Human Orf: An Under-recognized Entity

Dear Editor,

Orf, also called contagious ecthyma, contagious pustular dermatitis, sore mouth, or scabby mouth, is a highly contagious zoonotic disease which is caused by a double-stranded DNA virus, ORFV (Parapoxvirus genus of the family Poxviridae) (1). The infection is endemic to sheep and goats, and humans are infected either through direct transmission from active lesions on infected animals or through contact with fomites (2). Orf is an occupational hazard and the population at risk includes shepherds, butchers, farmers, wool shearers, and veterinarians (2,3). Professionals rarely seek medical attention as they are aware of its benign nature (4).

A 59-year-old woman presented with two painful plaques involving the dorsal aspect of her right



Figure 1. Two-centimeter and six-centimeter firm plaques with an erythemato-violaceous infiltrated rim and pale white center involving the dorsal aspect of the right thumb and the left first interdigital fold.

thumb and the first interdigital space of the left hand. On examination, targetoid appearance with a central umbilication surrounded by a violaceous bullous halo was observed (Figure 1).

The patient had been raising goats on her farm that recently presented udder and perioral crusted lesions. She did not wear gloves while performing this task. A skin biopsy was performed and showed elongation of the rete ridges, edematous papillary dermis, and eosinophilic intracytoplasmic inclusions in vacuolated cells (Figure 2). A diagnosis of human orf was established and the patient was started on a topical antiseptic solution to prevent secondary infection. The lesions healed without scarring after 6 weeks.

Orf is characterized by one or multiple nodules on the hands and fingers, but also on the feet, legs, neck, and face. After an incubation period of 3-7 days, the lesions evolve through six clinical stages (2-4): (1) maculopapular stage (days 1-7), with erythematous macules or papules; (2) target stage (days 7-14), with necrotic center and red outer halo; (3) acute stage (days 14-21), in which the nodule begins to weep; (4) regenerative stage (days 21-28), in which the nodule becomes dry; (5) papilloma stage (days 28-35), where the lesion become papilloma-like and forms a dry

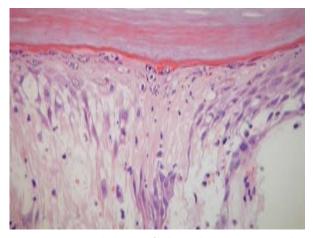


Figure 2. Histopathological findings showing eosinophilic intracytoplasmic inclusions in vacuolated cells; H&E x100.

crust; (6) regression stage (after 35 days). The lesions may be accompanied by lymphangitis, lymphadenopathy, malaise, fever, erysipelas, and occasionally erythema multiforme and bullous pemphigoid (3).

Orf is usually a clinical diagnosis (2-4). The characteristic clinical appearance and location of the lesions along with the history of contact with infected animals is sufficient to establish a diagnosis. The differential diagnosis includes milker's nodule, cutaneous anthrax, neutrophilic dermatosis, atypical mycobacterial infection, cutaneous leishmaniasis, pyogenic granuloma, keratoacanthoma, and fungal infection (4,5). Further investigations are performed only when the diagnosis is in doubt and include electron microscopy, virology, enzyme-linked immunosorbent assay, or PCR-based approaches (4). The histological findings depend on the stage of the lesion and include eosinophilic inclusion bodies, epidermal necrosis, vacuolated keratinocytes, a dense mixed dermal infiltrate, and delicate finger-like projections in the epidermis (6). There is no specific treatment since the disease resolves spontaneously within 6-8 weeks, but successful application of cryotherapy, topical imiquimod, and cidofovir has been reported without supporting evidence (4). Surgical debridement should be avoided because it prolonged the recovery period (5). For prevention, wearing nonporous gloves, washing after handling, and isolation of infected animals is effective (2,4).

In the present case, the diagnosis of orf was established in a straightforward manner after a good clinical examination and occupational history, allowing us to reassure the patient on its benign nature. The knowledge of this diagnosis prevents multiple complementary investigations (blood tests, histopathology, skin cultures, PCR detection, and electron microscopy) and unnecessary overtreatment. Although a rare entity, the predominant hand involvement in professionals can have significant morbidity that reflects on their productivity and quality of life. This reinforces the need for using appropriate measures to prevent animal-to-human transmission.

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