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REVIEW ARTICLE

A CASE OF PRIMARY CUTANEOUS ANAPLASTIC LARGE T-CELL LYMPHOMA PRESENTING AS LARGE ABDOMINAL MASS

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ABSTRACT

Primary cutaneous anaplastic large cell lymphoma is a small subset of anaplastic large cell lymphoma. It can only be differentiated from the other subtypes by exclusion of extracutaneous involvement and correlation of the clinical, histological, and immunohistochemical features. This case presentation describes a 76 year-old man with an abdominal mass larger than 5 centimeters that was found to be primary cutaneous anaplastic large cell lymphoma.

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INTRODUCTION

Anaplastic large cell lymphoma (ALCL) is a category of large cell lymphomas which are defined by their expression of CD30, characteristic pattern of tumor cell growth around sinusoids, and an anaplastic cytology. Cells are typically large lymphoid cells with abundant cytoplasm, little chromatin, a horseshoes-shaped nuclei with multiple nucleoi. Morphologic features, immunophenotype, and presentation are used to categorize ALCL subtypes (Stein, 2000) One subtype is cutaneous T-cell lymphoma, a heterogeneous group of non-Hodgkin lymphomas that are defined by clonal proliferation of malignant T-lymphocytes of the skin. 30% of these are primary cutaneous anaplastic large cell lymphoma (pcALCL) (Rosen, 2006). It is usually diagnosed in the sixth decade although it is also seen in children and adolescents (Geller, 2018). Primary cutaneous ALCL generally arises de novo as a solitary cutaneous or subcutaneous violet-colored lesion that may have ulceration (Stein, 2000) Excellent prognosis is achieved with local excision with or without radiation with a five-year overall survival (OS) of 90% (Stein, 2000; Savage, 2000). Spontaneous regression occurs in about 25% of cases although recurrences are experienced by up to 40% of treated patients. (Stein, 2000; Kempf, 2011) Progression of pcALCL to a systemic disease occurs in only 12-16% of cases and greater risk of this progression seems to correlate to those with disseminated cutaneous involvement (Kempf, 2015) These patients may benefit from systemic chemotherapy (Stein, 2000)

Case Presentation: A 76 year-old Caucasian man presented to a dermatologist office for initial evaluation of asymptomatic skin lesions that he stated had been present "for years." Review of systems was negative. Physical examination revealed various different pathologies on the patient's face and trunk including solar elastosis in sun-exposed distribution on the trunk, lentigines and irritated seborrheic keratosis on the forehead and in multiple areas of the trunk, and a 4 centimeter (cm) nodule on the neckas well asno palpable lymphadenopathy. The solar elastosis and the neck nodule did not require treatment. The dermatologist used liquid nitrogen for the lentigines and irritated seborrheic keratosis.Additionally, a 3 cm subcutaneous cyst with

prominent follicular pore was located on the right lateral abdomen which was locally excised. The patient was given prophylactic antibiotics to prevent infection after the excision. The tumor was found to be located in the subfascial plane and measured 5.2 cm. Dermatopathology report described the cells as having scant cytoplasm and large round to slightly irregular nuclei with granular chromatin and multiple nucleoli. Numerous apoptotic bodies and mitotic figures were present. The sample was positive for LCA, CD-3, CD-30, and MUM1 and had focal weak staining for CD-15.(Figure 1) One-hundred percent of the cells stained for Ki67. The pathologist suggested a diagnosis of anaplastic large T-cell lymphoma, a cutaneous anaplastic large cell lymphoma, or unspecified peripheral T-cell lymphoma. A bone marrow biopsy was performed by the consulted oncologist to determine a final diagnosis which showed no evidence of lymphoproliferation on histology and no chromosomal abnormalities were found on chromosome analysis. Positron Emission Tomography- Computed Tomography (PET-CT) Scan from skull base to mid-thigh was performed for complete staging and excluded malignant lymphomatous involvement. The patient was provided with a definitive diagnosis of cutaneous anaplastic large cell lymphoma and informed that it may recur. No further treatment was required or administered but the patient was scheduled for continued surveillance.

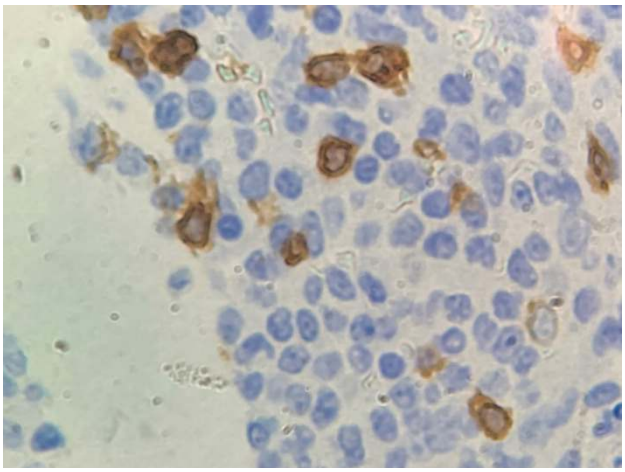

Figure 1a. Immunohistochemical staining of the biopsy sample for CD-3.

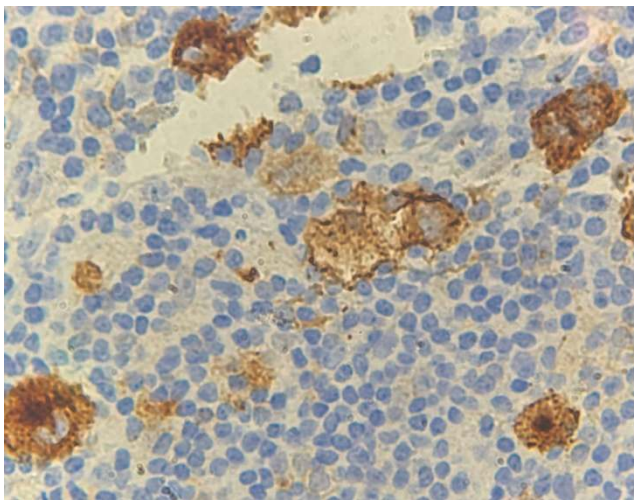

Figure 1b. Immunohistochemical staining of the biopsy sample for CD-30

DISCUSSION

In this case, our patient sought evaluation by dermatology for several different types of asymptomatic skin lesions which were present for years. All of these lesions were diagnosed and treated with either cryotherapy or observation,except one located on the abdomen which was suspected to be melanoma. The abdominal mass was biopsied and pathology revealed an anaplastic large cell lymphoma. Consultation to oncology resulted in exclusion of the systemic subtype by bone marrow biopsy analysis, whole body PET-CT, and immunohistochemical staining of tissue from the abdominal mass. Skin manifestations of cutaneous lymphoma often resemble common skin diseases and further evaluation may be initiated after failed treatment with standard therapies. Although pcACLC has a good prognosis, it is important to confirm the diagnosis as some cases can progress to systemic disease which portends a worse outcome (Zic, 2021). Primary cutaneous anaplastic large cell lymphoma can also present dermatologically similar to the more aggressive systemic anaplastic large cell lymphoma. These two diseases also share cytomorphologic features which can make diagnosis challenging. Consideration of clinical presentation, histology, and immunohistochemical features are necessary to differentiate the subtypes (Chen, 2019). Primary cutaneous ALCL is morphologically and immunophenotypically identical to systemic ALCLwith cutaneous involvement but systemic ALCL has a different clinical course and worse prognosis (Pina-Oviedo, 2021). In order to ensure an accurate diagnosis and adequate treatment, systemic ALCL should be excluded with whole body PET-CT (7).

CONCLUSION

Primary cutaneous ALCL is a rare subtype of ALCL which is defined as a CD30 positive clonal proliferation of skin-homing T-cells. Characteristic morphologic features include large atypical lymphocytes containing abundant cytoplasm, little chromatin, and horseshoe-shaped nucleus with multiple nucleoli. Because of its similarity to the other subtypes and its relatively better prognosis, it is important to distinguish pcALCL from other, more common pathologies.

Patient Consent: Written consent for this case report and its associated imageswas obtained from the patient.

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