

A Case with a Delayed Diagnosis of Extrafacial Rosacea

Ekstrafasiyal Rozaseli Tanısı Gecikmiş Bir Vaka

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ABSTRACT Rosacea is a chronic, inflammatory skin disorder involving the central part of the face. There are four main subtypes of the disease which are most commonly seen; but also, when the disease involves other parts of the body except the face, it's called extrafacial rosacea (ER). ER is rarely reported in the literature and it may involve the scalp, chest and the back. Herein, a man with extrafacially located rosacea who couldn't have been diagnosed for long years will be presented because of the rarity of the disease.

Keywords: Extrafacial rosacea; atypical rosacea; rosacea

ÖZET Rozase sıklıkla yüzün santral kısmını etkileyen, kronik, inflamatuvar bir deri hastalığıdır. Dört klasik formu sık görülürken; yüz dışındaki alanlar tutulduğunda ekstrafasiyal rozase (ER) olarak adlandırılmaktadır. ER, literatürde nadiren bildirilmiş olup; saçlı deri, göğüs, sırt gibi alanlarda görülebilmektedir. Burada uzun yıllar tanı koyulamamış atipik bir ekstrafasiyal yerleşimli rozase vakası nadir görülmesi nedeni ile paylaşılacaktır.

Anahtar Kelimeler: Ekstrafasiyal rozase; atipik rozase; rozase

Rosacea is a chronic, inflammatory skin disorder involving the central part of the face.¹ The main subtypes are erythematotelangiectatic, papulopustular, phymatous and ocular types, besides them, there is another form, which is called extrafacial rosacea (ER). ER is reported rarely and it tends to involve the scalp, neck, chest, axillary region, shoulders, arms and legs.²⁻⁴ The diagnosis is generally made through the clinical symptoms and the history of the patient, but it's difficult to make a diagnosis in atypical cases.⁴

Herein, a 66-year-old man with a severe chest and scalp involvement of ER and the clinical course of the patient will be presented.

CASE REPORT

A 66 years-old man applied to our outpatient clinic with the complaints of some acneiform lesions on his chest and scalp for the last ten years. He had widespread erythematous papulopustular lesions on his scalp, chin and chest (Figure 1). He had a clear history in terms of other illnesses. The hemogram,



FIGURE 1: Widespread papulopustular lesions in the chin and chest region.

biochemical parameters, microbiological and fungal culture values, which had been investigated in another medical care center before, were normal. He hadn't responded well to 100 mg/day systemic doxycycline, topical mupirocin cream and ketoconazole shampoo treatments in 3 months, then, we thought that the patient had seborrheic dermatitis and our treatment with 400 mg/day systemic itraconazole for one week in a month didn't relieve the symptoms at all and the patient refused to continue to treatment. Afterwards, a standardized skin surface biopsy was performed to see any demodex spp, but the result was negative. Hence, a biopsy was performed from the lesion on the chest with the prediagnoses of ER and acne conglobata. Histopathological findings indicated subcorneal pustule formation, subepidermal edema, mixed inflammatory cell infiltration, plasma cells, dilated vessels, and folliculitis. These findings were consistent with ER and 20 mg/day systemic isotretinoin and topical sodium sulfacetamide treatments were started. After three months of treatment, the lesions regressed substantially and then the patient stopped the treatment by his own decision because he thought that he was good enough (Figure 2). An informed consent has been obtained

to be able to share the data of the patient in any of the scientific journals or scientific meetings.

DISCUSSION

Rosacea is a common, chronic, recurrent, and inflammatory skin disorder.⁵ Mostly middle-aged women with Fitzpatrick skin types 1 and 2 are affected.⁶ According to the classification system, which has been composed by the National Rosacea Society Expert Committee in 2002; there are four main types (erythematotelangiectatic, papulopustular, phymatous and ocular) and one variant (granulomatous) of the disease.⁷

Generally, the convex parts of the face, except for the perioral and periocular region, are affected but the scalp, neck, chest, proximal back, shoulders, arms and proximal lower extremity involvements have been rarely reported.⁴ Extrafacial region involvement is encountered mostly in men, as in our case. While ER has been usually reported in granulomatous or acneiform type, the erythematotelangiectatic type of ER can also be seen.^{3,4,8} Here, in this case, the lesions were consistent with the papulopustular type of ER.

The etiopathogenesis of rosacea hasn't been fully understood yet. Naive immunity, neurovascular system disturbances, commensal microorganisms like demodex spp., topical steroids, systemic drugs like niacinamide, physical and psychological stress and environmental factors are considered to be among causes.^{1,4,6,8-10}



FIGURE 2: Mild erythema in the chest region after the final treatment.

Diagnosis of rosacea is easily made based on the clinical history of the patient, physical examination and elimination of the other differential diagnoses. But, in some cases which it is hard to diagnose, a biopsy may be required.^{10,11} Histopathology shows generally nonspecific findings. Here in our case, subcorneal pustule formation, sub-epidermal edema, mixed inflammatory cell infiltration, plasma cells, dilated vessels, and folliculitis findings were consistent with papulopustular type rosacea.^{9,11}

For the reason that the differential diagnosis depends on the type of ER, as for this case with papulopustular type of rosacea; acne conglobata, dermatophytosis, demodicosis, bacterial folliculitis, eosinophilic pustular folliculitis, halogenoderma, seborrheic dermatitis were among differential diagnoses. There were no comedonal lesions and no neutrophilic infiltration extending beyond follicle in the histopathology for acne conglobata.¹¹ Microscopic examination of a pustule smear with potassium hydroxide revealed no fungal elements for dermatophytosis. Histopathology and standardized skin surface biopsy were negative for demodex spp. as well. Microbiological culture was negative for bacterial folliculitis. There was no itching clinically and histopathology didn't show eosinophil folliculotropism and this made it easier to exclude the eosinophilic pustular folliculitis.¹² There was no drug history for halogenoderma. The patient didn't respond well to seborrheic dermatitis treatments also. After elimination of those differential diagnoses, with the support of clinical signs and histopathological findings, the patient was diagnosed as having ER.

Treatment options for ER are palliative rather than curative. Topical sodium sulfacetamide, azelaic acid, metronidazole and brimonidine, and systemic sub-antimicrobial-dose doxycycline might be

used in the treatment. The treatment must be supported with education of the patients about the disease and by promoting the sunscreen and emollient usage.^{9,13,14} Systemic isotretinoin with its sebum repressing and anti-inflammatory effects has been also found to be effective on resistant papulopustular rosacea, rhinophyma and ER.^{4,13} Our case had systemic doxycycline treatment for 1 month and systemic itraconazole treatment for 1-week course and there wasn't any sign of regression. Afterwards, a low-dose systemic isotretinoin treatment provided a great relief in 3 months and there was just minimal erythema on the former location of the lesions after 3-months follow up period.

In conclusion, ER diagnosis may be very difficult due to its atypical clinic presentation compared to rosacea, and that may be the reason of why the number of the reported ER cases on literature is very few.³ Eventually, we think that the more frequently it takes part in our differential diagnoses, the more cases will be reported.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

All authors contributed equally while this study preparing.

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