



The Role of Dermatologist in the Diagnosis of Systemic Langerhans Cell Histiocytosis in Adult Patient

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Langerhans cell histiocytosis (LCH) is a malign proliferation of dendritic cells which are able to infiltrate any organ and system. LCH could be restricted to a single organ. In adults, LCH is usually restricted to the lungs. Skin involvement of LCH in adults is rare. However, in pediatric group, skin is a one of the predominant organ which LCH involves solely or as a part of systemic disease. Pediatric cutaneous LCH demonstrates more clear clinical view compared to adult cutaneous LCH. Skin lesions in pediatric cutaneous LCH usually affects the seborrheic regions of the skin, therefore to recognize and diagnose cutaneous LCH in children is unchallenging compared to adult LCH.

In present article, a 33-year old patient with polydipsia and polyuria has been diagnosed as a central diabetes insipidus due to tumoral infiltration of sella turcica. The nature of the tumoral lesion has been revealed by histopathologic examination of single skin lesion which has been detected and sampled by dermatologist. In this case, diagnosis of adult type of LCH was challenging. However, detailed and total skin examination paved the way toward the correct diagnoses and avoided from the transsphenoidal intervention.

Since the skin biopsy is a low risk diagnostic tool, detailed skin examination should be implemented and unusual skin lesion should be excised for histopathologic examination in a challenging adult LCH cases.

Keywords: langerhans cell histiocytosis, adult, Hand-Schüller-Christian disease.

Introduction

Langerhans cell histiocytosis (LCH) is a proliferative disorder of dendritic cells with a capacity to infiltrate any organ and system. Three clinical subtypes of LCH were described up to now: localized, chronic disseminated and acute disseminated. In terms of organ involvement, LCH is classified as: single system (SS) and multisystem (MS). SS LCH could be unifocal and multifocal. Its equivalent to eosinophilic granuloma, localized form of LCH. MS LCH without risk organ (RO) is considered as a chronic disseminated (Hand-Schüller-Christian disease), however, MS LCH with RO are equivalent to acute disseminated form (Litter-Siwe disease) [1]. Bone and skin involvement is more common for SS in children, whereas lung infiltration of LCH is more common

in adults [2,3]. Skin as a SS involvement of LCH is rare in adults [2,3]. According to International Histiocyte Society Registry, skin infiltration of LCH as a part of MS occurs frequently in both, adults and children [2,3]. However, German registry demonstrates that skin involvement of LCH in adults is not common (17%) [4]. In children, LCH is usually limited to the seborrheic areas. But skin lesions of LCH in adult patients are not well defined [5]. So, diagnostic value of dermatologist in pediatric LCH cases is more prominent than in adulthood. Here, we report a case of LCH with hypophysis infiltration that was diagnosed by single skin lesion.

Case Presentation

A 33-year-old male patient referred to

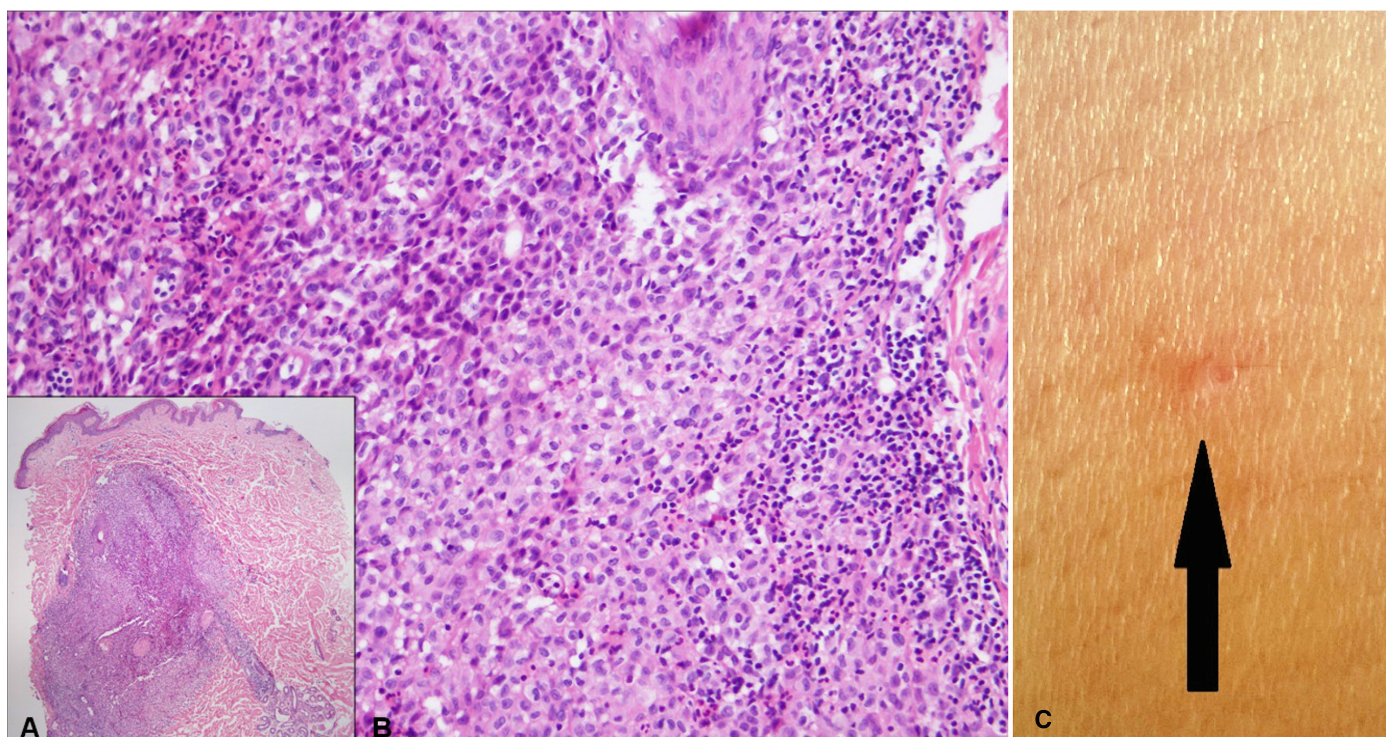


Figure 1. **A.** Nodular infiltration in the middle and deep dermis H&E x 40. **B.** Large, eosinophilic cytoplasm of Langerhans cells is evident H&E x 200. **C.** Single pink papule at the proximity of axillary fold.

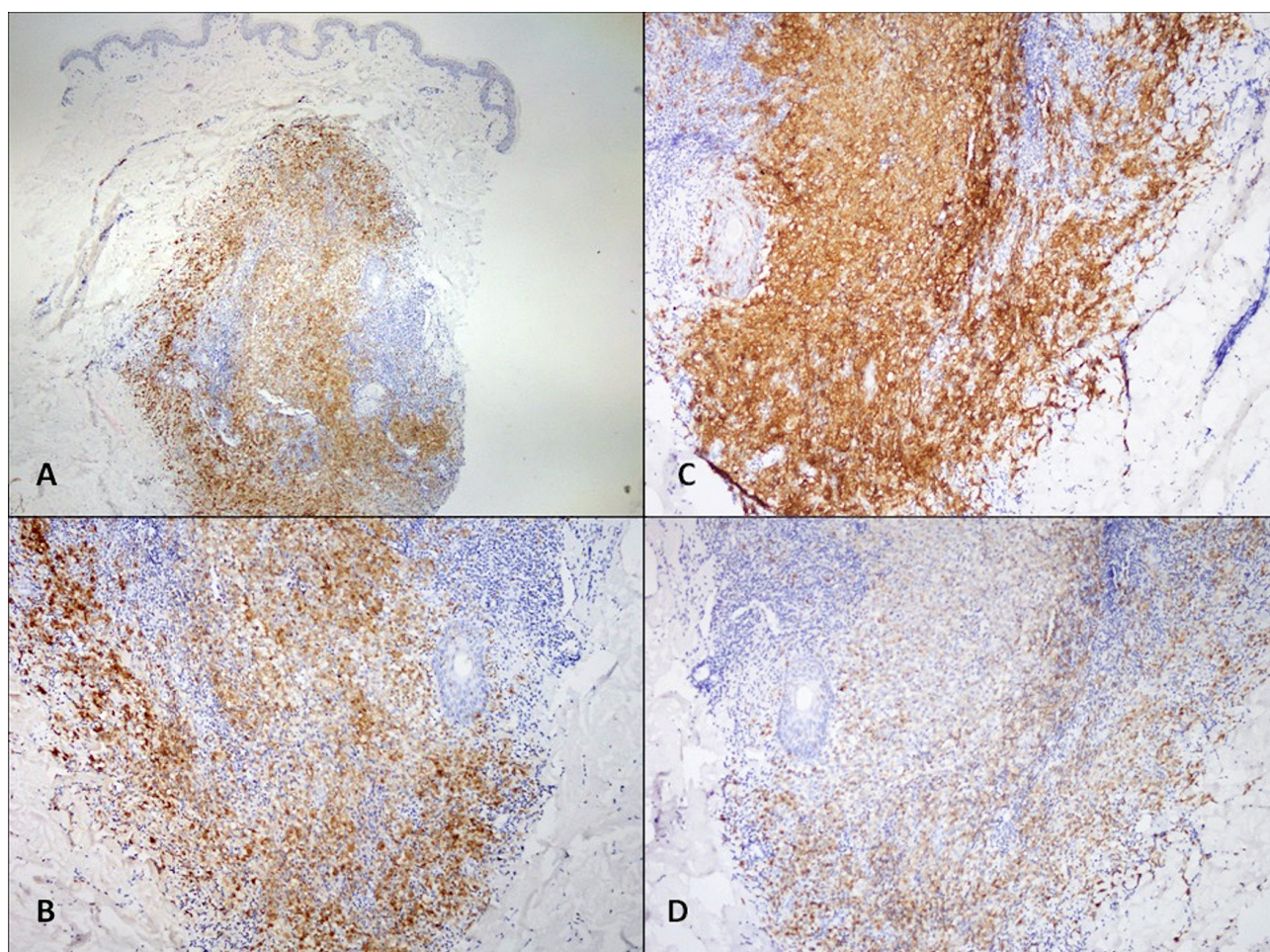


Figure 2. **A.** S100 positivity x 40. **B, C.** CD1 positivity x 100. **D.** Langerin positivity x 100.

dermatology department from endocrinology department to evaluate the patient in terms of skin involvement of LCH. He was hospitalized in endocrinology department due to polydipsia and polyuria that lasted for 6 month. After series examinations, patient was diagnosed with a central diabetes insipidus. Pituitary MRI revealed tumoral infiltration of sella turcica of unknown origin. LCH and sarcoidosis were the preliminary diagnosis. Systemic evaluation of the patient in terms of LCH and sarcoidosis revealed negative results. So, patient was referred to dermatology department for diagnostic purposes. Patient was thoroughly evaluated in terms of sarcoidosis and LCH. Single pink papula near the axillar region was found (Figure 1). "Apple jelly" sign was negative. It didn't look like folliculitis, hemangioma or melanocytic nevus. Seborrheic localization and atypical view of the lesion gave an idea to think about cutaneous LCH. The lesion was excised and histopathologically evaluated (Figure 1, 2). The nature of the hypophyseal infiltration was diagnosed by single LCH skin lesion in this patient. Ophthalmologic examination revealed minimal exophthalmos of the left eye. Macroscopic exophthalmos was absent. Lytic lesions were absent on bone survey. Lung and visceral organ involvement of LCH was negative.

Discussion

The role of dermatologist in the diagnosis of systemic LCH is very crucial, especially in adult cases. Therefore, in these group of patients, total skin examination should be performed thoroughly. Intertriginous regions should be carefully evaluated since LCH is prone to infiltrate seborrheic areas of the skin. Any atypical skin lesion should be sampled for histopathologic (HP) evaluation. In this case, systemic LCH was confirmed by single papula which was located at the proximity of seborrheic area. Because of detailed skin examination and HP evaluation patient escaped from invasive transsphenoidal intervention. Moreover, patient evaluated also in terms of chronic disseminated form of LCH, Hand-Schüller-Christian disease (HSCD). HSCD is more common in pediatric group. Exophthalmos, lytic bone lesions usually on skull, and diabetes insipidus due to pituitary stalk infiltration by LCH are classic triad of HSCD. However, classic triad presents only 25% of cases.[6] Probably, this case is an incomplete HSCD of adulthood which demonstrates skin involvement additionally.

Conclusion

In conclusion, total skin examination should be implemented thoroughly, in adults with systemic LCH suspicion. Skin lesions with an atypical view should be sampled for HP evaluation. Moreover, invasive diagnostic procedures such as transsphenoidal intervention bypassed in this patient due to skin biopsy technique which is a low risk diagnostic method. By presenting this article, it is emphasized once more that, the role of the dermatologists in the diagnosis of systemic LCH might be crucial.

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