

## Case Report

# Case report of primary cutaneous anaplastic large cell lymphoma of adult female patient visiting dermatology clinic of King Abdul-Aziz Medical City, National Guard Health Affairs, Jeddah, Saudi Arabia, 2018

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### ABSTRACT

Primary cutaneous anaplastic large cell lymphoma (PCALCL) is a rare T-cell cutaneous lymphoma consisting of CD30 positive typically anaplastic-appearing cells. PCALCL had been distinguished from systemic anaplastic large cell lymphoma (ALCL) and lymphomatous papulosis by World Health Organization classification of lymphoid neoplasms. Previous studies have shown PCALCL has an excellent overall survival with few patients dying from disease. This is a case report study design with detailed history, examination of 54-years-old female patient with a painful nodular lesion in the left thigh, which is misdiagnosed as Pyoderma Gangrenosum.

**Keywords:** Primary cutaneous anaplastic large cell lymphoma, Pyoderma gangrenosum, Lymphoma, Thigh

### INTRODUCTION

Primary cutaneous anaplastic large cell lymphoma (PCALCL) belongs to the spectrum of primary cutaneous CD30+ lymphoproliferative disorders.<sup>1</sup> It is considered a subtype of cutaneous T-cell lymphoma (CTCL) and non-Hodgkin lymphoma (cancer of the white blood cells) arises in the skin.<sup>2</sup> Extra cutaneous dissemination may occur, especially to regional lymph nodes. PCALCL is named from the description of the size and shape of the cells under the microscope (“anaplastic large cell”), and is characterized by the presence of a molecule on the lymphoma cells called CD30.<sup>2,3</sup>

The prevalence of Primary cutaneous anaplastic large cell lymphoma (PCALCL) is unknown but it accounts for approximately 9% of cutaneous lymphomas. The

male/female ratio is of 3:2.<sup>3</sup> It generally occurs in adults and rarely in children and adolescents.<sup>3</sup>

Most patients with PCALCL present with large solitary or multiple slow-growing erythematous skin plaques, nodules or tumors that usually develop in either a localized or multifocal distribution.<sup>3,4</sup> These lesions may ulcerate and/or itch.

In 10% of cases, PCALCL extends beyond the skin to lymph nodes and manifests as a painless swelling, especially in the neck, armpit or groin. It can also extend to extranodal sites.<sup>4</sup> General symptoms, usually present in cases with extracutaneous involvement, include loss of appetite, weight loss, fatigue and night sweats.<sup>5</sup>

Radiation therapy, removal of the lesion and/or low-dose methotrexate are the preferred treatments among patients with localized lesions.<sup>3</sup> Rapidly progressive or extra cutaneous disease should be treated with systemic polychemotherapy.<sup>4</sup>

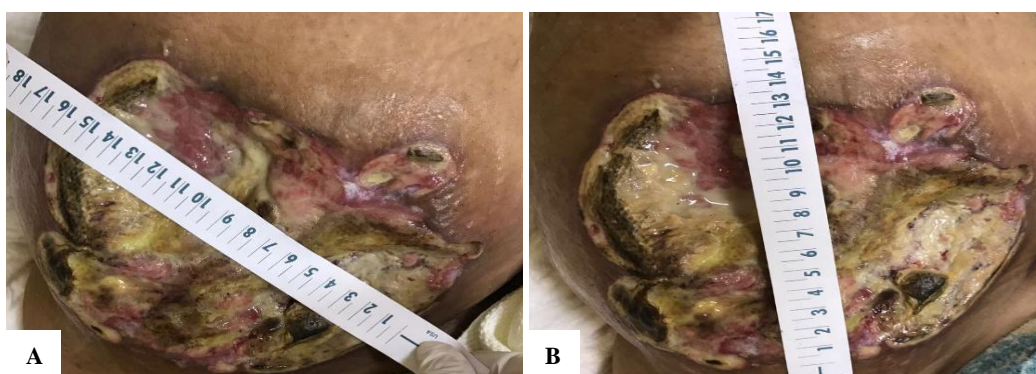
Low clinical suspicion of PCALCL may contribute to the delay of diagnosis especially for female patient other than male and patients with inflammatory bowel disease who usually have Pyoderma Gangrenosum with similar clinical features of PCALCL.<sup>4</sup>

This is a case report study design with detailed history, examination of 54-years-old female patient with a painful nodular lesion in the left thigh, which is misdiagnosed as Pyoderma Gangrenosum.

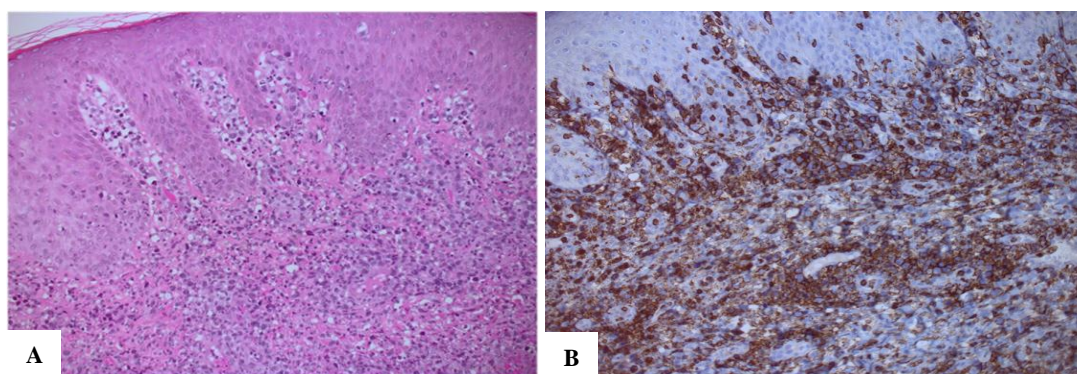
## CASE REPORT

54-year-old female came to the dermatology clinic, known case of chronic inflammatory bowel disease (IBD) on mesalazine and cyclosporine. She was complaining of skin ulcer on the left thigh. This skin ulcer started as a painful small nodule then for nine months its size progressed to form a well-defined ulcer.

Patient has regular follow up with gastroenterology clinic as a case of inflammatory bowel disease (IBD), when she was started to complain of skin lesion in her left thigh, physicians there diagnosed her as Pyoderma Gangrenosum. After that, her skin lesion got worse and the gastroenterology physician sent consultation request to dermatology department for kind care and further assessment.



**Figure 1 (A and B): Solitary well-defined ulcer over the left lateral thigh with violaceous border, undermined edges and necrotic center measuring 10×5 cm.**



**Figure 2 (A and B): Sections show Extensive infiltration of the dermis with neoplastic lymphoid cells. Right section showed Immunohistochemistry stain with target cells: CD30+, CD3+, LCA+, ki67 95%, CD20-, CD56-, CD10-, ALK-, CK-, BCL12-, BCL16-, and PAX5.**

Patient had seen by dermatology clinic (Figure 1) and examination of the skin revealed solitary well-defined ulcer over the left lateral thigh with violaceous border, undermined edges and necrotic center measuring 10×5 cm. In addition to that, she had also multiple post inflammatory hyperpigmentation on the lower limbs, thick crustations and a solitary erythematous non-scaly subcutaneous nodule on the medial side of the left leg.

Examination also showed clear oral mucosa and genital mucosa.

Histopathology showed (Figure 2) extensive infiltration of the dermis with neoplastic lymphoid cells having pleomorphic vesicular nuclei with nucleoli with frequent mitosis and extensive necrosis with no epidermotropism. The Immunohistochemical stain showed target cells including CD30+, CD3+, LCA+, ki67 95%, CD20-,

CD56-, CD10-, ALK-, CK-, BCL12-, BCL16- and PAX5. The results indicated large anaplastic T-cell lymphoma.

Patient had referral to oncology department and she was started on (CHOP) regimen, which consisted of cyclophosphamide, doxorubicin, vincristine, and prednisolone. After first cycle of chemotherapy there was

a significant reduction in the size of the lesion. However, her skin lesions had improved (Figure 3) but she found to have progressive diseases (metastasis to the oral mucosa) based on PET/ CT scan routine follow up after 4 months of end the cycle of chemotherapy.



**Figure 3 (A and B): Improvement in lesion after cycles of chemotherapy.**

## DISCUSSION

Primary cutaneous CD30+ lymphoproliferative disorders of T-cell origin, consist of lymphomatoid papulosis, and CD30+ anaplastic large cell lymphoma.<sup>6</sup> PCALCL affects older age group with median age of 55 years. Male-to-female ratio is 1.5:1.<sup>7</sup>

It commonly affects the trunk, extremities, and presents as solitary or multiple localized nodules with ulceration. Clinically, it resembles other dermatological diseases like eczema, pyoderma gangrenosum, pyogenic granuloma, morphea, and squamous cell carcinoma.<sup>8</sup>

Cutaneous CD30+ anaplastic large cell lymphoma (ALCL) is characterized by presence of CD30 expression in more than 75% of neoplastic cells.<sup>3</sup> Presence of cytogenetic abnormality, that is, nonrandom  $t(2; 5)(p23; q35)$  chromosomal translocation which involves fusion of anaplastic lymphoma kinase (ALK) with the nucleophosmin (NPM) gene is usually seen in systemic ALCL but only rarely in PCALCL.<sup>9</sup>

PCALCL usually presents as a solitary papulonodular lesion, which sometimes shows ulceration.<sup>4</sup> Our case presented with a large pyogenic granuloma-like lesion. Cheng *et al.* reported a case of systemic ALCL with local cutaneous involvement in the form of nonhealing pyogenic granuloma-like lesion in the right axillary area.<sup>9</sup> However, in our patient, systemic involvement was absent. Bains *et al* reported 35-year-old man with large pyogenic granuloma like lesion and regional lymphadenopathy at face, over the chin which is rare location other than the site of lesion in our case.<sup>4</sup>

A five-year survival rate of PCALCL is more than 90% but cutaneous disease arising from systemic ALCL has worse prognosis with five-year survival rate of less than 45%. ALK expression in cutaneous lesions usually indicates systemic disease but rarely, it can be positive in PCALCL. Also negative expression of ALK does not rule out systemic disease as 20%–60% of systemic ALCL can be ALK negative.<sup>10</sup> Twenty-five percent of the cases of primary cutaneous anaplastic CD30+ large T-cell lymphoma can also remit spontaneously.<sup>11</sup>

## CONCLUSION

ALCL can have varied cutaneous presentations. Being uncommon may lead to low clinical suspicion of PCALCL and the delay of diagnosis. As the cutaneous lesions can be the only manifestation in a case of systemic ALCL, clinicians should keep a high suspicion so that the disease can be detected at an early stage.

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