Partly Pigmented Papule on the Nose: A Friend or Foe?

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Case Presentation

An 81-year-old Caucasian man with fair, sun-damaged skin presented to a dermatologist with two nodular lesions on the right side of his face [Figure 1a]. The upper lesion, located beneath the right eye, exhibited clinical, dermatoscopic, and histopathological features consistent with nodular basal cell carcinoma, posing no diagnostic challenge. The lower lesion on the right ala nasi presented clinically as a firm, round, 6 mm blue nodule with a central crust.

In dermatoscopy [Figure 1b], the upper half of the nasal lesion displayed a pinkish–white appearance, whereas the lower half exhibited a blue–white structureless background. Within the lower portion of the lesion, radially arranged thin, hairpin-like vessels surrounded by a white halo were visualized. Given the suspicious characteristics of this lesion, although a definitive diagnosis was not possible, a biopsy was performed on the nasal lesion, and the obtained material was sent for histopathological evaluation. The patient provided written consent for this case to be published.

Question

What is Your diagnosis?

Provided Differential Diagnosis

- Angiokeratoma
- Keratoacanthoma
- Nodular pigmented basal cell carcinoma
- Nodular pigmented squamous cell carcinoma.
- Nodular pigmented spindle cell melanoma

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Correct Answer

Nodular pigmented spindle cell melanoma

Discussion

Histopathology revealed that this is a dermal tumor consisting of clusters of atypical pigmented spindle cells, with a mitotic rate of 3–4 mitoses/mm^2 [Figure 2a]. Immunohistochemical testing showed diffuse positivity for S100 [Figure 2b], HMB-45 and melan-A markers, whereas cytokeratin AE1/AE3 and CK5 markers [Figure 2c] were negative in the tumor cells. Based on the histopathological report, a final diagnosis of nodular pigmented spindle cell melanoma with a Breslow thickness of 5 mm was established.

Spindle cell melanoma (SCM) is a rare melanoma subtype that can often pose a diagnostic challenge due to its variable clinical and dermatoscopic characteristics. It has been noted in the literature that SCM frequently presents as a non-specific, amelanotic lesion, resembling a scar or inflammation, and it can mimic other types of tumors, often leading to delayed or incorrect diagnosis.[1]

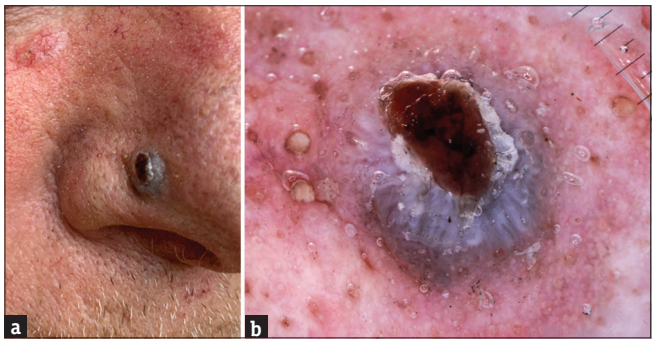

Figure 1: Initial findings–clinical and dermatoscopical image. (a) Clinical image showing nodular basal cell carcinoma under the right eye and nodular pigmented lesion on the right ala nasi. (b) Dermatoscopy of the lesion on the nose shows a central crust, a pink background on the upper part, and a blue-white structureless background on the lower part of the lesion. In addition, linear hairpin vessels surrounded by a white halo can be distinguished in the blue–white area

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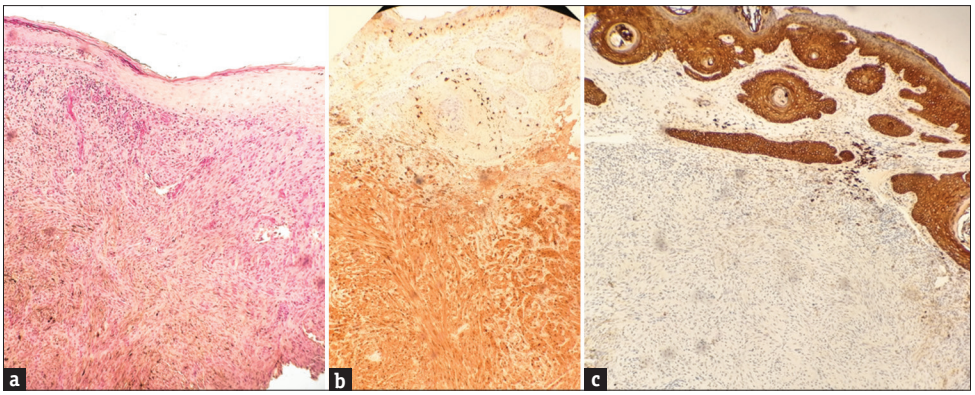

Figure 2: Histopathology. (a) Hematoxylin and eosin staining show dermal clusters of atypical spindle cells, some of which are pigmented, ×100. (b) Positive immunostaining for S100 in the dermis, ×100. (c) Immunostaining positive for cytokeratin AE1/AE3 in the epidermis, negative in tumor cells, (×10)

This case presented a diagnostic challenge because the clinical signs were more indicative of a keratinocyte tumor rather than melanoma. For instance, the presence of hairpin vessels with a white halo is a more characteristic sign for conditions such as keratoacanthoma, seborrheic keratosis, or squamous cell carcinoma. Although it has been documented in the literature that hairpin vessels can occasionally appear in amelanotic melanoma, the occurrence of a white halo around these vessels in melanoma, or any melanocytic lesion for that matter, is exceedingly rare.[2] Although a central crust can sometimes be observed in melanoma, it is also a more common feature in keratinocyte tumors, including squamous and basal cell carcinomas, as well as keratoacanthoma.[3] The sole significant sign in this case that suggests the possibility of melanoma is the presence of a unilateral, blue, asymmetrical, structureless area in the lower part of the lesion, which has been reported in the literature to be observed in up to 74.7% of melanoma cases.[4]

Diagnosing an SCM can be challenging, both clinically and histopathologically. Recent studies have identified immunohistochemical marker combinations that can aid in SCM diagnosis, particularly when distinguishing it from its close variant, desmoplastic melanoma (DM). Both tumors comprise atypical, spindled, malignant melanocytes and both can potentially mimic benign amelanotic processes. However, the prognosis and treatment approach for SCM and DM differ significantly. When looking for melanoma, the primary markers to assess are protein S100 and SOX-10. The common triad for SCM includes diffusely positive S100, along with positive HMB-45 and melan-A markers. In rare instances, HMB-45 may be negative in SCM. Conversely, desmoplastic melanoma often tests negative for HMB-45 and melan-A, with diffuse S100 positivity. Histologically, spindle cells in DM are separated by abundant collagen and fibrous fibers, in contrast to SCM, where spindle cells are organized in large clusters and fascicles, constituting at least 90% of the tumor.[5]

Our report shows an interesting case from dermatology practice in which melanoma exhibited several unusual clinical characteristics. This emphasizes the significance of collaboration with pathologists, as immunohistochemical investigations proved crucial in reaching an accurate diagnosis. This case serves as a reminder for dermatologists to consider rare melanoma subtypes when encountering clinically atypical and suspicious lesions.

Learning Points

1. Spindle cell melanoma, a rare tumor, may resemble various skin conditions including scars and inflammation; our case demonstrates its possible similarity to keratinocyte tumors.
2. Diagnosing spindle cell melanoma can pose a challenge, necessitating a histopathological examination for the differentiation of spindle cell melanoma from other skin lesions.
3. Important immunohistochemical markers to consider when diagnosing melanoma include S100, HMB-45, and melan-A, in conjunction with an evaluation of the organization of spindle cells within the tumor.
4. Further research is needed to enhance our comprehension of spindle cell melanoma and its clinical, dermatoscopic, and immunohistochemical manifestations.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and

due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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