Exophytic Scaly Nodule on the Wrist

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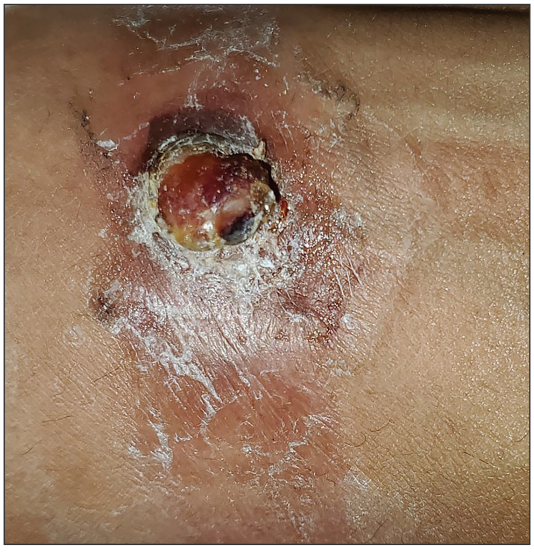

A 30-year-old Black man presented to the dermatology clinic with a rapidly growing, exophytic, scaly nodule on the right volar wrist of 2 months' duration. The patient's medical history was otherwise unremarkable. Physical examination revealed an irregularly bordered, red to violaceous, scaly, eroded, exophytic nodule on the wrist that was 2 cm in diameter with a surrounding adherent white-yellow crust. The patient had presumed the nodule was a wart and had been self-treating with over-the-counter salicylic acid and cryotherapy with no relief. He denied any bleeding or pruritus. The rest of the skin examination was unremarkable. A shave biopsy was performed for further evaluation.

WHAT'S YOUR **DIAGNOSIS?**

a. amelanotic melanoma
b. atypical Spitz tumor
c. bacillary angiomatosis
d. Kaposi sarcoma
e. pyogenic granuloma

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THE **DIAGNOSIS:** Atypical Spitz Tumor

The shave biopsy revealed extensive dermal proliferation with spitzoid cytomorphology containing large, spindled nuclei; prominent nucleoli; and abundant homogenous cytoplasm arranged in haphazard fascicles. The proliferation was associated with prominent pseudoepitheliomatous hyperplasia of the overlying epidermis, and anaplastic lymphoma kinase immunohistochemistry showed diffuse strong positivity. Fluorescence in situ hybridization confirmed fusion of the tropomyosin 3 (*TPM3*) and anaplastic lymphoma kinase (*ALK*) genes, which finalized the diagnosis of an *ALK*-mutated atypical spitz tumor. Due to the location and size of the lesion, Mohs micrographic surgery was performed to excise the tumor and clear the margins.

Spitz nevi are uncommon benign melanocytic neoplasms that typically occur in pediatric populations.[1] Atypical spitz nevi comprised fewer than 17% of all childhood melanocytic nevi in the United States and can be considered in the broader category of spitzoid tumors. Spitz nevi are divided into 3 classes: Spitz nevus, atypical Spitz nevus, and spitzoid melanoma. Atypical Spitz nevi have typical Spitz nevus and spitzoid melanoma features and often can be difficult to distinguish on dermoscopy. Malignant Spitz tumors typically occur in the fifth decade of life, though the age distribution can vary widely.[1]

Black patients are less likely to be diagnosed with Spitz nevi, potentially due to a lower prevalence in this population, thus limiting the clinician's clinical exposure and leading to increased rates of misdiagnoses.[2] Spitz nevi usually manifest as well-circumscribed, dome-shaped papules and frequently are described as pink to red due to increased vascularity and limited melanin content[1]; however, these lesions may appear more violaceous, dusky, or dark brown in darker skin types. Additionally, approximately 71% of patients in a clinical review of Spitz nevi had a pigmented lesion, ranging from light brown to black.[3] It is important for dermatologists to understand that the contrast in color between the nevus and the surrounding skin may not be as striking, prominent, or clinically concerning, particularly in darker skin types, such as in our patient.

Spitz nevi frequently manifest as rapidly growing solitary lesions most frequently developing in the lower legs (shown in 41% of lesions in one report).[4] However, a recent retrospective review indicated that Spitz nevi in Black patients most commonly were found on the upper extremities, as was seen in our patient.[2] Compared to typical and common Spitz nevi, atypical Spitz nevi often are greater than 10 mm in diameter and have features of ulceration.

Diagnosing atypical spitzoid melanocytic lesions requires adequate clinical suspicion and confirmation via biopsy. Under dermoscopy, typical Spitz nevi often display a starburst or globular pattern with pinpoint vessels, though it can have variable manifestations of both patterns. Atypical Spitz nevi can be challenging to distinguish from melanoma on dermoscopy since both conditions can have atypical pigment networks or structureless homogenous areas.[1] Consequently, there often is a lower threshold for biopsy and possible follow-up excision for atypical Spitz nevi. Histopathology of atypical Spitz nevi includes epithelioid and spindle melanocytes but can share features of melanomas, including areas of prominent pagetoid spread, asymmetry, and poor circumscription.[5] Furthermore, atypical Spitz nevi with *ALK* gene fusion, as seen in our patient, have been shown in the literature to demonstrate distinct histopathologic features, such as wedge-shaped extension into the dermis or a bulbous lower border that can resemble pseudoepitheliomatous hyperplasia.[6]

The differential diagnosis for this rapidly growing scaly nodule also should include pyogenic granuloma, bacillary angiomatosis, Kaposi sarcoma, and amelanotic melanoma. Pyogenic granuloma is a rapidly growing, benign, vascular tumor that often becomes ulcerated and can occur in any age group.[7] Pyogenic granuloma frequently appears at sites of trauma as a solitary, bright pink to red, friable, pedunculated papule and often manifests on the arms, hands, and face, similar to atypical Spitz nevi, though they can appear anywhere on the body. Histology shows a lobular capillary network with a central feeder vessel.[7]

Bacillary angiomatosis is an uncommon cutaneous infection associated with vascular proliferation and neovascularization due to the gram-negative organism *Bartonella henselae*.[8] Bacillary nodules typically are reddish to purple and appear on the arms, sometimes with central ulceration and bleeding. Patients may present with multiple papules and nodules of varying sizes, as the lesions can arise in crops and follow a sporotrichoid pattern. Most patients with bacillary angiomatosis are immunosuppressed, though it rarely can affect immunocompetent patients. Histologically, bacillary angiomatosis is similar to pyogenic granuloma, though Gram or Warthin-Starry stains can help differentiate *B henselae*.[8]

Kaposi sarcoma is a malignant vascular neoplasm that often manifests in immunocompromised patients as violaceous, purple, or red patches, plaques, and nodules on the skin or oral mucosa. Histopathology shows spindle cell proliferation of irregular complex vascular channels dissecting through the dermis. Human herpesvirus 8 immunohistochemistry can be used to confirm diagnosis on histopathology.[9] In contrast, amelanotic melanoma consists of lack of pigmentation, asymmetry with polymorphous

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vascular pattern, and high mitotic rate and is commonly found in sun-exposed areas. Dermoscopic features include irregular globules with blue-whitish veil.[10]

Treatment of atypical Spitz nevi depends mainly on the age of the patient and the histologic features of the nevus. Adults with atypical Spitz nevi frequently require excision, while the preferred choice for treatment in children with common Spitz nevi is regular clinical monitoring when there are no concerning clinical, dermoscopic, or histologic features.[8] Compared to common Spitz nevi, atypical Spitz nevi have more melanoma-like features, resulting in a stronger recommendation for excision. Excision allows for a more thorough histologic evaluation and minimizes the likelihood of a recurrent atypical lesion.[11] In all cases, close clinical follow-up is recommended to monitor for reoccurrence.

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