Disseminated Orf: An uncommon presentation

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Abstract

Orf or Ecthyma contagiosum is a viral dermatosis found mainly in animals, and in humans via zoonotic transmission from an infected animal. We herein, describe a case of Orf in an immunocompetent lady with a positive history of direct contact with a diseased animal in her neighbourhood. The patient presented to us in the out-patient department with multiple, large, ulcerated violaceous nodules on both her upper and lower limbs. The diagnosis was made on clinical grounds. It was later confirmed via histopathology. Where we might regard orf as a solitary appearing nodule, its unusual clinical patterns must not be missed in order to avoid unnecessary investigations and interventions.

Key words

Orf, poxvirus, ecthyma contagiosum.

Introduction

Ecthyma contagiosum, or Orf was initially reported in 1955 in sheep found in China.¹ It is commonly found in animals, cattle handlers, butchers, farmers, wool shearers, slaughterhouse workers, and veterinarians.² There is no predilection for age, sex or ethnicity of this disease. As seen in the literature, it has been reported from various parts of the world.

The causative organism, Parapoxvirus, is transmitted to humans via infected animals or infected meat. In dermatological settings it is commonly known as, contagious pustular dermatitis, infectious pustular dermatitis, or ecthyma contagiosum.² Mode of transmission is through simple breach in the skin integrity of the host. The virus is resistant to heat or cold, hence it can be transmitted from objects contaminated by the virus earlier on, such as fences, knives and clothing material. The virus has a high affinity for the epithelial lining, and results in a highly infective vesiculo-pustular lesion of the keratinous skin and the mucosal surface of the oral cavity.²,³ The virus remains localised to the epidermis without any systemic spread.

Diagnosis is usually clinical, but may require histopathological assessment, PCR or electron microscopy where the history is not clear and there is a confusion between other possible diagnosis. The histopathology findings are consistent with that of epidermal invasion by a virus. There is ballooning degeneration of the keratinocytes, cytoplasmic and nuclear vacuolation along with a dense inflammatory infiltrate of mixed cells. Epidermal hyperplasia and parakeratosis are also seen.¹ ⁴ Viral serology and PCR can also be done to confirm the diagnosis. Patients infected by this virus do not need any pharmacological intervention as the condition is self-limiting and resolves in 6-8 weeks. Treatment is needed if there is a superimposed bacterial infection, giant lesions, EM like reaction or other systemic symptoms.⁵

Aseptic technique and careful handling of infected cattle and meat must be practiced.

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Vaccination, currently seems to be an effective way of reducing the spread of the disease and decreasing the severity of the clinical presentation in animals. Attenuated tissue culture vaccines have proven to be more beneficial. As for humans, an effective vaccine against Orf is yet to be developed.

The case reported here is of a patient with multiple ulcerated lesions on her upper and lower limb extremities after coming in contact with a diseased farm animal.

Case report

A 35-year-old married female, belonging to Ormara, Baluchistan presented to the out-patient department with a 4-week history of multiple erythematous and ulcerated nodules and plaques on her hands and feet (Figure 1a & 1b).

She had a history of an acute febrile illness one week prior to the onset of the lesions. The lesions were painless and asymptomatic. The patient had first noticed a lesion on the dorsal aspect of the right hand, at the mid of the ring finger. The lesions then further kept developing at various sites on the hands and feet.

There was no history of pain or discharge from the lesions. There was no association of the lesions with trauma. Upon further questioning the patient informed that there was a goat in the neighbourhood, who had a disease around the mouth and that the patient was the primary caretaker of it. There was no significant past medical or surgical history. Patient denied usage of any new drugs before the onset of the symptoms.

On clinical examination, the lesions were erythematous nodules with raised centre and crusting. Some of the lesions showed ulceration (Figure 2a & 2b). They measured between 2-6 cm in diameter. There was no associated lymph node involvement, no restriction of movement and an unremarkable systemic examination. The differential diagnosis we thought for this patient were, pyoderma gangrenosum, cutaneous anthrax, herpetic whitlow, atypical mycobacterial infection, cutaneous leishmaniasis and primary inoculation tuberculosis but considering contact history with diseased animal and clinical presentation we made Orf as provisional diagnosis. Skin biopsy from one of the lesions was done for further evaluation and confirmation. The histopathological findings were; ballooning degeneration of the keratinocytes with eosinophilic inclusion bodies, which is diagnostic of Parapoxvirus (Figure 3). Rest of the blood workup to rule out immunosuppression was normal, including HIV serology.

Figure 1a & 1b Nodules and plaques over the dorsal aspect of the hand. Seen in a sporotrichoid pattern.

Figure 2a & 2b Ulceration seen in the centre of the lesions.
The patient was given reassurance about the benign and self-limiting nature of the disease and given topical antibiotic and antiseptic to apply, with further counselling of practicing caution while handling infected cattle. The patient reported back after 8 weeks with complete resolution of the symptoms and clearance of the disease.

Discussion

The common presentation of Orf is a solitary nodule on either of the extremities, but a more widespread pattern of such lesions should not be overlooked. Disseminated orf, like this case, has been reported in the literature in a few places.\(^7\),\(^8\) Moreover, Orf with associated erythema multiforme or bullous pemphigoid like lesions have also been reported in the literature, but rarely.\(^5\) The immune response of the body to orf is thought to be responsible for the erythema multiforme like lesions. Christopher et al. described a 4-year-old thermal burn patient with Orf, after autologous skin grafting. The possible source of virus was attributed to the use of contaminated fomites in the hospital used for the dressing.\(^9\) Afia et al. reported from Karachi of 19 probable cases in immunocompromised patients of the viral infection in 5 different burn units.\(^10\) Patients with underlying skin diseases are also prone to developing widespread infection, such as reported in a 13-year-old boy who developed multiple, super infected lesions all over the body after being treated with acupuncture for his atopic eczema.\(^7\)

Our case was unusual in certain ways;

(i) Multiple lesions in Orf are not common,
(ii) Morphologically the lesions resembled cutaneous leishmaniasis and cutaneous tuberculosis,
(iii) Lesions on right hand and arm exhibited a sporotrichoid pattern.

Diagnosis was likely to be missed if history would not have been probed and atypical presentation in form of multiple lesions would not have been thought of. Therefore, awareness of unusual clinical patterns of known entities is important to avoid unnecessary interventions.

In conclusion, Orf is a not very commonly seen problem by dermatologists, but in areas of cattle rearing and low socioeconomic setups, a high index of suspicion should always be kept in mind in case of atypical, disseminated or multiple lesions. Effective vaccines must be developed in order to make the animals disease free and to prevent further spread.

References

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