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Langerhans Cell Histiocytosis on the Vulva: A Case Report and Review of the Literature

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Langerhans cell histiocytosis (LCH) of the female genital tract is rare. Only 20 cases of primary vulva LCH have been previously reported in the medical literature. In this report, we describe an additional case of LCH on the vulva. A 28-year-old Chinese woman presented with a two-year history of ulcerous lesions on the left vulva. No associated temperature, tiredness, or general malaise was observed. A diagnosis of LCH had been made by biopsy. Histological and immunohistochemical findings were characteristic of LCH. After two weeks of combined medical therapy, including interferon, prednisone, and methotrexate (MTX), the lesion started to cicatrize. Now, the patient was well throughout a 18-month follow up, showing no symptoms or signs of local recurrence or systemic spread. The occurrence of LCH on the vulva is very unusual. It is necessary to perform a biopsy on the lesion, rule out the possibility of multiorgan involvement. There are no standard treatment options for this rare disease. The most effective treatment options remain elusive. In our case, combined medical therapy was proved to be effective.

Key words: Langerhans cell histiocytosis (LCH); vulva; treatment

Langerhans cell histiocytosis (LCH), also known as Histiocytosis X, is characterized by an organ-specific infiltration of cells with many morphological features and immunohistochemical markers of Langerhans cells. Clinically, LCH ranges from self-healing lesions to a multisystem involvement with organ dysfunction resistant to current therapies. The lesions appear in multiple organs, for example in the bones, skin, and lungs. But genital LCH as the only manifestation of this disease is very unusual. Initially, Lane et al.[1] reported LCH in a 6-year-old child in 1939. We here report an additional case of LCH on the vulva.

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Case report

A 28-year-old woman, gravida 2, para 1, discovered a ulcerous lesion on her left vulva in 2009. She felt pruritus in the area sometimes, but had no other cutaneous lesions, and she had no associated temperature, tiredness, or general malaise. She was given a course of oral antivirus medicine in local hospital, but the lesions had continued to spread. Physical examination revealed a ulcerous lesion of 1.0 cm × 1.5 cm on the left vulva (Figure 1). Regional lymph nodes were not enlarged.

A diagnosis of LCH had be made in March 2011 after biopsy. Biopsy and histological findings revealed that the lesions consisted of diffuse infiltrates of Langerhans cells with indented or grooved nuclei, some of Langerhans cells showed karyokinesis. Various numbers of eosinophils, lymphocytes, and neutrophils were also present (Figure 2). Immunohistochemical stains revealed that histiocytic cells were strongly positive for S-100 protein, CD1a, CD68 and vimentin, negtive for HMB-45 (Figure 3). These results led to a diagnosis of LCH.

The tests for the extent of disease including bone marrow biopsy, and PET-CT of bone, thorax and abdomen, but no abnormalities were detected in any other area. According to these results, the patient was diagnosed with LCH involving the vulva only.

Because the patient refused surgical excision, we decided to treat her with combined therapy. The treatment was commenced from April 2011, including interferon, prednisone, and methotrexate (MTX). The lesion started to cicatrize after two weeks therapy. Now, the patient was well throughout a 18-month follow up, showing no symptoms or signs of local recurrence or systemic spread.

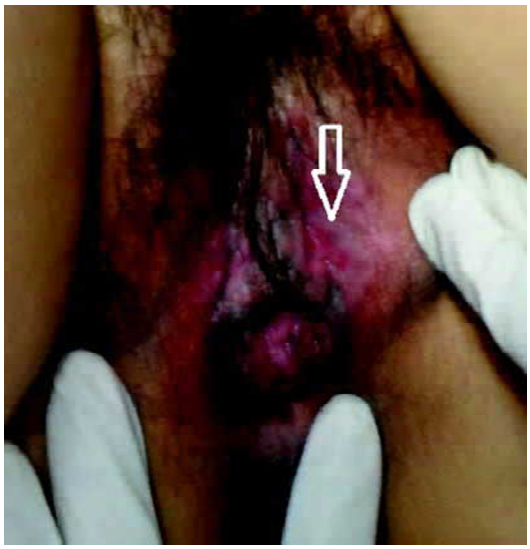

Figure 1 Ulcerous lesion on the left valva (↓)

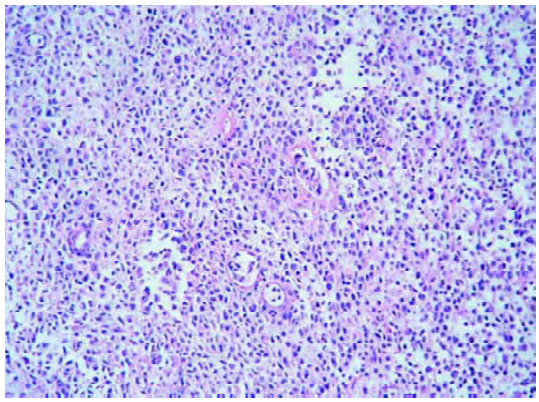

Figure 2 A biopsy specimen shows diffuse infiltrates of Langerhans cells and elevated number eosinophils, lymphocytes and neutrophils also can be seen (HE × 200)