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DERMATOLOGIC THERAPY WILEY

LETTER TO THE EDITOR

Adult Langerhans cell histiocytosis with recurrent vulvar ulcers

Dear Editor,

A 19-year-old female patient was referred to dermatology outpatient clinic with recurrent, erythematous papules and ulcers on her breast and sternum present for 8 months. She subsequently developed painless ulcers on the mons pubis and vulva. Physical examination was notable for erythematous papules of breast, sternum, and a 10mm ulcer of the mons pubis (Figure 1). There was no other skin abnormalities. Bilateral inguinal, cervical, axillary lymph nodes were unremarkable. Hepatomegaly and splenomegaly were not present. There was no additional past medical or surgical history.

Serologies viral infections, and bacterial, fungal cultures were normal. Intraoral examination was normal. Laboratory studies, including urine analysis, a complete blood cells count, erythrocyte sedimentation rate, and serum biochemistry were within the normal ranges. Abdominal ultrasound examination, a skeletal survey, and chest radiograph, and brain, thorax, abdominopelvic tomography were normal. Ear, nose, throat, eye, orthopedics examination were unremarkable. Pathergy test was normal.

FIGURE 1 Ulcers on the mons pubis and labium majus

Biopsies of the genital ulcer were consistent with Langerhans cell histiocytosis (LCH) suggesting a diagnosis of LCH with skin involvement (Figure 2).

This case points to the importance of LCH presenting as a genital ulcer. LCH is a complex, poorly understood, rare disorder characterized by the proliferation and accumulation of clonal dendritic cells called Langerhans cells.[1] Its incidence of 4 to 8 per million children and 1 to 2 cases per million in adults.[2] It is usually seen in children and young adults. The average age of onset is 1 to 3 years. The disease is observed more commonly in males.[2] Manifestations differ from single system to disseminated disease involving essentially any organ. Extra-skeletal involvement includes the skin, lymph nodes, lung, liver, spleen, bone marrow or central nervous system.[2] Although the potential for multisystem involvement may be seen, two thirds of patients have only single-system involvement, primarily of bones, skin, and lymph nodes.[1] Nearly, 25% to 50% of the LCH patients with skin involvement present with a skin lesion as the initial symptom.[3] The criteria for diagnosis of LCH require the demonstration of the presence of Birbeck granules by electron microscopy or CD1a antigen expression by immunohistochemistry.[2]

Generalized papules, nodules, localized scalp lesions or ulcers in skin folds can be observed in cutaneous LCH. The most common sites of cutaneous involvement are the scalp, flexural areas, external genitalia and glabrous skin.[4] The development of systemic findings may be also seen months to years after the initial presentation of skin manifestations.[4] The prognosis and treatment of LCH rely on the extent and severity of disease.[5] The skin-limited disease may heal

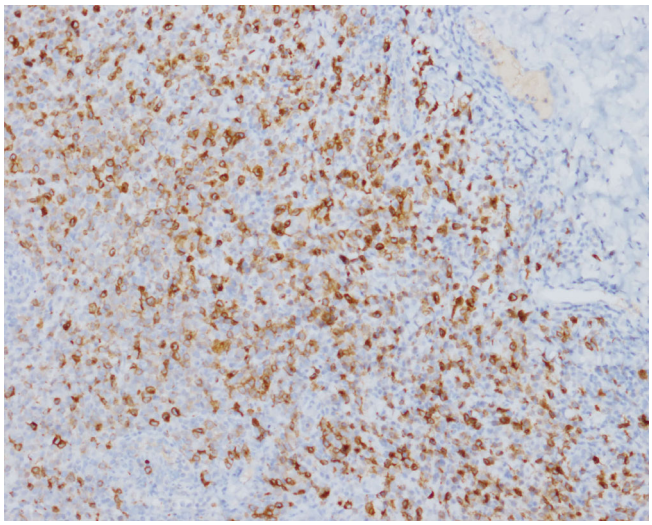

FIGURE 2 LANGERIN positivity in histiocytic cells

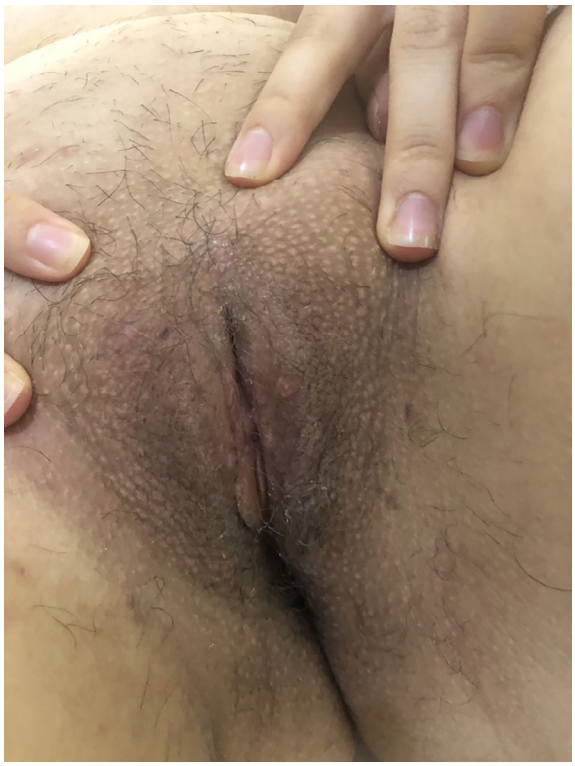

FIGURE 3 Scars on the mons pubis and labium majus

spontaneously but multisystem disease often requires chemotherapy because of the risk of mortality.[5]

Our patient has been on regular follow up for over 8 months. After diagnosis, we treated her with topical %0.05 clobetazol 17-propionate, twice in a day. We observed the lesion every month.

After 3 months beginning this treatment, complete improvement was seen and we are going on follow up patient (Figure 3).

We presented this case because it is rare and important to see LHH in an adult patient with recurrent ulcers and without systemic involvement.

CONFLICT OF INTEREST

The authors declare to potential conflict of interest.

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