

A LADY WITH RECURRENT PERIANAL ULCER: A CASE REPORT

Nawshirwan Gafoor Rashid ^a



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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare systemic disorder characterized by clonal proliferation of bone-marrow-derived Langerhans cells in various systems. Lesions on the genital tract are uncommon, with only a few reports of isolated vulvar LCH ⁽¹⁾. It is characterized by accumulating CD1a+/Langerin+ LCH cells and wide-ranging organ involvement ⁽²⁾. The presentation can vary from single organ involvement to disseminated, multi-system disease. Clinical aggressiveness is equally variable. In 1939 Andrews first described LCH of the female reproductive tract. Subsequently, four patterns of involvement have been identified: (a) pure genital LCH, (b) genital tract LCH with subsequent multi-organ involvement, (c) oral or cutaneous LCH with subsequent genital and multi-organ involvement, and (d) diabetes insipidus with organ involvement ⁽³⁾. here our case at the beginning she had only perianal skin involvement which was completely resected but after two years she relapsed with perianal & valvar ulcer, again totally resected and the result of histopathology & immunohistochemistry with positive CD1a, CD 207 & S100 confirms recurrence of the same pathology.

Keywords: *Recurrent ulcer, Perianal ulcer*

^a Hiwa Hemato-Oncology Hospital-Slemani-KRI-Iraq, Kurdistan Board of Medical specialities, Slemani Hematology Center.

Correspondence: rashid.nawshirwan1973@gmail.com

CASE PRESENTATION

A 42 years old lady who is a primary school teacher presented to Hiwa hospital in Slemani City-Kurdistan Region-Iraq in July 2019 with a perianal lesion for one month, associated with constipation, proctalgia, and hematochezia, not associated with weight loss, polyuria, polydipsia, headache or neurological problem or exophthalmos. Her pain was not aggravated by anything but relieved by analgesia. After one month of her condition referred to a surgeon, and total resection was done, and a biopsy was taken from the perianal lesion and diagnosed as Langerhans Cell Histiocytosis.

After a thorough workup including complete blood count, blood film, inflammatory markers, biochemical test, viral screen, CT scan of chest, abdomen & pelvis, bone marrow aspiration & biopsy all are normal; we put her on regular follow. However, unfortunately, she lost intentionally from our follow-up and returned after 26 months from her first presentation with pain and two small ulcers on her anal & vulvar region; we sent the patient again for a full workup, including colonoscopy, which was normal apart from a lower rectal polyp that was unrelated to anal & vaginal ulcer the biopsy was normal, this time PET scan & MRI of the brain were done to exclude distant spread and diabetes insipidus.

PET scan shows FDG avid soft tissue thickness noted in the upper anal canal with interval decrease in FDG uptake and size, with stable lung changes in the form of

ground glass haze, multiple sub-centimetre cysts and some interstitial lung reticulations.

No other FDG avid hypermetabolic lesions were seen in the rest of the PET scan, MRI of brain & pituitary gland were normal, concomitantly we sent for the tissue biopsy of the vulvar & anal lesions, the lesions removed completely surgically & the result of the biopsy & immunohistochemistry showed positivity for CD1a, CD68 & S100, which confirmed the diagnosis of Langerhans cell histiocytosis again.

After explaining the diagnosis again to the lady & asking her to not skip her treatment & her follow-up, unlike the first time, as she has multifocal involvement in one area, we started her on a combination of daily oral prednisolone, weekly methotrexate & local topical steroid with regular follow up. Biopsy from perianal & vulvar ulcer

Sections (A. & B.) show ulcerated skin lesions with lichenoid inflammatory infiltrates at the dermo-epidermal junction, composed of lymphocytes, neutrophils intermixed with bland appearing histocytes composed of large cells having eosinophilic cytoplasm with grooved nuclei mixed with large numbers of clusters of eosinophils.

C. Immunohistochemistry performed on both lesions with positive controls showed CD1a & S100 positive with moderate & diffuse cytoplasmic staining patterns.

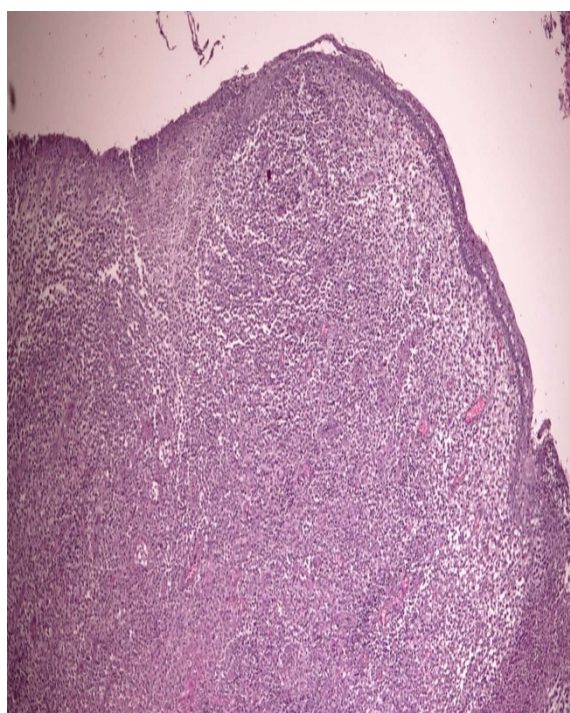


Figure A.

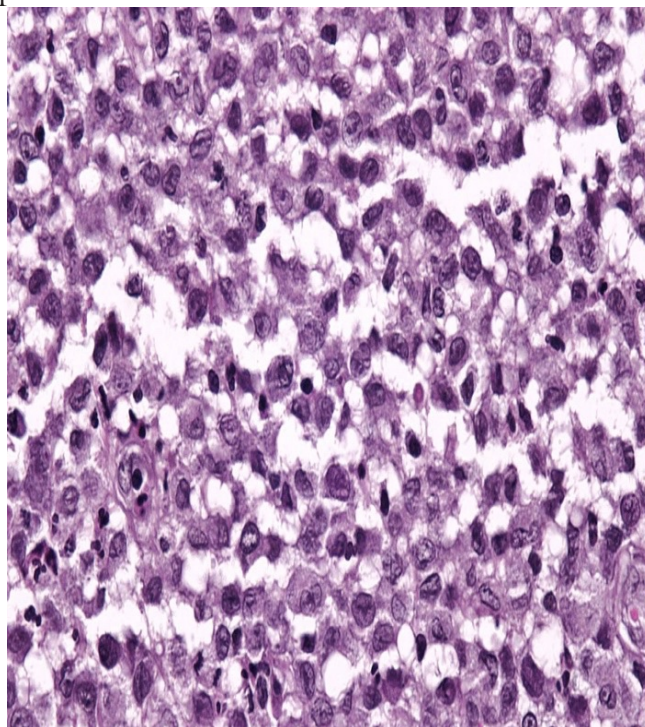


Figure B.

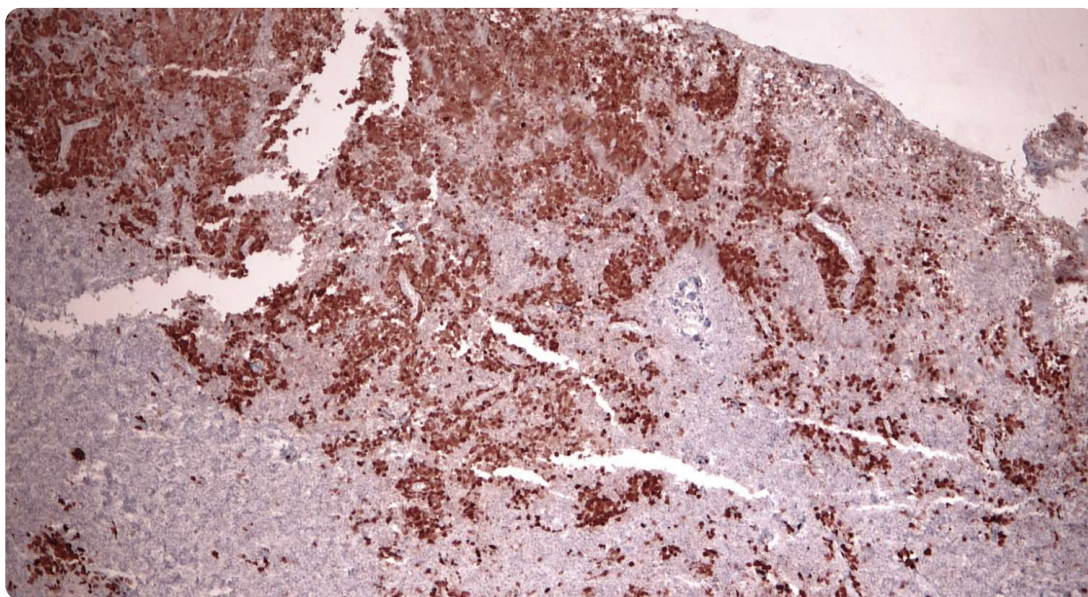


Figure C.
Biopsy from perianal & vulvar ulcer

DISCUSSION

Langerhans Cell Histiocytosis (LCH) can occur at any age, but mainly in children of 1~4-year-old. The incidence of LCH in adults is 1–2 cases per million. Most LCH patients are males. The sex ratio (M: F) was 2:1⁽⁴⁾. Pure LCH of the female genital tract is rare, with only 32 reported cases after a worldwide literature search on Pubmed and Medline restricted to the English language ⁽¹⁾. Here we report a 42 years old still menstruating woman who has a history of perianal ulcers and was completely resected without systemic involvement. However, she lost intentionally from follow-up for two years. When she came back, she had pain in the perianal region with two ulcers in the perineum & vulvar region; again, complete surgical resection & tissue biopsy was done and showed recurrent Langerhans cell histiocytosis with two focal involvements in the perianal region without multisystemic involvement. She was started on treatment according to NCCN guidelines with weekly oral methotrexate and daily oral & topical steroids with regular follow-up. Unfortunately, we do not have molecular studies for detecting BRAF V600 E or MAPK pathway gene mutation. Neither do we have new targeted therapies to start for first-line or subsequent lines of treatment.

We learned from this lady with Langerhans cell histiocytosis that although we warned this patient

that these skin lesions are a type of cancer & they can spread to other parts of the body & you may need systemic chemotherapy or radiotherapy in future & it could be fatal. However, the patient neglected all of these even though the patient is a primary school teacher. So, we thought that probably there were two reasons behind her noncompliance. The first one is that the disease at the start was only a single skin ulcer & removed. So, the patient thought that it would unlikely reappear again. The second reason she complains of financial difficulties is she lives about 150 Km far away from our centre, which makes her difficult to return for monitoring especially in the last two years as we went through a difficult time because of economic crisis & burden of recent COVID-19 pandemic on public life. So, raising the people's health education with better economic status will increase people's compliance with medical services.

We concluded that to increase patients' compliance with medical follow-up & treatment, we need to raise the standards of health education for non-medical people, and better financial life will help improve their compliance.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Consent

A formal written consent was obtained from this patient for sharing & publishing her medical information.

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