Primary Cutaneous Anaplastic Large Cell Lymphoma Presenting With Corkscrew Vessels On Dermoscopy

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Introduction

Corkscrew (linear helical) blood vessels on dermoscopy have been described as features of melanoma and cutaneous metastases[1]. The etiology of these vessels is unknown but the association with high-risk neoplasms is ominous and may be related to features unique to cancers with a high metastatic potential. We present a case of primary cutaneous anaplastic large cell lymphoma (ALCL) with prominent corkscrew blood vessels on dermoscopy.

Case Presentation

An 86-year-old man with a past medical history of basal cell carcinoma and prostate adenocarcinoma presented to clinic for a routine skin cancer screening. Upon exam, a 0.8x0.5-centimeter (cm) tan pink papule with minimal overlying scale was observed on his left posterolateral foot (Figure 1). Dermoscopy revealed a tan pink background with central grouped corkscrew (linear helical) vessels and a small amount of overlying white/yellow scale (Figure 2).

Histopathologic evaluation (Figures 3a and 3b) revealed a dermal lymphoid proliferation include a population of large atypical cells with prominent nucleoli and variable amounts of amphophilic cytoplasm. Multinucleate forms and occasional horseshoe-shaped forms were present and mitoses were conspicuous. There was a mixed inflammatory infiltrate.

Immunohistochemistry showed large cells staining positively for CD30 (Figure 3c), TIA-1, CD4 (Figure 3d), and CD2. These cells were negative for ALK1, nuclear PAX-5, CD20, CD3, CD5, CD7, CD8, CD10, P40, AE1/AE3, PSA, Mart1, and S100. The histologic diagnosis of a CD30 positive lymphoproliferative disorder was made with the differential diagnosis of lymphomatoid papulosis (LyP), primary cutaneous anaplastic large cell lymphoma (ALCL), or systemic ALCL with cutaneous involvement.

The patient was referred to hematology and oncology where he underwent an extensive work up to include imaging and a bone marrow biopsy with no evidence of systemic disease. This negative workup and the lack of other similar lesions argued against systemic disease and LyP, and the diagnosis of primary cutaneous anaplastic large cell lymphoma was made. The patient was treated with radiation therapy and has had no evidence of recurrence or new lesions on subsequent exams.

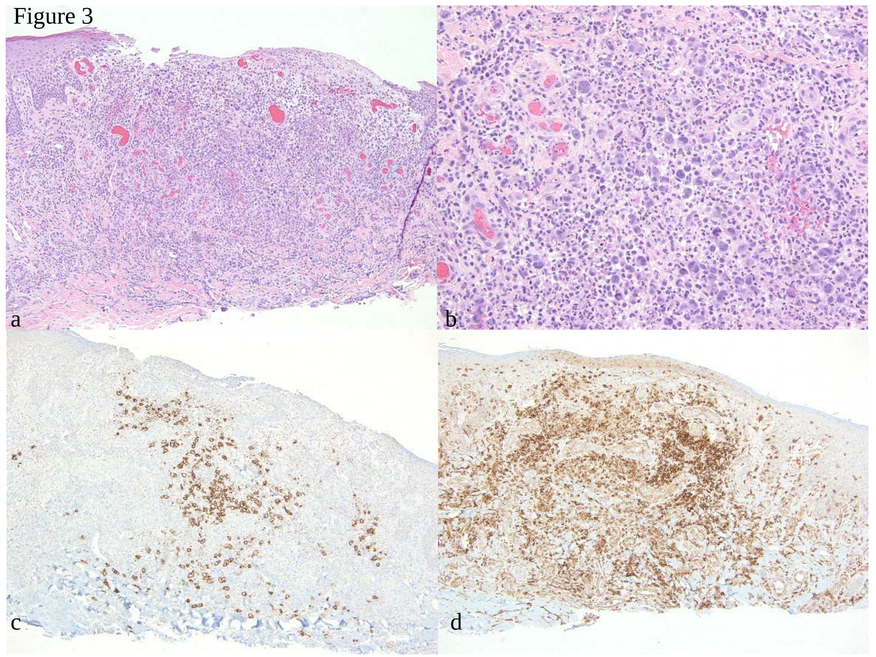
Clinical and Dermatoscopic Images

Figure 1

Figure 2

Histology

Figure 3



Discussion

This is the first time that large tortuous, corkscrew, or linear helical blood vessels have been described in the context of ALCL. They have been noted in the context of nodular melanoma, desmoplastic melanoma, and cutaneous melanoma metastasis[2]. It is unclear why these vessels form but they appear to have a predilection for cancers that have either already metastasized or have a high rate of metastasis. Thus this finding may be related to features associated with tumor invasion. Arterial tortuosity has been associated with reduced axial tension or elongation of arteries[3]. Additionally, weakening of the arterial wall and degradation of surrounding connective tissue, which occur in rapidly growing malignancies have been shown to induce a corkscrew shape in the vessel[3]. On a molecular level, alterations in matrix metalloproteinase (MMP) expression have been shown to alter normal angiogenesis[4] and contribute to the progression of melanoma[5]. This represents a possible mechanistic connection between progressing neoplasms and the presence of corkscrew blood vessels on dermoscopy.

The discovery of this new vascular pattern in an ALCL serves to introduce this entity as a member of the differential diagnosis for linear helical or corkscrew shaped vessels on dermoscopy. Additionally, the possible connection between this dermatoscopic vascular pattern and abnormal MMP expression suggests a possible clinical clue to an increased risk of metastasis. In our case the recognition of these concerning vessels served to ensure that this otherwise benign appearing lesion was biopsied.

References

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