SkIndia Quiz 33

Solitary pigmented plaque containing a blue-gray papule

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A 35-year-old woman presented with a new dark papule that had arose within a previously stable tan plaque on her right thigh. There was no history of itching or bleeding from the lesion. Her past medical and family history were unremarkable. On examination, there was a 1.1 cm sharply demarcated tan plaque on the right anterior thigh containing an eccentric blue-gray papule [Figure 1].

Low-power histopathologic examination revealed a nested proliferation of cytologically bland appearing melanocytes focally along the dermoepidermal junction and more extensively in the dermis with a second population of pigmented epithelioid appearing melanocytes with surrounding melanophages in the upper reticular dermis [Figure 2]. High-power viewing confirmed the presence of two populations of melanocytes; the first was the small amelanotic population and the second consisted of localized nests of epithelioid melanocytes with conspicuous cytoplasmic melanization. There were prominent pigmented melanophages [Figure 3].

What is THE diagnosis?

A. Cellular blue nevus
B. Clonal nevus (inverted type A nevus)
C. Common blue nevus
D. Deep penetrating nevus
E. Epithelioid blue nevus

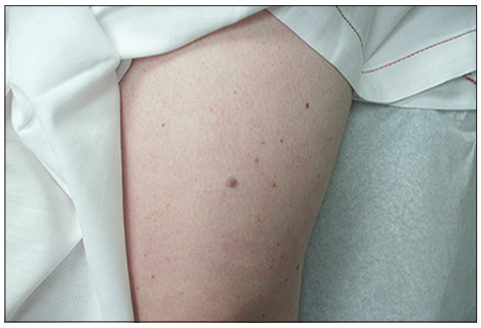

Figure 1: Tan plaque containing an eccentric blue-gray papule on the right anterior thigh

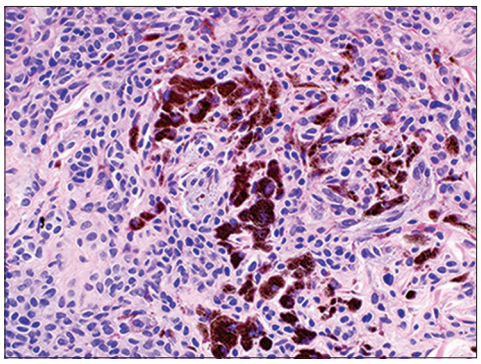

Figure 3: Focus of pigmented epithelioid appearing melanocytes with enlarged atypical appearing nuclei and surrounding pigmented melanophages with adjacent nests of smaller nonpigmented type B melanocytes (H and E, original magnification ×400)

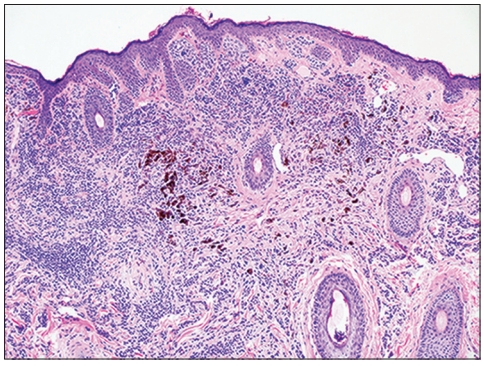

Figure 2: Compound melanocytic nevus with a focus of pigmented epithelioid appearing melanocytes with surrounding melanophages (H and E, original magnification ×100)

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Quick Response Code:

Answer

Clonal nevus.

Discussion

Known by a number of other designations, including inverted type A nevus and melanocytic nevus with focal atypical epithelioid components, clonal nevus is a distinct entity in the differential diagnosis of pigmented, dermal, melanocytic lesions, including malignant melanoma. However, careful histopathologic inspection will reveal distinctive features of clonal nevi that distinguish these benign lesions from malignant melanoma and other pigmented dermal melanocytic lesions.

Clinically, a clonal nevus appears as a biphasic, tan to light brown nevus with a focal macular or papular region of darker pigmentation, which may appear blue-gray to blue-black [Figure 1].[1,2] This dark area commonly develops amidst a preexisting banal compound or dermal nevus, either centrally or eccentrically, and can be alarming to an affected patient or physician who recognizes the change. Histopathologically, this dark focus corresponds to clusters of similar appearing, atypical, epithelioid-appearing melanocytes surrounded by heavily pigmented melanophages [Figure 2].[1,2] These melanocytes are characterized by dusty brown cytoplasmic melanization, enlarged nuclei with prominent nucleoli, and (in stark contrast to the mitotically active melanocytes of melanoma) rare, if any, mitotic figures.[1] The nuclei of these melanocyte "clones" are uniform and slightly larger than those of the melanocytes within the surrounding ordinary melanocytic component of the lesion [Figure 3].[1,2] The pigmented epithelioid melanocytic component demonstrates HMB-45 expression, whereas the surrounding nonpigmented, nonepithelioid dermal melanocytes associated with the lesion typically do not.[3] A histopathologically similar lesion is the deep penetrating nevus. DPN assumes a symmetrical, inverted wedge shape composed of atypical appearing spindled to epithelioid melanocytes with dusty brown cytoplasm and associated heavily pigmented melanophages.[4] In addition, the pigmented dermal melanocytes in DPN are often associated with a background conventional appearing nevus.[4] Although DPN and clonal nevus demonstrate similar cytomorphology, and a background of ordinary appearing melanocytic component, they primarily differ in their respective depth of dermal involvement and presence of periadnexal/neurovascular growth with DPN extending into the reticular dermis in an inverted wedge shape along the adnexal/neural vascular structures.[1,4] In contrast, clonal nevi remain confined to the superficial dermis.[1] Interestingly, one study identified lesions of intermediate depth, between that of clonal nevi and DPN: A finding that supports the notion that these two lesions may in fact represent different points on a continuum for the same entity.[5] Finally, some authors favor the term combined nevus (monomorphic variant), for those lesions showing features of DPN.[6] Despite their depth of involvement and cytologic atypia, DPNs have never been found to metastasize and, just like clonal nevi, are considered to be benign lesions.[4]

Other pigmented, dermal melanocytic lesions include the common blue nevus, cellular blue nevus, and epithelioid blue nevus. Clinically, common blue nevi are acquired blue or blue-black macules or dome-shaped papules occurring on the backs of the hands and feet, buttocks, face, or scalp.[7] Onset may be in childhood or later in life.[7] Histopathologically, the common blue nevus appears as a symmetric, well-demarcated proliferation of dendritic melanocytes with darkly staining nuclei and elongated cytoplasmic processes. The process is often centered on adnexal structures, and the surrounding stroma can demonstrate varying degrees of fibrosis.[7]

Cellular blue nevi can be biphasic, like clonal nevi, and in that setting are usually paired with a component of common blue nevus.[7] They are larger than common blue nevi, often 1–3 cm or larger, and most commonly present on the buttocks—less often on the head, neck, and extremities—as firm, dermal nodules with a bluish-gray hue. Like DPN, cellular blue nevi show extension along neurovascular and adnexal structures sometimes into the deeper, subcutaneous tissue. Pale-staining oval or spindled melanocytes form fascicles or sheets in "cellular" areas, and "wreath-like" giant cells can be found.[7]

Epithelioid blue nevus was originally described presenting in childhood as a component of the Carney complex, a familial lentiginosis and low-grade, multiorgan neoplasia syndrome consisting of endocrine overactivity, patchy skin pigmentation, myxomas, and psammomatous, melanotic schwannomas.[7,8] However, since then, sporadic lesions have been described in both children and adults as a dome-shaped darkly pigmented papule on the trunk or extremities.[9] Histopathologically, epithelioid blue nevi present as an oval or wedge-shaped dermal tumor composed of pigmented epithelioid melanocytes and pigmented spindled melanocytes similar to common blue nevi.[7-9] The epithelioid melanocytic component demonstrates large vesicular nuclei with prominent nucleoli in association with heavily pigmented globular melanophages.[7-9] Epithelioid blue nevi may occur in association with other blue nevi as part of a combined nevus.[10] Some consider it to represent the same entity as pigmented epithelioid melanocytoma, which is classified as a low-grade melanocytic tumor with metastatic potential.[7]

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