phymas.[1,2] The etiology is unknown.[1] It is believed that tissue damage, oxidative stress, decreased superoxide dismutase, and production of vasoactive substances such as serotonin, prostaglandins and substance P are involved in its pathogenesis, as well as opioid peptides and Helicobacter pylori infections, Demodexbrevis and Demodex folliculorum.[1,2] Phyma is considered the last evolution stage of rosacea. [1-6] It affects mostly men and is characterized by fibrosis, sebaceous glands hyperplasia and lymphedema.[3] It most frequently affects the nose (rhinophyma), and rarely the chin (gnatophyma), forehead (metophyma), ears (otophyma) and eyelids (blepharophyma).[2,3] This report is about gnatophyma, a rare form of phyma with both clinical and histopathological characteristics and a satisfactory response to treatment, but which is difficult to diagnose because of the rarity of its occurrence.

CASE REPORT

A 41-year old female patient presented growth and redness of the chin two years ago. Dermatological examination showed well-defined infiltrated, erythematous, edematous, asymptomatic plaque on the chin and lower lip. The skin was thickened on the chin and the rest of the face was unaltered (Figures 1 and 2). The patient reported periods of exacerbation but was unable to relate it to psychological factors, stress and hot food or alcohol. She had been taking captopril, hydrochlorothiazide, metformin and fluoxetine for five years. The initial diagnosis was contact dermatitis, so she was instructed to stop wearing makeup, creams, nickel jewelry and nail polish, associated with oral corticosteroids for 30 days, but no improvement was

Received on 01.06.2011.
Approved by the Advisory Board and accepted for publication on 28.09.2011.
* Study carried out at the University of Santo Amaro (Universidade de Santo Amaro - UNISA) – São Paulo (SP), Brazil.
Conflict of interest: None
Financial funding: None

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noted. A skin biopsy of the chin showed corneal plugs, dense lymphocytic perifollicle, periadnexal and perivascular infiltrate in the dermis, sebaceous glands hyperplasia, hypertrophied hair follicles, dilated vessels and outline of giant cells (Figures 3 and 4). This histopathological result suggested Syndrome of Mekelsson-Rosenthal, thus, in addition to oral corticosteroids, we introduced intralesional steroids as part of the treatment, but there was no improvement. Another possible diagnosis was rosacea, in its rare variant, gnatophyma. The treatment prescribed was tetracycline 1 g/day, ivermectin 12 mg single dose and metronidazole cream 1 %, with improvement of the clinical status (Figure 5). Through clinical and histopathological correlation and a satisfactory therapeutic response the diagnosis of gnatophyma was confirmed.

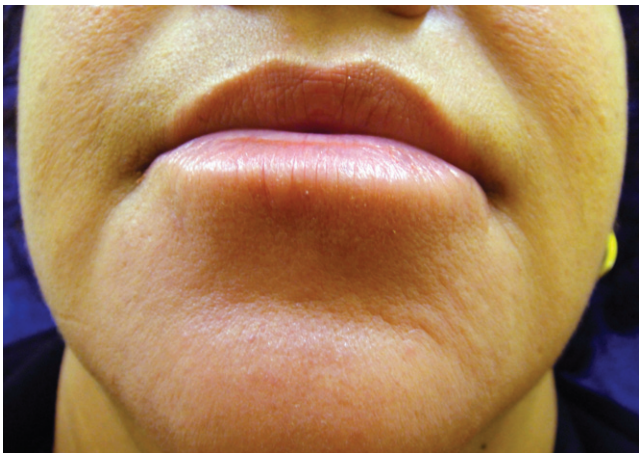

Figure 2: Erythematous and edematous plaque on chin and lower lip. Skin looks like orange peel

DISCUSSION

Rosacea is a common disease that affects mostly middle aged women. It is manifested by transient or persistent erythema, telangiectasia, edema, papules and / or pustules on the face, most typically in the middle. There are several subtypes that include erythematotelangiectatic, papular-pustular, nodular-infiltrative, fulminating, ocular, granulomatous and phymas. [1,2] Phymas, which means swelling or masses, are described as the last stage of rosacea, due to edema and chronic inflammation, which result in tissue hypertrophy and hyperplasia of sebaceous glands. [2,3] They affect mostly males above 40 years of age. [3] Rhinophyma is the most common form of phyma and rarely affects the chin (gnatophyma), forehead (metophyma), one or both ears (otophyma), and one or both eyelids (blefarophyma). [3] The clinical severity of the phyma variant is based on the deformity degree. [4] Gnatophyma has been rarely reported, there are only three cases in the scientific literature, and it may manifest alone or coexist with rosacea lesions. [4,5,6] In those reported cases, one patient is male and two are female, and all of them have injuries exclusively on the chin. The patient reported here is female and also has lesions exclusively on the chin. The process of diffusion and extension of the chin can be attributed to lymphedema, which is a chronic inflammation, due to mechanical failure of the lymphatic system, caused by persistent inflammation of rosacea. [7] Demodex was found in two of three cases mentioned in scientific literature and its presence in rosacea may be due to localized immune deficiency caused by lymphoedema and / or Demodex could serve as a source for antigenic persistent inflammation.[4,5,6] These hypotheses

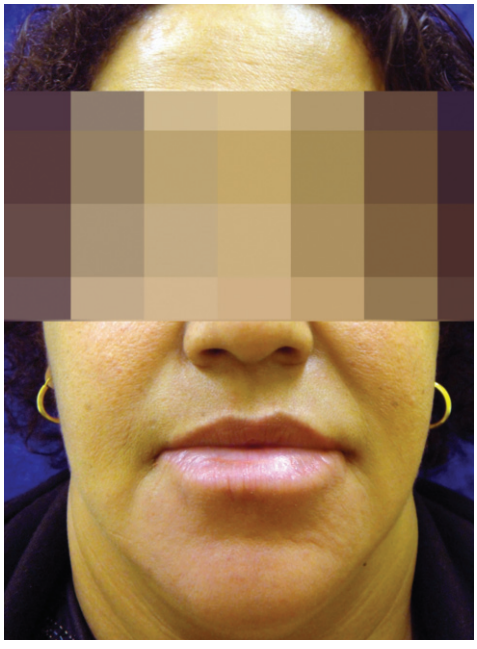

Figure 1: Infiltrated erythematous and edematous plaque on chin and lower lip, without other facial changes

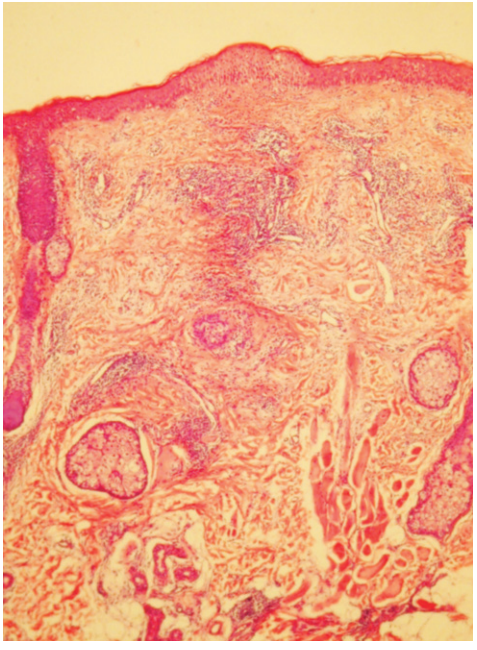

Figure 3: HE – perianexal lymphocytic infiltrate, hypertrophied follicles, sebaceous glands and increased dilated vessels

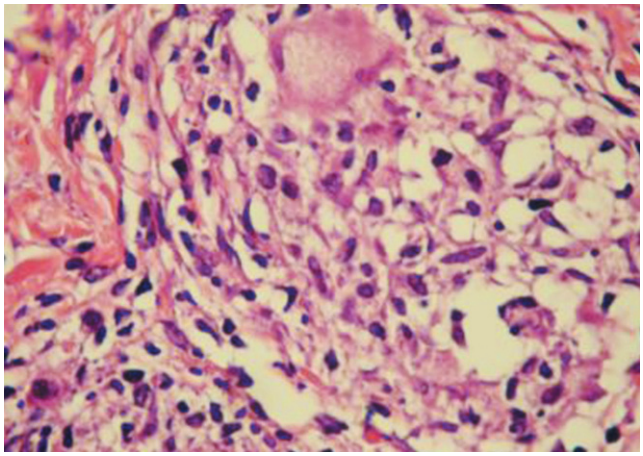

FIGURE 4: HE- Outline of giant cell

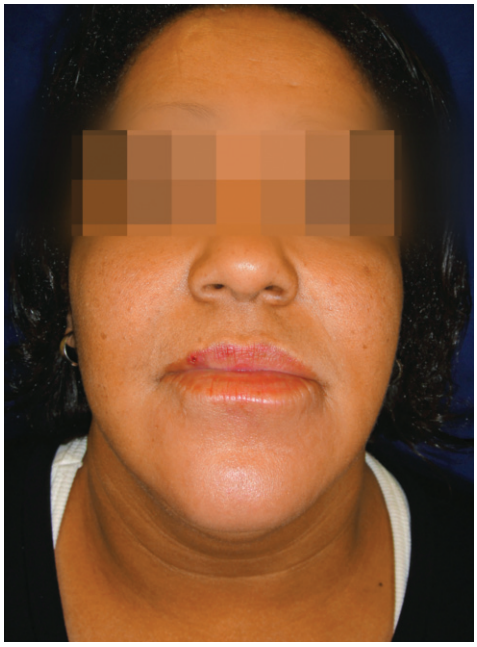

FIGURE 5: Patient after treatment

explain the predisposition to the development of skin cancer on phyma. [7] There are few reports in literature that show association between rosacea and dental diseases. [5,8] They indicate a significant clinical improvement in rosacea after a dental treatment. [8] Histologically, phyma can be divided into the common type, which shows fibrosis and hyperplasia of sebaceous glands, and the severe forms, which resemble elephantiasis caused by chronic lymphedema. [3,7] Our patient had histopathological fibrosis and hyperplasia of sebaceous glands, corresponding to the common type. An important consideration in this case as a differential diagnosis is the Syndrome of Mekelsson-Rosenthal, which is characterized by lip edema, plicated tongue and facial paralysis associated with epithelioid granulomas in histopathology. The classical triad occurs in less than 30% of the cases, and the remainder is represented by mono or oligosymptomatic forms. [9] However, in this case, this diagnosis was excluded because of the other findings in histopathology, hyperplasia of sebaceous glands, perifollicle, periadnexal and perivascular infiltrate, dilated vessels and hypertrophy of the hair follicles associated with non-response to treatment for Syndrome of Mekelsson-Rosenthal, leading to the diagnosis of rosacea. Possible treatments for rosacea include metronidazole cream, azelaic acid or permethrin, atenolol, clonidine, oral antibiotics, including metronidazole, minocycline, doxycycline, clarithromycin, cephalosporins, ivermectin, oral isotretinoin and treatment of Helicobacter Pylori. [1,2,10] However, only antibiotics and oral isotretinoin are capable of reducing the phymas. [1,2,3] To treat the severe type of phyma one can make use of carbon dioxide laser and surgery. [3,4,5,6] Due to the rare occurrence of gnatophyma, the ideal treatment still represents a challenge. The treatment prescribed was tetracycline 1g / day, ivermectin 12 mg single dose, and metronidazole cream 1%. Clinical improvement was observed after one month treatment. ❑

REFERENCES

1. Crawford GH, Pelle MT, James WD. Rosacea: I. Etiology, pathogenesis, and subtype classification. J Am Acad Dermatol. 2004;51:327-41.
2. Wilkin J, Dahl M, Detmar M, Drake L, Feinstein A, Odom R, Powell F, et al. Standard classification of rosacea: report of the National Rosacea Society Expert Committee on the Classification and Staging of Rosacea. J Am Acad Dermatol. 2002;46:584-7.
3. Rohrich RJ, Griffin JR, Adams WP Jr. Rhinophyma: review and update. Plast Reconstr Surg. 2002;110:860-9.
4. Ilyas EN, Hanson MR, Lawrence N, Green JJ. Gnatophyma. J Am Acad Dermatol. 2006;55:165-6.
5. Vidigal MR, Kakihara CT, Gatti TRSR, Tebcherani AJ, Pires MC. Gnatophyma: a rare variant of phyma. Clin Exp Dermatol. 2008;33:743-4.
6. Ezra N, Greco JF, Haley JC, Chiu MW. Gnatophyma and Otophyma J Cutan Med Surg. 2009;13:266-72.
7. Carlson JA, Mazza J, Kircher K, Tran TA. Otophyma: A case report and review of the literature of lymphedema (Elephantiasis) of the ear. Am J Dermatopathol. 2008;30:67-72.
8. Lesclous P, Maman L. An unusual case of a relationship between rosacea and dental foci. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1999;88:679-82.
9. Zanini M, Martinez MAR, Machado Filho CDS. Sindrome de Melkersson-Rosenthal: relato de dois casos e revisão da literatura. Med Cutan Iber AM. 2005;33:113-7.
10. Trindade Neto PB, Rocha KBF, Lima JB, Nunes JCS, Oliveira e Silva AC. Rosácea granulomatosa: relato de caso – enfoque terapêutico. An Bras Dermatol. 2006;81(5 Supl 3):S320-3. An Bras Dermatol. 2006;81(Supl 3):S320-3.

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How to cite this article: Macedo ACL, Sakai FDP, Vasconcelos RCF, Duarte AA. Gnatophyma - a rare form of rosacea. An Bras Dermatol. 2012;87(6):903-5.