

A Case of Human Orf Complicated with Atypical Erythema Multiforme

Atipik Eritema Multiforme ile Komplike Olmuş Bir Orf Olgusu

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ABSTRACT Human orf is a viral zoonotic infection caused by a parapoxvirus. *Parapoxviruses* are a genus of *Poxvirus* family that infect different species of wild and domestic animals which may serve as reservoirs for human infections. Human parapoxvirus infections are almost invariably arise as a result of occupational exposure. Farmers, veterinarians, butchers are at greatest risk for acquiring infections through contact with an infected animal or an animal product. Although orf is a self-limited disease most commonly resolves spontaneously within weeks, atypical presentations and complications of orf are well known. Here we report a case of human orf who had been presented with atypical targetoid lesions of erythema multiforme. After clinical and histopathological investigations a diagnosis of human orf complicated with atypical erythema multiforme was made. The patient was given symptomatic treatment in conjunction with single dose of prednisolone and within weeks all of the lesions almost completely resolved. Through the presentation of this case report we wanted to emphasize a rare complication of orf.

Key Words: Orf virus; erythema multiforme

ÖZET Orf bir çeşit parapoksvirüsün neden olduğu viral zoonotik bir hastalıktır. *Poksvirüs* ailesine ait bir genus olan *Parapoksvirüsler*, insanlar için rezervuar görevi görebilecek yabani ve evcil çok sayıda hayvan türünü enfekte eder. İnsan parapoksvirüs enfeksiyonları hemen hemen her zaman mesleki maruziyet neticesinde ortaya çıkar. Enfekte hayvan ya da hayvan ürünlerine temas etmeleri nedeniyle çiftçiler, veterinerler, kasaplar yüksek risk grubunda yer alır. Orf her ne kadar birkaç hafta içinde kendiliğinden düzelen selim seyirli bir hastalık olsa da, orfa ait atipik prezentasyonlar ve komplikasyonlar bilinmektedir. Biz de burada, atipik eritema multiforme lezyonları ile başvuran bir orf olgusu sunuyoruz. Olguya klinik ve histopatolojik incelemelerin ardından atipik eritema multiforme ile komplike olmuş orf tanısı konuldu. Hastaya tek doz prednizolonla birlikte semptomatik tedavi verildi ve haftalar içerisinde lezyonlarda tama yakın gerileme izlendi. Biz de bu olgu sunumuyla orfun nadir bir komplikasyonuna dikkat çekmek istiyoruz.

Anahtar Kelimeler: Orf virüsü; eritema multiforme

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Orf is a viral zoonosis caused by a *Parapoxvirus* belonging to the family *Poxviridae*, which is ubiquitous in livestock such as sheep and goats. Infection in humans is primarily occupational in origin, mainly acquired through contact with an infected animal or a fomite.^{1,2} Both humans and animals share similar clinical features, which include limited cutaneous disease typically manifests as contagious pustular dermatitis and mucosal lesions in zoonotic hosts and one or a few number of papulonodular lesions on the infection site in humans.¹⁻⁴ Orf is a self limited benign dis-

ease that generally ends up with spontaneous resolution. However, there are certain numbers of complications which may interfere with the course of the disease. Erythema multiforme (EM) is one of the most commonly reported complication of human orf up to date.¹⁻⁵ Here a case of human orf complicated with atypical EM is presented.

CASE REPORT

A 50-year-old woman came to our outpatient clinic with chief complaints of a painless wound on her hand and a pruritic eruption over distal parts of her limbs. She told that one week after a contact with a healthy-appearing lamb which had been slaughtered for the Feast of the Sacrifice a red lesion appeared on her right hand and on the following 10 th day the pruritic eruption has emerged. She had no other diseases other than well-controlled hypertension and no other family members had any similar skin lesions. Her physical examination was unremarkable. Upon dermatological examination on right hand of the patient a centrally crusted nodule with yellowish raised border approximately 2,5x2,5 in size and on the medial aspect violaceous and flesh-coloured infiltrated papules, some of which had punctum like central depression were observed (Figure 1). On dorsum of both hands flesh-coloured papules and confluent papulovesicular lesions up to 1 cm in size were noticed (Figures 1, 2). On extensor sides of forearms there were slightly elevated

poorly demarcated erythematous lesions, few of which had dusky centers (Figure 3). In addition, especially insteps and dorsum of the feet there were erythematous to violaceous targetoid lesions and on extensor sides of lower legs there were flat violaceous macules (Figure 4). Histopathological examination of one of the targetoid lesions revealed vacuolar interface dermatitis with hydropic degeneration of basal keratinocytes and intraepidermal vesiculation which is consistent with EM (Figures 5, 6). Based on history, clinical and histopathological examinations a diagnosis of human orf complicated with atypical EM was made. Since the history and



FIGURE 1: A fully developed solitary orf lesion with typical central crusting encircled by a peripheral yellowish border, papulovesicular lesions and violaceous targetoid lesions.

(See color figure at <http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/>)



FIGURE 2: Multiple flesh-coloured papules and papulovesicular lesions on dorsum of the hands.

(See color figure at <http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/>)



FIGURE 3: Targetoid lesions on right forearm.

(See color figure at <http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/>)



FIGURE 4: Palpable and non-palpable targetoid lesions on ankle, instep and dorsum of the right foot.

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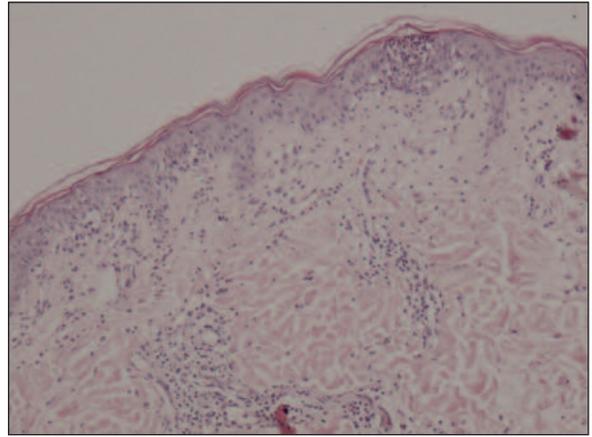


FIGURE 5: Basal hydropic degeneration (H&Ex100).

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dermatological findings of the patient were so demonstrative, we did not perform laboratory investigations like serological tests, virus culture, polymerase chain reaction and electron microscopy which were not available at our hospital. The patient was given wound care and a treatment consisting of topical corticosteroid, systemic antihistamines and a single dose of intramuscular prednisolone (80 mg). At the 15th day of follow-up, the patient showed significant clinical improvement (Figure 7).

DISCUSSION

Orf, also known as ecthyma contagiosum or contagious pustular dermatitis, is a viral infection most commonly affects farmers, shepherds, veterinarians, and butchers as a result of occupational exposure.^{2,4,6}

On the other hand, children are also at higher risk for being infected after petting a diseased animal in a zoo or a fair.³ Although orf occurs worldwide, exceptional outbreaks develop after the Feast of Sacrifice in most of the Muslim countries.^{3,7-9} After an incubation period of 1 or 2 weeks, the typical initial papule of human orf evolves on the contaminated abraded skin.^{1,3,4,10} Though unusual localisations have been described, hands are the most frequent site of involvement.^{2,3} This initial erythematous papule slowly enlarges into a haemorrhagic pustule or bulla which consistently crusts centrally. Indeed orf progresses through six sequential stages each with its own characteristics.^{1-4,10,11} As a fully developed lesion, in the target stage, orf typically has a crust over the umbilicated center and greyish white

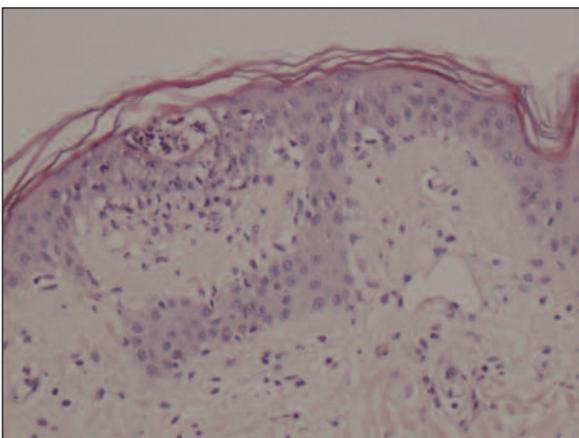


FIGURE 6: Intraepidermal vesiculation (H&Ex200).

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FIGURE 7: Clinical improvement of the patient after the treatment.

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raised rim surrounded by outer erythema. Gradually, weeping nodular lesion regresses, papillomas and dry crust appear atop the lesion and eventually orf spontaneously resolves without scarring within 3-6 weeks.^{1,4,10}

Orf is a self-limited benign disease. Nonetheless it tends to cause some mild but fairly common complications.^{2,4,10} EM is one of the most common complications of human orf.^{1-3,5,10,12-14} In fact EM is a type of hypersensitivity reaction triggered by certain kinds of infections and drugs.¹ Recently Joseph et al. indicated that in the literature up to date 10 cases of human orf complicated with EM have been reported and about 30 similar cases could be identified from the epidemiological and immunological studies. In addition, they affirmed the estimated incidence of EM in association with orf as 7-18%.¹ On the other hand, the exact mechanism underlying this association remains unresolved. It appears to be related to virulence factors of *Parapoxvirus* modulating the host's immune system and response.¹

Other than EM, an obscure complication, papulovesicular eruption after orf infection have been described.^{2,15,16} In 1977 Wilkinson reported a family with orf infection who suffered unusual complications including widespread papulovesicu-

lar eruption consisting of scattered and isolated vesicles or papulovesicles over face, scalp, trunk, limbs and mucosal surfaces.¹³ Bassioukas et al. also published similar cases demonstrating analogous eruption, two of which had widespread distribution and one with papules, vesicles and papulovesicles on dorsal hands, forearms and lateral aspect of the neck.² Like EM the pathogenetic mechanism of the papulovesicular eruption has not been clarified, although it has been suggested to be an unusual pattern of a vesicular exanthem.^{15,16}

Here, we present a case of human orf complicated EM with typical history, clinical and histopathological findings. Even though orf is a fairly common disease and EM is a prevalent complication of orf, we assume that with that distinctive atypical targetoid lesions and papulovesicular eruption, our case is particularly unique. To the best of our knowledge, in the literature there is not any instance of human orf complicated with atypical EM and papulovesicular as in our case. More importantly, since orf can easily be misdiagnosed and unnecessarily overtreated, we think that case reports of human orf should appear in the literature more commonly to overcome underdiagnosis and maltreatment of orf.

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