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A case of primary cutaneous anaplastic large cell lymphoma presenting with corkscrew vessels on dermoscopy

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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.

An 86-year-old man with a past medical history of basal cell carcinoma and prostate carcinoma presented to the dermatology clinic for a routine full body skin examination. An 8 by 5mm tan pink papule with minimal overlying scale was observed on his left posterolateral foot (Fig 1). Dermoscopy demonstrated a tan pink background with central linear helical or corkscrew and coiled vessels and a small amount of overlying light-yellow scale (Fig 2).

Histology revealed a dermal lymphocytic proliferation that included a population of large atypical cells with prominent nucleoli and variable amounts of amphiphilic cytoplasm. This was in the context of a mixed inflammatory background with prominent neutrophils. The large atypical cells demonstrated multinucleate forms and occasional horseshoe shaped nuclei were present with conspicuous mitoses. These cells stained positively for CD30, TIA-1, CD4, and CD2 and 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 with 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.

This is the first time that large tortuous, corkscrew, or linear-helical blood vessels have been described in the context of an ALCL. It has been previously reported that the finding of corkscrew vessels on dermoscopy has been associated with nodular melanoma, desmoplastic melanoma, and cutaneous melanoma metastasis[2]. While it is well established that linear helical vessels are associated with malignant lesions, a definitive cause of this pattern has not been established. It is unclear why these vessels form but as they appear to have a predilection for cancers that have either already metastasized or have a high rate of metastasis, they may be related to features associated with tumor invasion. This theory is supported by the fact that increased axial tension decreases tortuosity, so that when this tension is decreased such as in a rapidly growing vessel, the tortuosity increases[3]. Additionally, weakening of the arterial wall and degradation of surrounding connective tissue, which occurs 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 after linear helical or corkscrew shaped vessels are identified via 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.

References:

1. Kittler H, Marghoob A, Argenziano G, *et al*. Standardization of terminology in dermoscopy/dermatoscopy: Results of the third consensus conference of the International Society of Dermoscopy. *J Am Acad Dermatol* 2016; 74(6): 1093-1106.
2. Togawa Y. Review of vasculature visualized on dermoscopy. *J Dermatol* 2017; 44(5), 525-532.
3. Han H. Twisted blood vessels: symptoms, etiology and biomechanical mechanisms. *J Vasc Res* 2012; 49(3): 185-197.
4. Sounni N, Paye A, Host L, Noël A. T-MMPS as regulators of vessel stability associated with angiogenesis. *Front Pharmacol* 2011; 2: 111.
5. Luca M, Huang S, Gershenwald J, *et al.* Expression of interleukin-8 by human melanoma cells up-regulates MMP-2 activity and increases tumor growth and metastasis. *Am J Pathol* 1997; 151(4): 1105-13.

Fig 1. Rounded erythematous papule with overlying scale on the left posterolateral foot.

Fig 2. Dermoscopic features include a tan pink background with central grouped corkscrew vessels and overlying yellow/white scale.