


Nevo de Spitz Angiomatoide

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RESUMO – O nevo de Spitz angiomatoide é um tumor raro que associa os aspetos clássicos do nevo de Spitz a uma componente vascular marcada. Clinicamente, manifesta-se como uma lesão papular rosada ou acastanhada, habitualmente solitária, mais frequente nas extremidades de adultos jovens. À histologia, caracteriza-se por uma proliferação de melanócitos epitelioides ou fusiformes num estroma fibroso denso, onde é evidente uma assinalável proliferação de pequenos vasos. O diagnóstico diferencial com melanoma maligno pode ser difícil, particularmente com a variante desmoplásica ou com aqueles com aspetos de regressão. O seu comportamento é benigno, comprovado pela ausência de recidivas locais ou metástases à distância durante o seguimento a longo prazo.

PALAVRAS-CHAVE – Neoplasias da Pele; Nevo de Células Epitelioides e Fusiformes.

Angiomatoid Spitz Nevus

ABSTRACT – Angiomatoid Spitz nevus is a rare tumor that combines the classic aspects of Spitz nevus with a prominent vascular component. Clinically, it presents as a pink or brownish papule, usually solitary, in the extremities of young adults. On histology, it is characterized by a proliferation of epithelioid or spindle cell melanocytes embedded in a fibrous stroma, where a dense proliferation of small vessels is evident. The differential diagnosis with malignant melanoma can be difficult, particularly with the desmoplastic variant or with those with marked regression. Its behavior is benign, as suggested by the absence of local recurrences or distant metastases during long-term follow-up.

KEYWORDS – Nevus, Epithelioid and Spindle Cell; Skin Neoplasms.

INTRODUCTION

Spitz nevus is a benign melanocytic proliferation characterized histologically by the presence of large melanocytic cells with spindle and epithelioid morphology.¹ Several variants have been described to date, such as the pigmented, desmoplastic or atypical Spitz nevus.² Because some of its aspects may resemble those of a melanoma, the differential diagnosis between the two is often difficult.

CASE REPORT

A 27-year-old woman presented to the Dermatology Department with an asymptomatic pigmented lesion on the thigh that had been present since infancy. On examination she had a 5-mm homogenous brown dome-shaped papule with an ill-defined border. On dermoscopy, a homogenous brown-grayish pattern with a central brighter area and small

linear vessels were seen. No pigment network was evident. The clinical impression was that of a dermatofibroma but, due to its nonspecific character, the lesion was excised.

On histology, findings were striking. The lesion, predominantly located in the dermis, was composed of spindle and epithelioid melanocytes and a prominent vascular network dissecting through collagen fibers (Fig. 1A). The large melanocytes contained voluminous pale eosinophilic cytoplasm, enlarged oval nuclei and a prominent nucleolus. They were distributed diffusely throughout the lesion as solitary units among thick collagen bundles. No mitoses were observed. A striking feature was the presence of numerous small blood vessels with round lumina lined by a thin wall of endothelial cells without cellular atypia (Fig. 1B). Epidermis showed no significant changes, other than some degree of hyperplasia. No Kamino bodies were present. The lesion was devoid of

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Caso Clínico

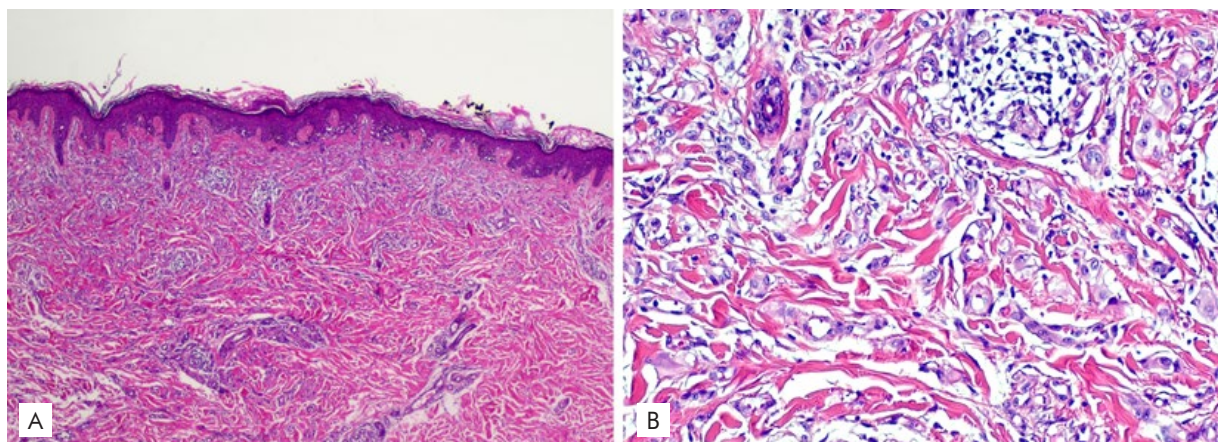


Figure 1 - Histology: The lesion is predominantly located in the dermis (A) and composed of spindled and epithelioid melanocytes with a prominent vascular and fibrogenic stromal response (B) [H&E, original magnification 20x (A) and 100x (B)].

significant inflammatory infiltrate and melanin pigment. Immunostaining was positive within the melanocytes for both Melan-A (Fig. 2A) and S100 (Fig. 2B), and negative for p63 and HHV8. From these findings, the lesion was diagnosed as angiomaatoid Spitz nevus (ASN).

DISCUSSION

ASN is a rare histological variant of desmoplastic Spitz nevus. First described in 2000 by Diaz-Cascajo,³ it is characterized by a proliferation of epithelioid and spindle-shaped melanocytes embedded in a prominent fibrous stroma with many densely arranged small blood vessels. Clinically, it presents as a popular dome-shaped lesion, usually solitary, often located on the extremities of young adults. Dermoscopic findings include an atypical pigmented network, white lacunar zones, reabsorption areas and short linear telangiectatic vessels.⁴ On histology, although some junctional or compound lesions have been described, the majority of tumors are purely dermal. Melanocytes,

distributed mostly as solitary units, typically possess a plump nucleus with prominent nucleoli and abundant eosinophilic cytoplasm. Mitotic figures are rare and, when present, are confined to the upper dermis. Melanin pigment (present in 40% - 83% of cases^{3,5}) is usually sparse and limited to the superficial component of the lesion. The fibrous stroma is composed of thick collagen bundles and a large number of small blood vessels. These are composed by plump endothelial cells without cellular atypia, lining a round or oval lumina.⁵ This vascular component consisting of a dermal proliferation vascular channels dissecting through collagen fibers may resemble a microvenular hemangioma. Vessel walls can be thick, thin, or mixed, potentially reflecting the tumor evolutionary stage. A mild to moderate perivascular inflammatory infiltrate consisting predominantly of lymphocytes is seen in most lesions.³

Differential diagnosis with malignant melanoma may be difficult, particularly with those with desmoplastic features or partial regression.^{6,7} However, melanocytes in

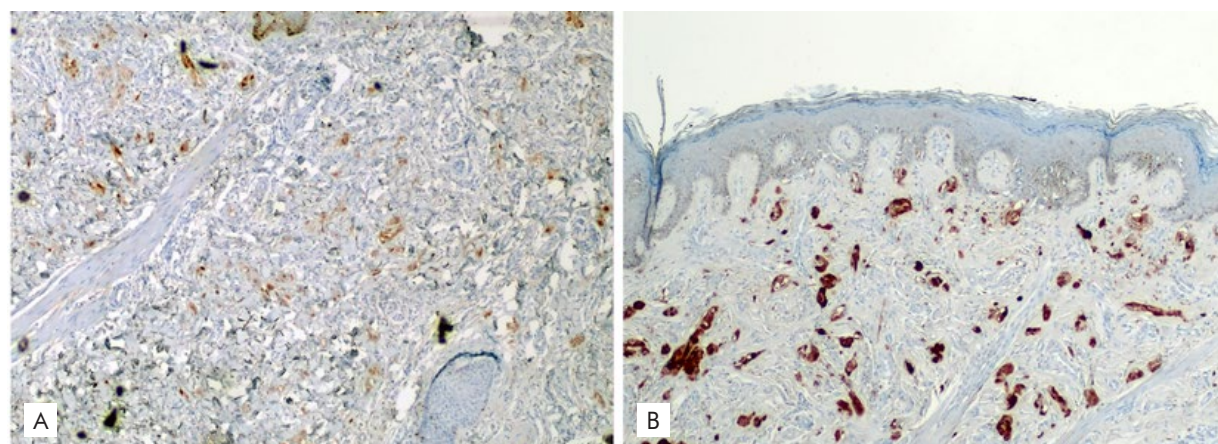


Figure 2 - Immunostaining: Positive within the melanocytes for both Melan-A (A) and S100 (B) (Original magnification, 40x).

malignant melanoma often show more significant cytologic atypia and tendency to cluster, mitoses are more frequent and located in the deeper dermis, asymmetry is more pronounced and an epidermal component is often seen. Other differential diagnosis includes vascular tumors such as hemangiomas, sclerosing hemangioma type dermatofibroma and epithelioid cell histiocytoma³

Regarding the possible relation between the two proliferations (vascular and melanocytic), Helm and Helm⁸ challenge the idea of a simple coincidence. The authors point out that *“the matching and intermingling silhouettes of the two proliferations provide compelling circumstantial evidence”* that they may be related. Research on serpins shed a light on how melanocytes and blood vessels might interact and reveal a complex interaction between the tumor and surrounding stroma.⁹

The benign character of these lesions is supported by the absence of recurrences and metastases after complete excision during long-term follow-up.^{3,7}

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