

# Langerhans cell sarcoma of the skin in association with superficial atypical Langerhans cell proliferation

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## Abstract

Langerhans cell sarcoma of the skin is a rare tumor with aggressive behavior. There are reports of Langerhans cell sarcoma involving the skin in patients with underlying systemic Langerhans cell histiocytosis. However, to our knowledge, sarcomatous transformation of skin Langerhans cell histiocytosis, has not been previously described. We report a case of Langerhans cell sarcoma of the skin representing a probable transformation from Langerhans cell histiocytosis. The lesion had a clinical and pathological biphasic pattern, presenting as a plaque and nodule in the lower leg of a 66 year-old female. The plaque area showed a superficial epidermotropic proliferation of atypical Langerhans cells. The associated deep nodule had an overtly sarcomatous Langerhans cell proliferation. The immunohistochemistry profile was characteristic of Langerhans cells in both components. We consider this case a unique example of deep nodular Langerhans cell sarcoma of the skin, in which the associated superficial Langerhans cell proliferation is consistent with the precursor lesion.

## Introduction

Langerhans cell sarcoma was first described as a multisystemic aggressive process termed malignant histiocytosis X.<sup>1,2</sup> Few cases of primary cutaneous Langerhans cell sarcoma have been reported.<sup>3</sup> Unlike other neoplastic processes, the association of a precursor lesion and transformation into a overtly malignant process has been elusive in Langerhans cell sarcoma. A case of pulmonary malignant Langerhans cell transformation in the lung,<sup>4</sup> and association of Langerhans cell sarcoma with systemic histiocytosis X have been reported.<sup>5</sup> We report here a case of deep nodular Langerhans cell sarcoma arising in

association with a superficial plaque of Langerhans cell proliferation, consistent with the precursor lesion.

## Case Report

The patient is a 66 year old female, who presented with a rapidly growing 3×1.5 cm reddish plaque on the anterior distal left lower leg. Beneath the distal portion of the plaque there was a 1 cm hard, deep and relatively well circumscribed, spherical nodule (Figure 1). Separate biopsies were obtained from both the plaque area and from the deep nodule. Histological evaluation of the biopsy from the plaque showed a superficial proliferation of atypical mononuclear cells with epidermotropism (Figure 2A). Melanocytic, B-cell lymphocytic, and epithelial markers were negative. The cells in both the superficial and deep component had the characteristic features described in proliferations of Langerhans cells,<sup>6</sup> but exhibited cytologic atypia. The cells from the deep nodule exhibited marked atypia, areas of necrosis and abnormal mitoses (Figure 2B-D), as has been described in overtly malignant Langerhans cell tumors.<sup>7</sup> Immunohistochemistry studies performed in both biopsies showed markers typical of Langerhans cells, including expression of CD1a (Figure 3A,B), S-100, CD68, CD4 and CD45. CD30 is expressed in both the superficial and deep components (Figure 3C,D), with the highest expression in the superficial component. CD3 is expressed in both the superficial (Figure 4A) and the deep component (Figure 4B). CD207 (Langerin) is expressed in the superficial component (Figure 4C) but not in the deep component (Figure 4D). Beta and Gamma T-cell receptor monoclonality was not detected by PCR in neither the superficial or deep components. No BRAF gene mutation was detected by PCR, which was performed on both specimens.

## Materials and Methods

The biopsies were processed by routine tissue histology methods. Immunohistochemistry was performed at Miraca Life Sciences with appropriate controls using a Benchmark Ultra Ventana automated stainer, Ventana Medical Systems, Inc., Tucson, AZ. The antibodies were from Cell Marque, Rocklin, CA., and Ventana Medical Systems, Inc., Tucson, AZ. Immunohistochemistry for Langerin was performed at Associated Regional and University Pathologists, Inc. (ARUP), Salt Lake City, UT. Beta and Gamma T-cell receptor clonality and BRAF gene mutation

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analysis was performed by PCR on both specimens at Miraca Life Sciences, Phoenix, AZ.

## Discussion and Conclusions

Langerhans cell sarcoma may arise in the skin as a primary cutaneous neoplasm,<sup>3,8</sup> or represent a cutaneous manifestation of a systemic malignant process.<sup>9-11</sup> In either condition, understanding the process of malignant transformation in Langerhans cell neoplasms has proven difficult, and few cases have reported a bona fide precursor lesion.<sup>4</sup> In addition, the neoplastic transformation of Langerhans cells is even more complex given the potential of different cell progenies to acquire phenotypic and immunologic features of Langerhans cells by mechanisms known as clonal plasticity and transdifferentiation.<sup>12-15</sup> The process of clonal plasticity and transdifferentiation may explain the expression of CD30 and CD3 in our case. However, there are lesions described as resembling Langerhans cell histiocytosis, some of which may be neoplastic or reactive, in association with lym-