Caso Clínico

Cellular cutaneous neurothekeoma associated with a café au lait spot *

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Resumo: É relatado um caso de neurotecoma cutâneo celular associado com uma mancha "café com leite" na região dorsal.
Palavras-chave: Neurotecoma cutaneo celular

Summary: A case of cellular cutaneous neurothekeoma associated with a *café au lait* spot on the dorsal region is reported.
Key words: Cellular cutaneous neurothekeoma

INTRODUCTION

Cutaneous neurothekeoma (CNT) is a rare, recently described benign cutaneous neoplasm, which has been reported under various names, including neurothekeoma, pacinian neurofibroma, bizarre cutaneous neurofibroma and cutaneous lobular neuromyxoma.[1] CNT has a fairly distinctive histological appearance and characteristic clinical features, affecting preferentially women (4.3:1), usually arising during childhood or early adult life (mean age of 21.6 years). [2] These lesions are non typical papules or nodules, in general solitary, located on the upper part of the body, with a predilection for head, neck, and shoulders,[3] with documented but rare occurrences on the trunk and lower extremities.[2] CNT has a benign course and recurrence is not seen since excision is complete. Low-grade cytological atypia and mitotic activity are common in cases of CNT, but a malignant form has never been described.[3] Based on the pathological appearance CNT have been subclassified into two main histological varieties, i.e. cellular (CCNT) and myxomatous variants, the latter being referred to as nerve sheath myxoma (NSM).[4,5,6] However, some authors believe that neurothekeoma encompasses a wider spectrum of lesions, with the myxoid and cellular subtypes falling at either end of the morphologic spectrum.[4,7]

The histogenesis of CNT is controversial. The classical NSM has neural (mainly Schwann cell) differentiation. However, in CCNT the neural features are not fully expressed, since CCNT is predominantly composed of undifferentiated cells with partial features of Schwann cells, smooth muscle cells, myofibroblasts and fibroblasts, suggesting a divergent differentiation.[8] Despite confusing immunohistochemical and ultrastructural data, most authors admit that neurothekeoma can express schwannian or perineurial differentiation's, and may also be derived from an undifferentiated mesenchymal cell of neural crest origin.[5]

CASE REPORT

A 28-year-old woman was seen with a history of a few papules. Each of them measures approximately 0,5 to 1 cm in diameter, located on the right dorsal region associated with a five centimeter *café au lait* zone of hyperpigmentation (*café au lait spot*), she had since childhood. Tumors were asymptomatic but grew over several months. They were firm and well circumscribed, with reddish to yellowish color (Figure 1). Adult xanthogranuloma and verrucous nevus were clinically suspected and one lesion was excised.

Histologically there was a circumscribed tumor located in the dermis. The epidermis was flattened with a Grenz zone

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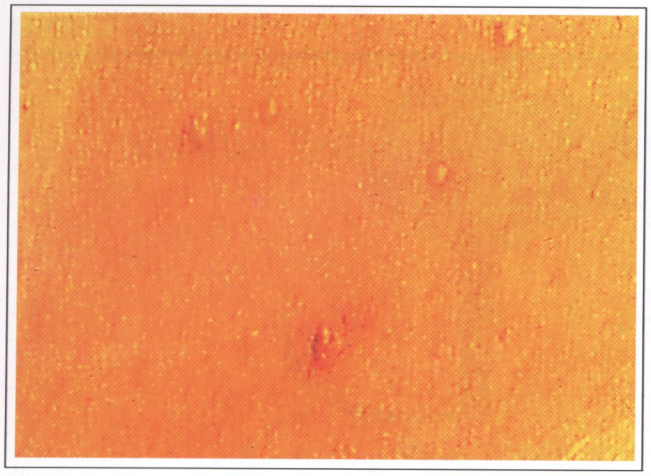

Figure 1: Dorsal papules, associated with a café au lait *zone of hyperpigmentation (café au lait spot)*

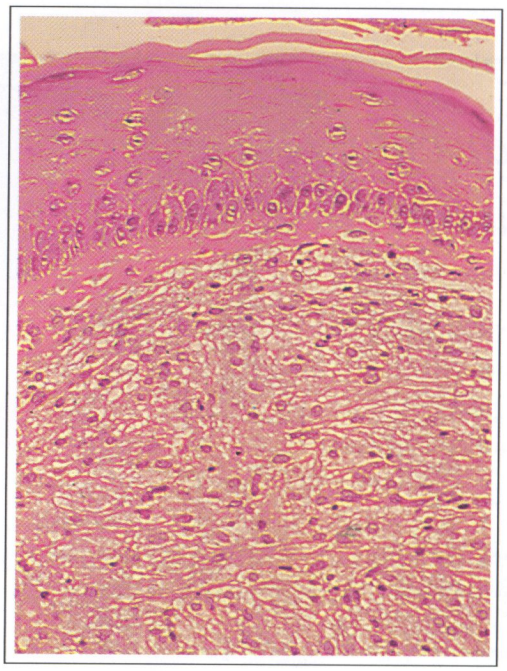

Figure 2: Circumscribed dermal tumor. The epidermis is flattened with a Grenz zone (HE, 120x)

(Figure 2). The tumor showed a fairly discrete fascicles ill-defined multilobular pattern and small nests arranged in a linear or pale eosinophilic concentric array (Figure 3), predominantly epithelioid cells, with some spindle-shaped cells (Figure 4) between scanty hyalinized matrix that fibrous appearance but staining for mucopolysaccharide acid (extracellular mucin). Purely myxoid lobules with scant amounts of mucin were sparsely seen (Figure 5), concurring with the cellular ones. Immunohistochemically, vimentin was diffusely positive (Figure 6), but keratin and S-100 protein were negative. The histological diagnosis of CCNT was made.

DISCUSSION

The differential diagnosis of CNT usually includes spindle and epithelioid cell (Spitz) nevus, malignant melanoma (particularly desmoplastic-neurotropic melanoma), cellular blue nevus, and fibrohistiocytic proliferation. The histological differentiation of CCNT from the myxoid variant of neurothekeoma is based on its prominent cellularity, the lesser degree of myxomatous change, and the less pronounced plexiform compartmentalization by fibrous septae.

Some authors have found S-100 protein in CNT, whereas others failed to detect it, as in the case under discussion. The reason for this may be that CNT are composed of different stages or lines of differentiation of a precursor cell. The immunohistochemical findings of this case support the view that the origin cells of this kind of tumor may be perineurial cells rather than Schwann cells.

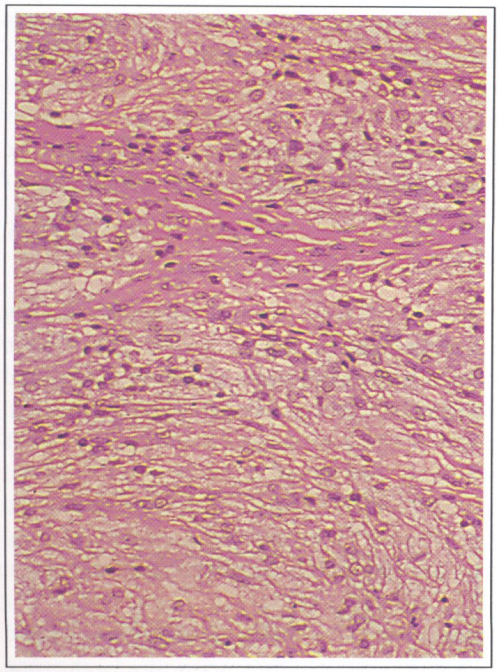

Figure 3: Multilobular pattern of fairly discrete fascicles and small nests arranged in a linear or concentric array (HE, 120x)

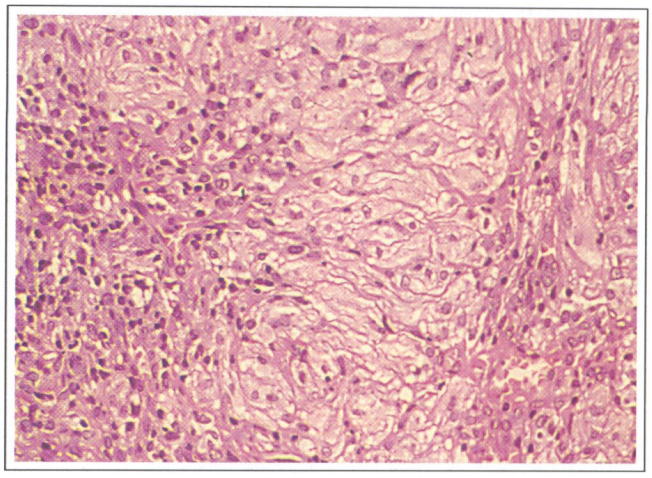

Figure 4: Lobules and fascicles of pale eosinophilic epithelioid cells and some spindle-shaped cells between scanty hyalinized matrix (HE, 250x)

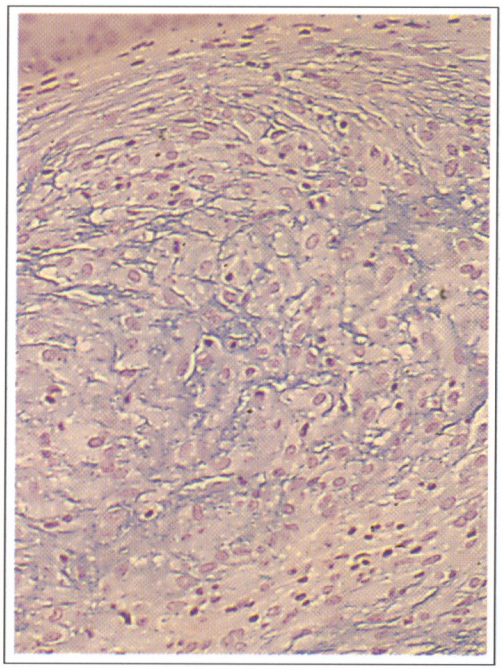

Figure 5: Myxoid lobules with scant amounts of mucin were sparsely seen. Alcien blue (120x)

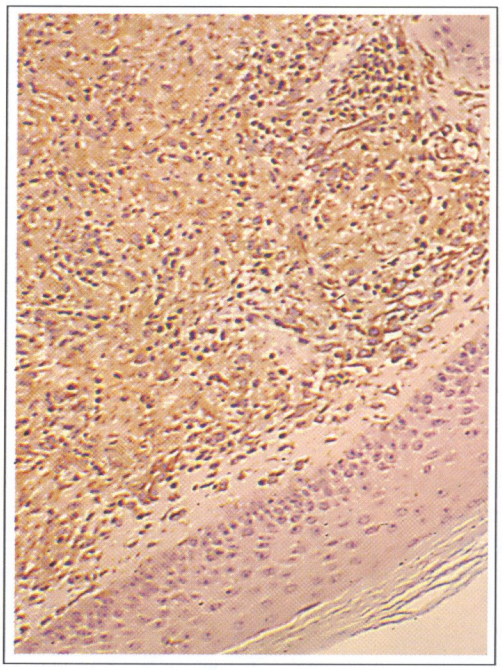

Figure 6: Aggregates of cells positively stained for vimentin. StreptAB Complex/HRP techique (100x)

So far, we think this is the first case described in Brazil and probably the first case in which this type of tumor has been reported to develop associated with a *café au lait* spot, in spite of the common believe that there is no association of these neoplasms with von Recklinghausen's disease.[2]

Because of their rarity and unusual histological appearance, it is advisable the dermatopathologist to be familiar with these lesions; they may be mistaken for other myxoid skin lesions. ❑

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