ADULT-ONSET LANGERHANS CELL HISTIOCYTOSIS MASQUERADING HIDRADENITIS SUPPURATIVA

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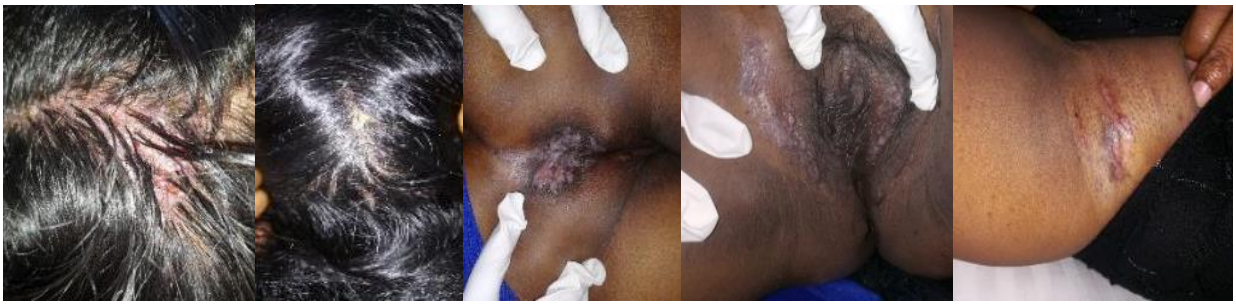
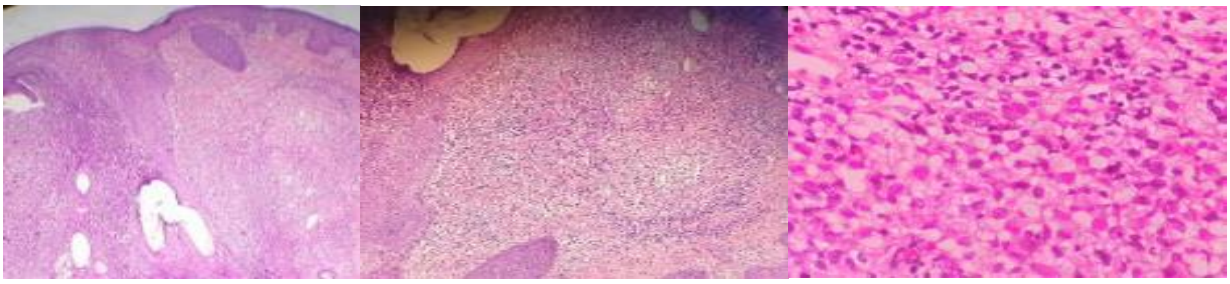
A 53 year old lady presented with recurrent erythematous papules, ulcerating nodules of the intertriginous areas and crusted plaques of the scalp for 3 years duration.

Figure 1: Erythematous plaques with crusted erosions and maceration involving scalp and intertriginous areas



She has been clinically diagnosed and treated for hidradenitis suppurativa and dissecting cellulitis of the scalp. Poor response to therapy prompted us to arrange histological evaluation (Figure 1). Extensive evaluation with biochemical, histological and radiological workup was arranged in order to out the systemic involvement. She was found to have complete cranial diabetes insipidus, but there were no liver, lung or bone marrow involvement. she was referred to oncologist, started on chemotherapy with a satisfactory response

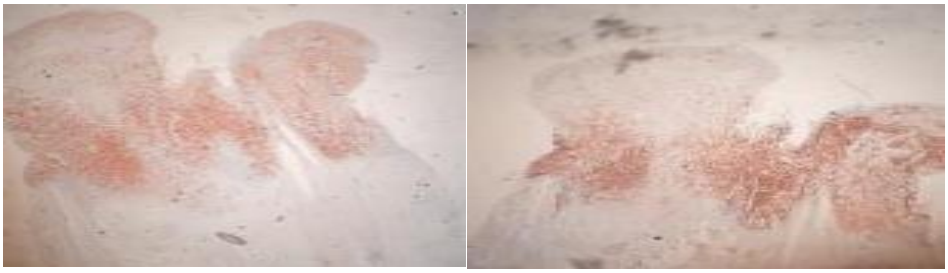
Figure 2: Skin biopsy findings compatible with Langerhans cell histiocytosis



Langerhans cell histiocytosis (LCH), is a clonal proliferative disorder of bone marrow-derived histiocytes with three sub types(1) (single-system unifocal, single-system multifocal, and multisystem disease). This is a rare disease, with unclear etiopathogenesis(2). This entity may

occur at any age, but mainly in children of 1~3-year-old. The incidence in adults is 1–2 cases per million(3).

Figure 3: Skin biopsy revealed dense dermal collections of histiocytes with S100 and CD1A positivity suggestive of adult onset LCH.



Simultaneous involvement of bones, lungs, skin, oral–genital mucosa, and endocrine glands are recognized(4). Treatments are based on organ involvement and extension of the disease and the prognosis is closely related to age of onset and number of involved organs(5). Though the clinical picture in our patient was not typical for hidradenitis, LCH was not considered as a differential diagnosis initially as it is rare among adult population(6). However, if we have arranged a skin biopsy sooner the diagnosis would have been made earlier in this case. Skin is the second most affected organ but uncommon nature of this entity leads to high rate of misdiagnosis and missed diagnosis, hence the correct clinical suspicion, histopathology and immunohistochemistry play a key role in diagnosis of LCH(7).

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