Dermatology Practical & Conceptual

EMA-Positive Superficial ALK–Rearranged Myxoid Spindle Cell Neoplasm (SAMS): A Case Report of a Rare Entity and Dermoscopic Findings

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Introduction

Superficial anaplastic lymphoma kinase (ALK)–rearranged myxoid spindle cell neoplasm (SAMS) is a recently described and rare cutaneous soft tissue tumor characterized by concentric spindle cell whorls and cords embedded in an abundant myxoid to collagenous stroma [1]. Immunohistochemically, SAMS typically demonstrates ALK positivity with frequent coexpression of CD34 and S100 [1,2]. Although epithelial membrane antigen (EMA) immunostaining was initially reported as negative in most cases, recent studies suggest that EMA expression may occur more frequently than previously recognized [2,3].

Because of its rarity and its histopathologic overlap with peripheral nerve sheath tumors, SAMS represents a diagnostic challenge. Moreover, dermoscopic features of SAMS have not yet been described in the literature. Herein, we report a case of EMA-positive SAMS and provide, to our knowledge, the first description of its dermoscopic appearance.

Case Presentation

A 56-year-old male patient presented to the outpatient dermatology clinic with a solitary, asymptomatic, soft pink nodule on the right shoulder, present for two months. Clinical examination revealed a 1-cm dermal nodule (Figure 1A). Dermoscopic evaluation showed a peripheral pigment network with central yellow-white structures and polymorphic vessels (Figure 1B).

An excisional biopsy was performed. Gross examination revealed a small tan-whitish dermal nodule. Histopathological analysis demonstrated a relatively well-circumscribed dermal spindle cell proliferation embedded in a myxoid stroma. Tumor cells were arranged in loose concentric whorls and

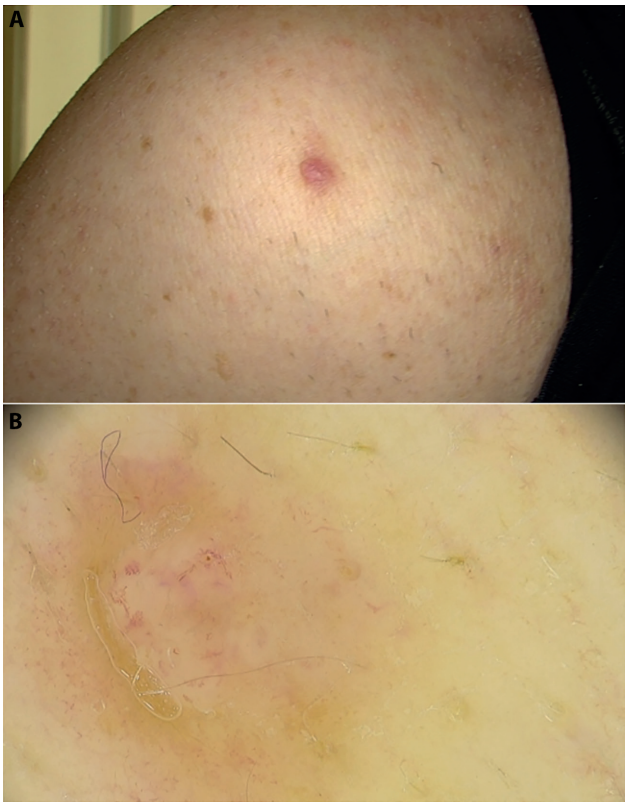

Figure 1. A) Clinical presentation, B) Dermoscopic image: peripheral pigment network with central yellow structures and polymorphic vessels.

short fascicles within a richly vascular background. No cytologic atypia, mitotic figures, or necrosis were identified, and surgical margins were clear (Figure 2A–B).

Immunohistochemical staining showed tumor cell positivity for S100 and EMA (Figure 2C), while CD34, SOX10, PRAME, desmin, and cytokeratin CAM5.2 were negative. ALK (D5F3) immunostaining demonstrated diffuse positivity (Figure 2D), supporting the diagnosis of EMA-positive SAMS. Fluorescence in situ hybridization could not be performed due to limited residual tissue. No additional treatment was required, and no recurrence has been observed to date. Written informed consent was obtained.

Conclusion

SAMS is a rare cutaneous neoplasm with an indolent clinical course that may closely mimic peripheral nerve sheath tumors, including perineurioma, schwannoma, and hybrid variants, which share overlapping spindle cell morphology and myxoid stroma [4]. Importantly, these entities are consistently negative for ALK immunostaining, making ALK expression a key discriminating feature [1,4].

In cases of EMA-positive SAMS, the diagnostic process may be particularly challenging, as EMA immunoreactivity

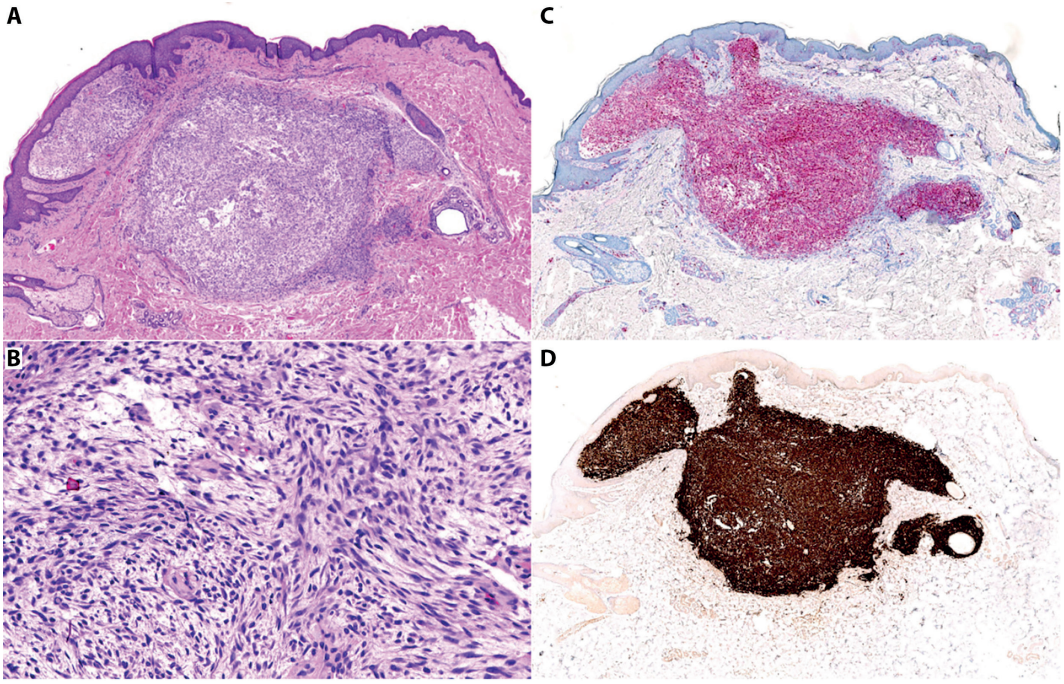

Figure 2. A) Histological examination: relatively well-circumscribed dermal spindle cell mesenchymal proliferation in a myxoid stroma (Hematoxylin and eosin, 2x); B) tumor cells arranged in a loose concentric whorls and fascicles and embedded in a richly vascular mixoid stroma (Hematoxylin and eosin, 40x); C) Positive staining of EMA, (2x); D) Diffuse positive staining for ALK (2x)

was originally considered uncommon. While early reports described EMA positivity only in isolated cases [3], more recent studies have demonstrated EMA expression in a substantial proportion of SAMS cases, suggesting that this feature is more frequent than previously recognized [2]. This highlights the risk of misdiagnosis, particularly if ALK immunohistochemistry is not included in the diagnostic panel.

This report also provides the first description of dermoscopic features of SAMS. The central yellow-white area may correlate with spindle cell proliferation and myxoid or fibrotic dermal stroma, resembling the dermoscopic appearance of dermatofibroma [5]. Peripheral pigmentation may reflect hyperpigmented rete ridges at the tumor margin, while polymorphic vessels could be attributed to tumor-associated neovascularization, as described in other cutaneous tumors [6]. Although histopathologic evaluation remains essential, dermoscopy may offer useful non-invasive diagnostic clues. Further studies are needed to better define the dermoscopic spectrum of SAMS.

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