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Disseminated extrafacial rosacea with papulonecrotic lesions

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Abstract

Background: Rosacea is a common skin disease and predominantly affects on the face of middle-aged women. It exceptionally occurs on the extrafacial areas such as ear, neck, axilla, and upper extremities, and has been reported as disseminated rosacea.

Main observation: A 40-year-old Japanese female presented with one-month history of erythematous skin eruption with burning sensation on the face, neck, and upper limbs. Physical examination showed rosacea-like eruption on the face as well as multiple papules disseminated on the neck, forearms, and hands. These extrafacial lesions demonstrated papulonecrotic appearance. Bilateral conjunctiva showed marked hyperemic which was consistent with ocular rosacea. Corneal opacity was also seen. Histology of the umbilicated papule on the neck revealed necrobiotic granulomas around the hair follicle with transepidermal elimination. Another tiny solid papule on the forearm suggesting early lesion also demonstrated necrobiosis with palisading granuloma but no transepidermal elimination. Systemic administration of minocycline and topical tacrolimus therapy promptly improved the skin lesions. Topical application of fluorometholone in temporary addition with levofloxacin improved ocular involvement 12 weeks after her 1st visit. The clinical course of the skin lesion and ocular symptoms mostly correlated. Then, the skin lesion and ocular symptoms often relapsed. Rosacea uncommonly associates with the extrafacial involvement as disseminated rosacea. The present case is characterized by the disseminated papulonecrotic lesions of the extrafacial areas histologically showing transepidermal elimination of necrobiotic granulomas.

Conclusions: Dermatologists should recognize that papulonecrotic lesions of the neck and upper extremities might be extrafacial rosacea when the patient has rosacea on the face. (*J Dermatol Case Rep*. 2016; 10(4): 68-72)

Introduction

Rosacea commonly occurs on the central face of the middle-aged woman and categorized into four subtypes; erythematotelangiectatic rosacea (Subtype 1), papulopustular rosacea (Subtype 2), phymatous rosacea (Subtype 3), and ocular rosacea (Subtype 4).[1] The extrafacial lesion of rosacea is extremely rare.[2] We herein report an unusual case of papulopustular rosacea with the disseminated extrafacial lesions histologically showing necrobiotic granuloma with transepidermal elimination.

Case report

A 40-year-old Japanese female presented with a two-month history of erythematous rash with burning sensation on the face and upper extremities. She also complained of tearing, dryness, and foreign body sensation as her eye symptoms. On physical examination, numerous erythematous papules, pustules, and indurated erythemas were symmetrically distributed on the face (Fig. 1a, b). The papulopustular lesions spared the upper and lower eyelids. In addition, multiple, umbilicated papules with an admixture of solid tiny papules

were scattered on the forearms, palms and dorsa of the hands as well as the neck and anterior chest (Fig. 1, 2). These extrafacial lesions on the neck and upper extremities showed papulonecrotic appearance (Fig. 2). Bilateral conjunctiva revealed markedly hyperemic which was termed as blood shot appearance (Fig. 3). Histologic features of an umbilicated papule on the neck demonstrated a defect of epidermis and transepidermal elimination of necrobiotic granuloma around the damaged hair follicle (Fig. 4a, 4b). Another histology of a non-umbilicated papule among the papulonecrotic lesion from the right forearm revealed palisading granuloma consisting of lymphocytes and histiocytes with a few multinuc-

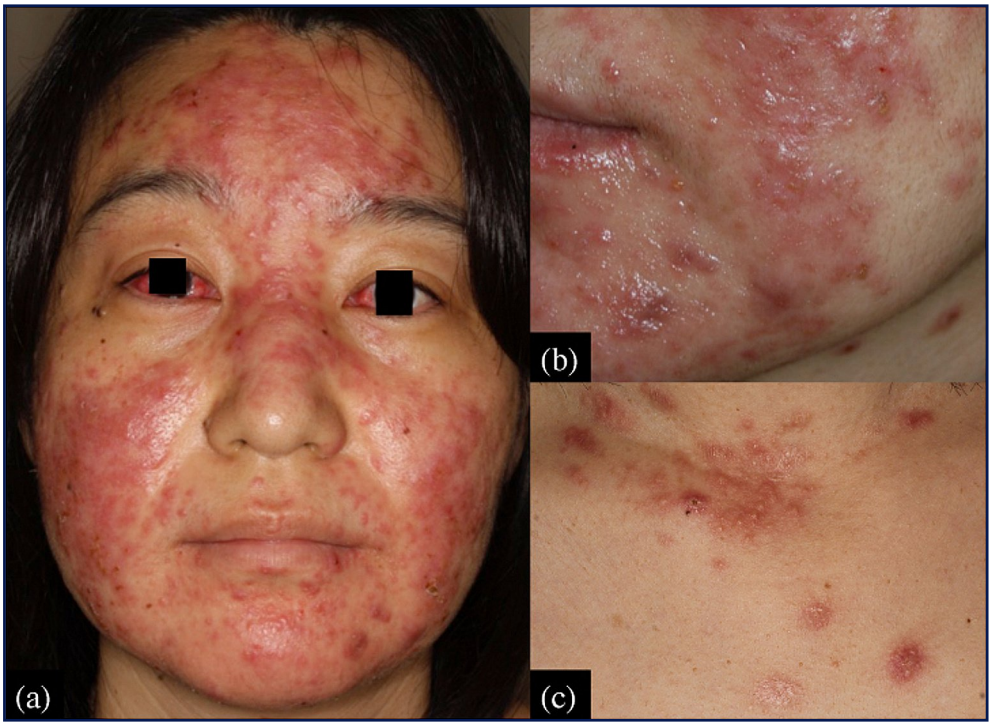

Figure 1

Clinical view of head and neck.
(a) *Multiple papules and nodules were symmetrically distributed on the face. The periocular areas were spared.*
(b) *Close up view of the facial lesion. Papulopustular eruption could be seen around the left cheek.*
(c) *Reddish papules clustered on the neck and anterior chest. Some papules revealed central atrophy or crusting.*

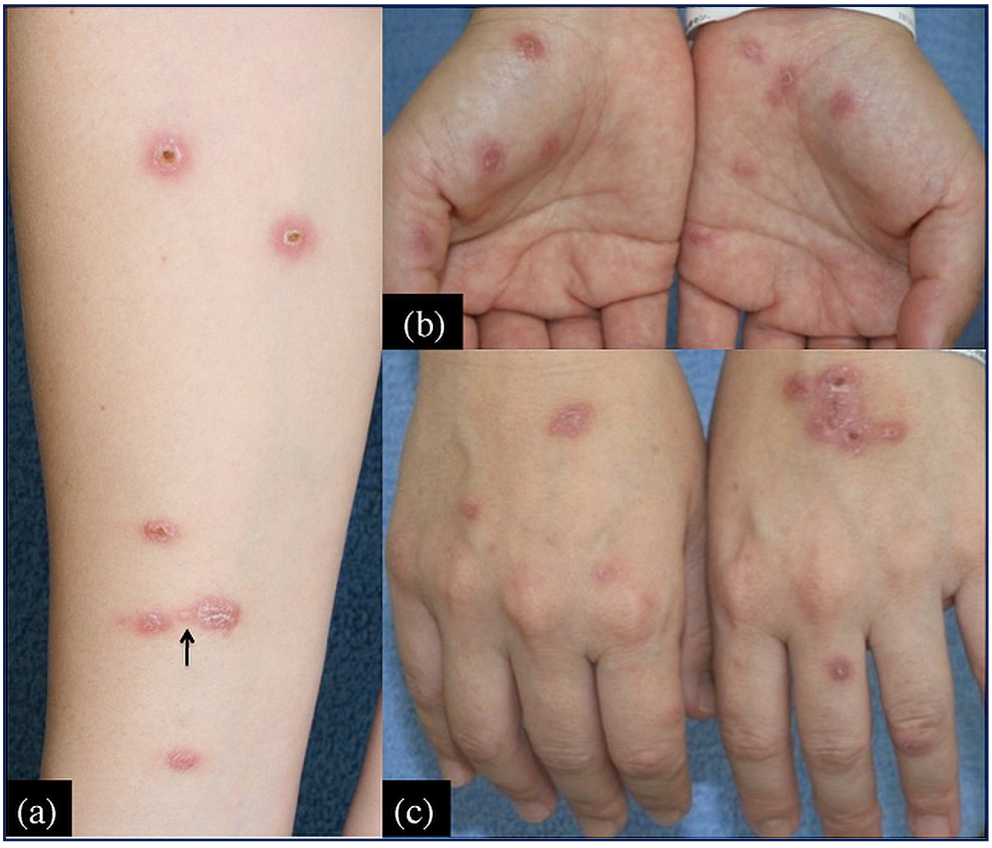

Figure 2

Clinical view of the forearm and hands.
(a) *Extrafacial lesion of the forearm. Multiple, umbilicated and/or crusted papules were scattered on the right forearm. A small papule was taken as biopsy sample of the initial lesion (arrow).*
(b) *Extrafacial lesion of the palms.*
(c) *Extrafacial lesion of the dorsal hands. These lesions demonstrated papulonecrotic appearance.*

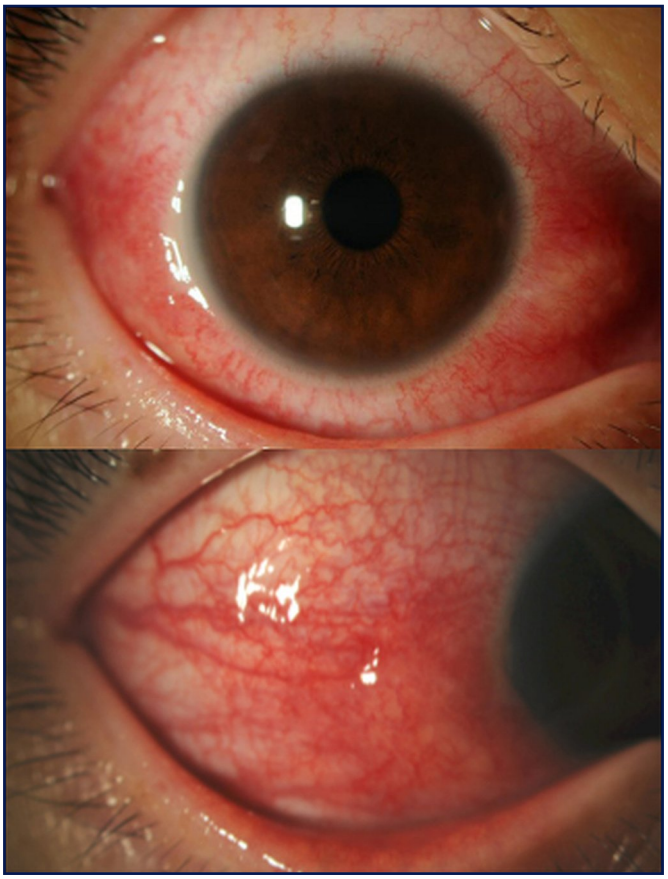

Figure 3
Ocular involvement.
Conjunctiva showed markedly hyperemic, which was consistent with "blood shot appearance" of ocular rosacea.

leated giant cells in the dermis (Fig. 4c, 4d). Transepidermal elimination was not seen in the tiny papule suggesting early lesion. Tuberculosis, sarcoidosis, syphilis, or systemic lupus erythematosus was excluded by blood tests including serum antinuclear antibody, angiotensin converting enzyme, interferon gamma release assay, and radiologic evaluation. She did not have diabetes mellitus. Ophthalmologic examination confirmed the diagnosis of severe conjunctivitis with corneal opacity due to ocular rosacea. Topical tacrolimus and systemic minocycline 200 mg a day for 3 weeks caused marked improvement of the skin lesions. Keratoconjunctivitis was gradually improved by topical application of fluorometholone in temporary addition with levofloxacin 12 weeks after her 1st visit. Visual acuity has not been disturbed. Both facial and extrafacial lesions recurred when minocycline was discontinued. Then, the skin lesion and ocular involvement had repeated the remission and exacerbated. The course of the skin lesion and ocular involvement mostly correlated.

Discussion

Rosacea is a chronic cutaneous disorder affecting primarily the convexities of the central face and presents with a variety of clinical findings.[1] Skin manifestation of rosacea consists of telangiectasia, erythema, papules, pustules, and sebaceous gland hypertrophy. The present case is characterized by (1) papulopustular rosacea-like lesion on the face (2) multiple umbilicated papules disseminated on the neck and upper extremities (3) necrobiotic granulomas with transepidermal elimination around the damaged hair follicle of the extrafacial umbilicated papule. (4) keratoconjunctivitis consistent with ocular rosacea. Similar cases have been reported as disseminated rosacea.[2] Disseminated rosacea occurs not only on the face but also appears on the extrafacial

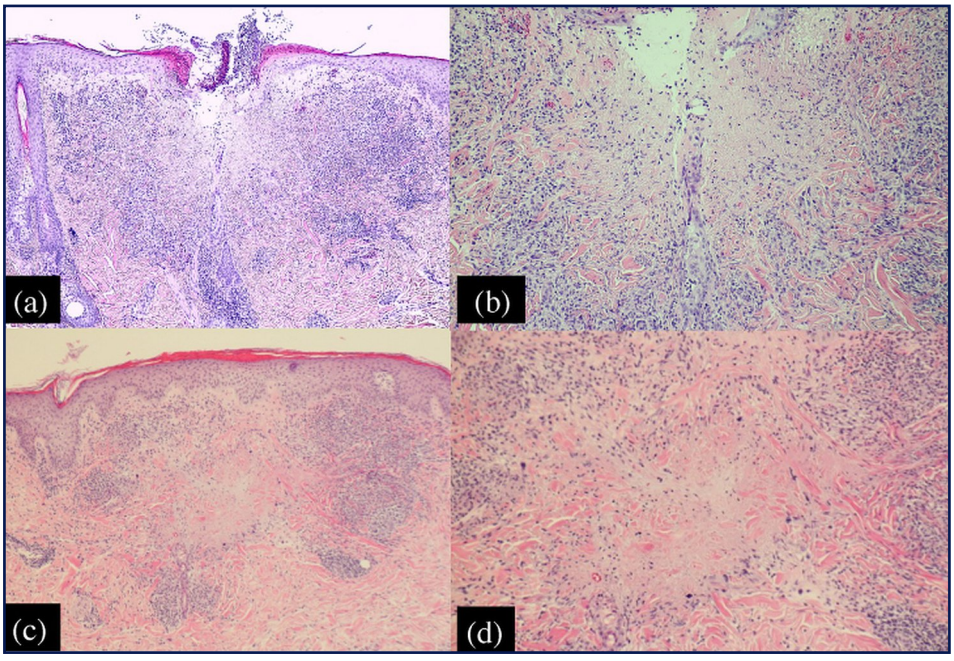

Figure 4
Histology of the matured lesion from the neck and of the early lesion from the forearm.
***(a)** Histologic features of an umblicated papule on the neck. Histology showed a defect of the epidermis through which necrobiotic collagen was eliminated. Inflammatory cell infiltration around the necrobiotic material could be seen. HE x 40.*
***(b)** Higher magnification. Lymphohistiocytic infiltration with an admixture of neutrophils was observed around palisading granulomas. A damaged hair follicle was seen at the center of the necrobiosis. HE x100.*
***(c)** Histologic features of a non-umbilicated small papule on the forearm. Necrobiotic area was surrounded by histiocytes and lymphocytes in the dermis. HE x40*
***(d)** Higher magnification. A few multinucleated giant cells were observed in the granuloma around the necrobiosis. HE. X100.*

areas such as ear, neck, axilla, and upper extremities.[2] Marks & Jones[2] described 14 patients with disseminated rosacea that developed papular lesions predominantly on the upper limbs, wrists, and hands similar in appearance to those on the face in the course of typical facial rosacea. In the literature there are several articles concerning extrafacial lesions of rosacea.[3,4] Extrafacial lesions of rosacea generally revealed multiple, discrete, papules or nodules similar to those of facial lesions.[3,4] Pereira *et al*.[3] reported an unique case of papulopustular lesion extensively developed on the upper limb and arms. Helm *et al*.[4] also described that extrafacial lesions occurred in 15% of the patients with granulomatous rosacea. Disseminated rosacea or extrafacial rosacea might be misdiagnosed or underreported. Extrafacial rosacea clinically presents a variety. Predominant scalp involvement of extrafacial rosacea has been reported.[5] Lupus miliaris disseminatus faciei also occasionally associates the extrafacial lesion such as axilla.[6] The present case is clinically different from lupus miliaris disseminatus faciei because the papulopustular lesions spared the upper and lower eyelids.

Histology of rosacea might reveal granulomatous reaction around the disrupted hair. In rosacea, granulomatous infiltrates are reported to occur in about 10% of rosacea, and caseation necrosis has been identified in about 10% of the patients.[7] Lupus miliaris disseminatus faciei, considered as a variant of rosacea, histologically reveals a dermal granulomatous reaction with central areas of necrosis and occasionally degenerated hair follicles. Marks & Jones[2] also observed focal necrobiotic changes in cellular areas in 2 cases of 14 patients with disseminated rosacea. Thus, rosacea histologically might reveal necrobiotic granuloma as well as caseating granuloma.[4] Necrobiotic materials with palisading granuloma is usually seen in granuloma annulare. Generalized perforating granuloma annulare[8] or sarcoidosis with transepidermal elimination[9] could be considered as a differential diagnosis. Samlanska *et al*.[8] described that perforating granuloma annulare and perforating sarcoidosis may be difficult to differentiate because of a similar clinical appearance and the presence of sarcoidal granulomas in biopsy specimens. These disorders could not be excluded by the histology alone. The present case had no evidence of diabetes mellitus or systemic sarcoidosis by systemic investigation including radiologic survey. Granuloma annulare or cutaneous sarcoidosis had neither rosacea-like eruption on the face nor keratoconjunctivitis. Accordingly, a characteristic ocular involvement, granuloma surrounding the hair in histology, and the effectiveness of oral minocycline also support the diagnosis of rosacea in the present case.

The present case showed severe keratoconjunctivitis without disturbance of visual acuity. There is no diagnostic test for ocular rosacea. Accordingly, the diagnosis relies on the physician's clinical judgment. The clinical course of skin lesion and ocular manifestation mostly correlated in the present case. The diagnosis of ocular rosacea was confirmed by clinical manifestation and clinical course in the present case. Ocular rosacea clinically presents with redness of bulbar and/or palpebral conjunctivae, telangiectasia of conjunctiva and lid margin.[1] Serious corneal complication might cause loss of vision. The symptoms include a foreign-body sensation, dryness, and burning sensation. Ocular involvement is estimated to occur in 6 to 72% of patients with rosacea depending on the series.[10] Systemic treatment is generally with tetracycline. Keratoconjunctivitis in the present case responded relatively well to oral minocycline in combination with topical steroid therapy in spite of corneal complication.

The exact pathogenesis of rosacea is still unknown. Microorganisms such as *Demodex folliculorum*, *Staphylococcus epidermidis*, *Helicobacter pylori*, and *Bacillus oleronius* have been hypothesized to play a role in the pathogenesis of rosacea.[11] It has been suggested that cathelicidin and kallikrein 5 might be involved in the development of the disease. Such inflammatory granulomatous reaction against the damaged hair follicles might play some role in the development of necrobiotic granuloma with transepidermal elimination of the present case.

Conclusions

Pereira *et al*.[3] described that extrafacial rosacea may be more common than it is thought. Dermatologists should recognize that papulonecrotic lesions of the trunk and upper limb might be the extrafacial lesions when the patient has severe rosacea.

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