A Case of Human Orf Contracted From a Deer

John T. Kuhl, BS; Christopher J. Huerter, MD; Hisham Hashish, MD

GOAL

To gain a thorough understanding of orf

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

- 1. Recognize the clinical presentation of orf.
- 2. Identify the histopathology of orf.
- 3. Understand the stages of orf.

CME Test on page 323.

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Orf, or contagious ecthyma, is a rare viral dermatosis caused by a member of the genus Parapoxvirus. The typical lesion consists of solitary or multiple papules that progress through a series of stages, terminating in complete resolution. This zoonotic disease is most commonly transmitted to humans from infected sheep or

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Mr. Kuhl and Drs. Huerter and Hashish are from Creighton University School of Medicine, Omaha, Nebraska. Mr. Kuhl is a medical student, and Dr. Hashish is a resident in the Department of Pathology. Dr. Huerter is Associate Professor and Head, Division of Dermatology.

Reprints: Christopher J. Huerter, MD, 601 N 30th St, Suite 5850, Omaha, NE 68131 (e-mail: chuerter@creighton.edu).

goats. We report a case of human orf, likely contracted from exposure to deer carcasses.

rf, also known as contagious ecthyma, scabby mouth, soremouth, contagious pustular dermatitis, is caused by a member of the genus Parapoxvirus in the family Poxviridae.^{1,2} The virus typically infects sheep or goats and produces papulovesicular lesions on non–wool-bearing areas (eg, gums, lips, nose, groin).^{3,4} The infection can be transmitted to humans either by direct contact with an infected animal or by indirect contact with contaminated fomites.^{4,5} After an incubation period of 3 to 7 days, an erythematous maculopapular lesion



Erythematous boggy nodule characteristic of later stage orf.

develops.^{1,4} Orf lesions are benign and progress to complete resolution in 6 weeks.⁴

Case Report

A 51-year-old woman presented to her primary care physician for evaluation of a lesion on the metacarpal-phalangeal joint of the left second digit. A raised annular lesion was identified, and the patient was started on dicloxacillin therapy. On reexamination 4 days later, an enlarging 2-cm nodule with signs of central necrosis was found. The lesion was incised, and a bacterial culture of purulent exudate was obtained. Gram stain and initial culture results were negative. The patient also complained of diarrhea, which was attributed to use of dicloxacillin; this antibiotic was replaced with clarithromycin. The patient was reevaluated the next day for signs of cellulitis and ascending lymphangitis. She denied constitutional symptoms or fever and chills. The lymphangitis was present on the dorsum of the left hand to the level of the wrist. Ceftriaxone was injected intramuscularly. Three days later, an enlarging lesion with serous discharge was identified. The patient was referred for further dermatologic evaluation.

On examining the patient, we found a 2.5-cm boggy fluctuant nodule over the metacarpalphalangeal joint of the left second digit (Figure). The patient complained of pain, and the hand showed signs of lymphangitic spread. When her history was taken, the patient indicated that she worked at a deer station. Her job involved handling deer carcasses, and her bare hands had been exposed to blood from slaughtered deer. She denied having any contact with sheep, goats, cattle, and farm structures containing those animals. Results of bacterial cultures were negative, and a differential diagnosis of deep fungal infection, atypical mycobacterial infection, or contagious ecthyma was entertained. Two 3-mm punch biopsies were performed. Stains for acid-fast bacilli and fungi were negative. Results of microscopic examination showed vacuolation of cells in the upper third of the malpighian stratum, eosinophilic inclusion bodies, and massive dermal infiltrate.

Because of the patient's clinical presentation, microscopic findings, and negative culture results, orf was diagnosed. Oral antibiotic therapy was discontinued, and daily cleansing and use of bacitracin ointment were started. The lesion resolved completely over the ensuing 3 to 4 weeks.

Comment

The diagnosis of orf is suggested by a characteristic skin lesion and a history of exposure. The typical lesion begins as a solitary papule on a finger, a hand,³ or the face. Orf lesions classically progress through a series of 6 clinical and histopathologic stages, each lasting approximately 1 week. In the initial maculopapular stage, an erythematous macule or papule erupts. In the target stage, the lesion becomes a papule with a red center, a white middle ring, and a red halo. In the acute stage, the lesion becomes a weeping nodule. In the regenerative stage, the lesion dries, a thin yellow crust develops, and small black dots form on the surface. In the papillomatous stage, tiny papillomas form on the surface. In the final, regressive stage, a dry crust forms. 1,3,4,6 Uncomplicated lesions rarely leave a residual scar.4

The histopathology of an orf lesion evolves with the clinical stages and helps to secure the diagnosis. The maculopapular and target stages are characterized by vacuolated epidermal cells. In the maculopapular stage, cells have intracytoplasmic inclusions; in the target stage, they have both intracytoplasmic and intranuclear inclusions. The acute stage is marked by patchy areas of lost epidermis, reticular degeneration of the epidermis, and a

dermal infiltrate composed primarily of lymphocytes. The regenerative stage involves epidermal regeneration and extrusion of pyknotic hair-follicle cells that form the small black dots on the surface of the lesion. In the papillomatous and regressive stages, fingerlike downward projections of epidermis are evident.^{1,4,6}

The diagnosis of orf is further supported by a history of exposure to infected animals. The most compelling history involves exposure to sheep or goats, but the orf virus has infected other animals, including musk oxen⁷ and camels.⁸ Experimental inoculation has produced contagious ecthyma lesions in mule deer, white-tailed deer, pronghorn, and wapiti.⁹ That wild deer could contract orf and that the infection could be transmitted to humans through direct contact seem reasonable speculations. Our patient's clinical picture suggests that this mechanism may have been involved in her contracting the disease.

Electron microscopic views of characteristic viral particles in the cytoplasm of keratinocytes provide the definitive diagnosis of orf.^{1,10} Other diagnostic studies are tests of viral culture, complement fixation, and immunofluorescence.^{1,4,6} These tests are used mainly for epidemiology rather than clinical diagnosis.⁶

Complications of orf are generally rare but may include fever, chills, rigor, drenching sweat, malaise, lymphadenopathy, and lymphangitis. 1,6,10 Secondary bacterial infection is the most common complication. Cases of erythema multiforme also have developed in the presence of orf. 11

As a benign self-limited disease, orf requires no specific treatment. Antibiotics should be administered in cases of secondary bacterial infection but are otherwise unnecessary. 1,2,6 Regression of the lesion may be accelerated by application of idoxuridine. 10 Surgical excision also can bring about rapid resolution but is generally contraindicated because the lesion spontaneously regresses without leaving a scar. 2 Corticosteroids and other immunosuppressive drugs should be avoided because they can exacerbate the lesion in its papillomatous stage. 6,12 Use of

topical cidofovir has been beneficial in treating patients who are immunocompromised.¹³

Although orf is a benign disease with a striking presentation that is easy to spot, early diagnosis can prevent unnecessary diagnostic workup and treatment. Orf should be considered in patients presenting with the characteristic skin lesion and a history of exposure to sheep or goats. As with our case, however, when a patient presents with a characteristic lesion and a history of exposure to other animals, orf should be kept in the differential diagnosis.

REFERENCES

- 1. Huerter CJ, Alvarez L, Stinson R. Orf: case report and literature review. Cleve Clin J Med. 1991;58:531-534.
- Chahidi N, de Fontaine S, Lacotte B. Human orf. Br J Plast Surg. 1993;46:532-534.
- Leavell UW, McNamara MJ, Muelling R, et al. Orf: report of 19 human cases with clinical and pathological observations. JAMA. 1968;204:109-116.
- 4. Mendez B, Burnett JW. Orf. Cutis. 1989;44:286-287.
- 5. Wilkinson JD. Orf: a family with unusual complications. Br J Dermatol. 1977;97:447-450.
- Bodnar MG, Miller OF, Tyler WB. Facial orf. J Am Acad Dermatol. 1999;40:815-817.
- 7. Zarnke RL, Dieterich RA, Neiland KA, et al. Serologic and experimental investigations of contagious ecthyma in Alaska. *J Wildl Dis.* 1983;19:170-174.
- 8. Azwai SM, Carter SD, Woldehiwet Z. Immune responses of the camel (*Camelus dromedarius*) to contagious ecthyma (orf) virus infection. *Vet Microbiol*. 1995;47:119-131.
- 9. Lance WR, Hibler CP, DeMartini J. Experimental contagious ecthyma in mule deer, white-tailed deer, pronghorn and wapiti. *J Wildl Dis.* 1983;19:165-169.
- 10. Lo C, Mathisen G. Human orf in Los Angeles County. West J Med. 1996;164:77-78.
- 11. Agger WA, Webster SB. Human orf infection complicated by erythema multiforme. *Cutis.* 1983;31:334-338.
- 12. Mohr BW, Katz D. Orf: a case report. Henry Ford Hosp Med J. 1989;37:79-80.
- 13. Geerinck K, Lukito G, Snoeck R, et al. A case of human orf in an immunocompromised patient treated successfully with cidofovir cream. *J Med Virol*. 2001;64:543-549.

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