Orf: case report and literature review

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■ Ecthyma contagiosum, or orf, is an uncommon dermatosis resulting from cutaneous infection with sheep pox virus. It is generally a benign and self-limited condition. Early clinical recognition is paramount to avoid unnecessary surgical intervention or extensive diagnostic workup. Diagnosis is usually based on a clinically typical skin lesion, characteristic histology, and a history of sheep exposure. We report the case of an infected sheep farmer and review the literature.

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ECTHYMA CONTAGIOSUM, or orf, an uncommon dermatosis resulting from cutaneous infection with sheep pox virus, is a generally benign, self-limited condition which can be confused with other more dangerous conditions. Early clinical recognition helps to avoid unnecessary surgical intervention or extensive diagnostic workup. We report a case of orf in a sheep farmer, and present a review of the literature.

CASE REPORT

A 26-year-old sheep farmer presented to his family doctor with complaints of an enlarging skin lesion over the right chin area present for several weeks. The patient reported the lesion to be asymptomatic and denied fever or chills. A bacterial culture was performed, and the patient was admitted to the hospital for treatment of a presumed staphylococcal abscess. Bacterial culture grew a moderate amount of *Staphylococcus epidermidis*, and the patient showed no improvement on intravenous nafcillin.

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A dermatology consult was obtained. A 2-cm erythematous, crusted nodule was noted over the right chin area (*Figure 1*). The diagnosis of orf was entertained and a partial shave excision was performed for biopsy.

Skin biopsy revealed focal areas of full thickness epidermal necrosis with ulceration and multilocular subcorneal vesicles (*Figure 2*). Within the dermis, dilated hair follicles containing pyknotic epidermal cells and surrounded by a dense lymphohistiocytic infiltrate were noted (*Figure 3*). These findings were consistent with a diagnosis of ecthyma contagiosum in the acute stage.

At a follow-up visit 2 weeks after biopsy, the nodule had decreased in size. The lesion had become more dry with superficial overlying scaling. He denied any associated symptoms at that time. The patient reported over the next few weeks that the skin lesion continued to resolve. The only topical therapy was bacitracin ointment applied after the shave biopsy.

DISCUSSION

Classified as belonging to the paravaccinia subgroup of the pox virus,[1] scabby mouth, sore mouth, ovine pustular dermatitis, contagious pustular dermatitis, and orf are all synonyms for ecthyma contagiosum, a nonpustular lesion.[2]

FIGURE 1. Erythematous, crusted nodule typical of the acute stage of orf infection.

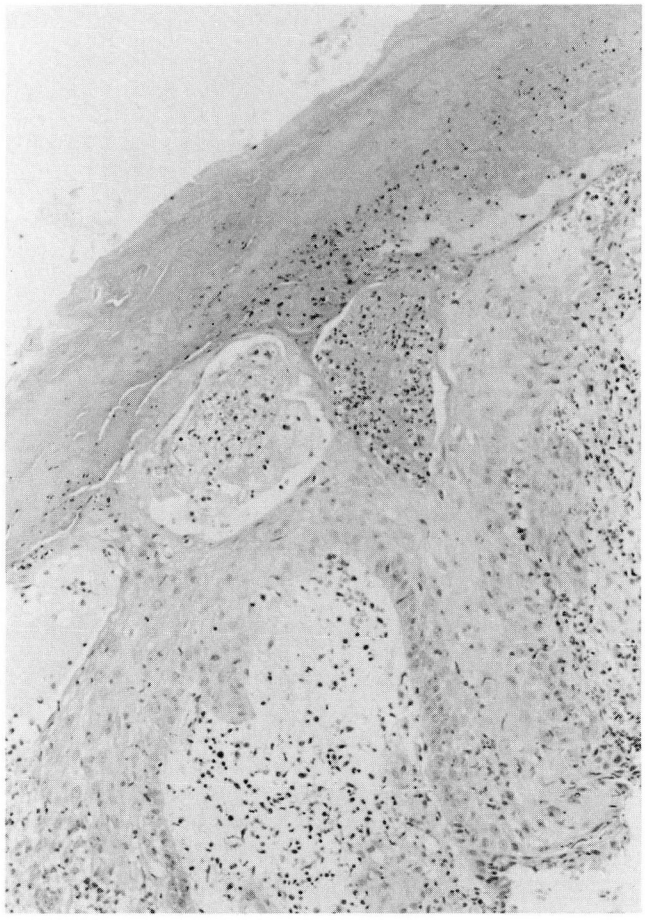

FIGURE 2. Epidermal necrosis, multilocular subcorneal vesicle formation and an inflammatory infiltrate consisting of lymphocytes, neutrophils, and eosinophils (hematoxylin-eosin, x 100).

The designation "orf" is derived from the Anglo-Saxon name for cattle and is actually a misnomer for the disease which, though naturally occurring in sheep, goats, and camels,[3] has never been reported in cattle. However, milker's nodule, an endemic pox virus disease in cows, presents with a clinical and histologic picture identical to orf,[4] as well as having a similar viral ultrastructural appearance.[5]

First reported in sheep in 1787 by Steeb, it was later described in goats in 1879.[5] A clinical description of the disease in humans was first made by Newson and Cross in 1934.[6] Though it was initially thought not to be transmissible by human-to-human contact, it became apparent that such a route of infection was possible.[7]

Leavell and associates[8] described six stages of orf in sheep and in man, both closely paralleling one another. Each stage lasts approximately 7 days. After an incubation period of 3 to 7 days, the first stage presents with an erythematous macule which subsequently becomes vesicular. Next, a "bulls-eye" lesion with an erythematous center, white middle ring, and surrounding red halo indicates the presence of the target stage. From this stage, a nodule then develops. Drying, resolution, and the final complete healing of the lesion occur progressively during the regenerative, papillomatous, and regressive stages, respectively. When uncomplicated, the lesions resolve without scarring.

Transmission in man generally occurs by contact with an infected animal, whether dead or alive. The saliva of such an animal is highly infectious, and dried saliva crusts may remain infective for several months.[7] The fomite route of infection is relatively frequent. At room temperature, viral infectivity has remained for greater than 15 years, and with more than 22 years of refrigeration infectivity still existed.[5]

The diagnosis of orf can usually be deduced by a characteristic clinical appearance of the asymptomatic lesion and a history of direct or indirect handling of sheep. The lesions are usually single, but can be multiple, and most frequently occur on the hands and forearms.[2] Definitive diagnosis is most often made by electron-microscopic visualization of the characteristic virus within the cytoplasm of keratinocytes.[9] However, identification of the pox virus by viral culture,[10] a progressive increase in the serologic titer of orf virus antibodies,[2] immunofluorescent detection of viral an-

tigens,[11] and complement fixation[12] are other methods of diagnosis.

Routine histopathology is helpful in securing a diagnosis. The first two stages are characterized by vacuolization of epidermal cells which contain eosinophilic intracytoplasmic and intranuclear inclusions. Our patient presented in the acute stage, which reveals focal epidermal necrosis, multilocular epidermal vesicle formation, dermal inflammation, and pyknotic keratinocytes within hair follicles. In the final three stages, there is epidermal regeneration and the development of epidermal papillomatous projections into the dermis.

Although rare, accompanying systemic symptoms can be severe, and include rigors, drenching sweat, fever, and leukocytosis.[13] Additional findings associated with orf infection may include malaise,[14] urticaria,[15] lymphadenopathy, lymphadenitis, and lymphangitis.[8]

Varied presentations reported include a generalized bullous orf beginning 1 month after an initial lesion and requiring 1 month to clear.[14] A generalized, pruritic, varicelliform eruption involving skin and mucous membranes has been reported, as well as the development of "metastatic lesions" 24 hours after typical lesions developed on the hands.[13,15] Ocular,[16] scrotal,[17] and perianal[17] sites of vesicular eruption have been reported. Permanent blindness has resulted from infection of the eye.[5] A case of orf occurring during pregnancy was reported.[18] The involved woman gave birth to a healthy child with no signs of fetopathy caused by the virus.

Secondary bacterial infections seem to be the most common complication of ecthyma contagiosum. Erythema multiforme developing soon after the appearance of orf has been described, and in 119 cases studied by Johanessen, it was found to be a relatively frequent complication and the main reason for seeking medical attention.[5,19,20] Stevens-Johnson syndrome has been cited less frequently.[5,21] The resemblance of the orf lesion to keratoacanthoma, granuloma pyogenicum, and giant molluscum contagiosum may result in misdiagnosis. In one instance, the rapid growth of a lesion was mistaken for malignancy, and a finger was amputated.[5] Ecthyma contagiosum also needs to be distinguished from a felon to avoid overtreatment.[22]

The number of cases of orf does not correlate with exposure to a higher concentration of sheep.[5] It can be speculated that those with chronic exposure are more

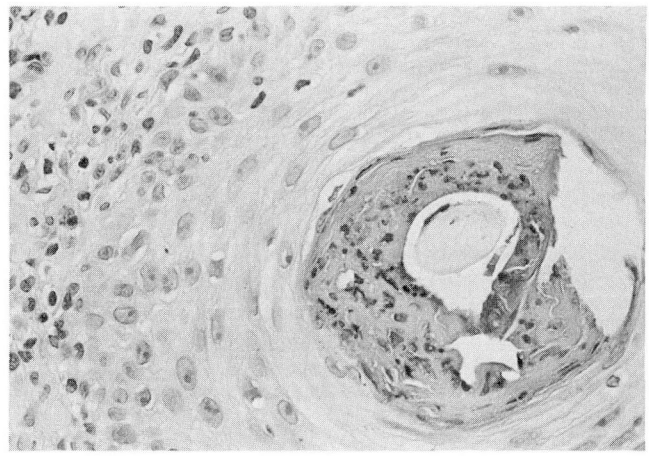

FIGURE 3. Dilated hair follicle containing pyknotic epidermal cells and surrounded by a lymphohistiocytic infiltrate (hematoxylin-eosin, x 400).

apt to acquire protective antibodies. Pask et al[23] demonstrated that solid immunity of unknown duration occurred following primary infection, and no recurrences were reported in all 19 cases reported by Leavell.[8] Contradictory are the reports of relatively frequent reinfection.[24,25] Sanchez and associates[26] reported a case in a temporarily immunosuppressed patient with viral hepatitis. The histologic features in this patient were not typical and suggested the possibility of malignancy. However, the patient recovered with no complications.

Because of the benign, self-limited nature of the disease, no treatment is required. Antibiotics to prevent superinfection have been employed with the addition of hot compresses, if desired. Nonetheless, the use of idoxuridine resulting in the more rapid resolution of lesions has been reported.[27] Surgical excision is another alternative,[28] with advantages which include rapid removal of the lesion, prevention of lesion extension and contagion, and accelerated healing. Lesion location, size, and implications of scar formation are obvious considerations before proceeding with surgery. The use of corticosteroids is not indicated.[29]

In summary, ecthyma contagiosum is a benign, self-limited, occupational viral infection associated with exposure to sheep and goats. Complications are infrequent and treatment is generally not indicated. However, correct diagnosis is important to avoid unnecessary testing and the more severe consequences of improper treatments.

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