



MUCOSAL DISEASES (ORAL, ANOGENITAL), EXTERNAL EYE DISEASE

AN UNUSUAL RECURRENT ERUPTION OF PAINFUL VULVAL PAPULES

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Background: Langerhans cell histiocytosis (LCH) is a rare disease characterised by abnormal proliferation of Langerhans cells. Isolated LCH confined to the vulva is rare; only a handful of cases have been reported to date. It usually presents as persistent ulcers or erythematous plaques, and less commonly as nodules, papillomas or eczematous patches, often leading to a missed or delayed diagnosis. It is important to consider LCH as a diagnosis of chronic, atypical vulval lesions as it may be part of a multiorgan disease. The gold standard treatment remains elusive due to the scarcity of reported cases, but suggested regimens range from local surgery to chemotherapeutic agents and are not necessarily successful.

Observation: We report the case of a 46-year-old female who was first seen in 2010 with recurrent itchy and painful papules on the labia minora that had been treated empirically as herpes simplex virus infection for many years. Several papules would occur intermittently, clinically resembling sebaceous adenitis and subsequently healing with depressed scars. Repeated biopsies revealed macrophage-like cells staining strongly with antibodies to CD1a, S100 and Langerin, confirming LCH. Systemic investigations showed no evidence of disseminated disease and she has remained well for 8 years of follow-up. Treatment with long-term oral lymecycline reduced the recurrence of the painful papules.

Key message: Isolated vulval LCH is rare and can often be mistaken for other vulval diseases. We report an unusual presentation of this rare condition that clinically resembled sebaceous adenitis, which to our knowledge has not been reported previously. Chronic, atypical vulval lesions should be investigated and LCH should be considered as it has the potential for aggressive clinical behaviour, either as local recurrence or as disseminated disease. Antimicrobials with anti-inflammatory properties such as lymecycline may prove useful in treating other cases of vulval LCH presenting in a similar way.

