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Case Report

Cellular Neurothekeoma – Rare but exists

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KEYWORDS

Cellular Neurothekeoma, benign skin tumor, dermoscopy, histopathological diagnosis

ABSTRACT

Cellular neurothekeoma is a rare, benign cutaneous neoplasm with uncertain histogenesis, that most commonly develops on the head or neck. It predominantly affects females in their third or fourth decade. Usually, they do not have any symptoms, but there may be pressure-related pain. Since cellular neurothekeomas are typically rare and do not have well defined clinical symptoms, it is often difficult to distinguish them from other types of skin tumors. We present the case of a 40-year-old female with a slowly growing violaceous nodule on her scalp, noted since childhood but recently exhibiting growth. We describe the diagnostic and therapeutic approach. This case highlights the diagnostic challenges posed by cellular neurothekeoma due to its histological and clinical variability. Accurate diagnosis through histopathology and immunohistochemistry is crucial to avoid misdiagnosis and ensure appropriate management.

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1. Introduction

Cellular neurothekeoma is a rare, benign cutaneous neoplasm of uncertain histogenesis. Unlike dermal nerve sheath myxomas (formerly known as classic or myxoid neurothekeomas), cellular neurothekeomas are not related to tumors of nerve sheath origin. They typically appear as a slow-growing, painless nodule on the head and neck. Most common in women during their third or fourth decade of life, it can be challenging to diagnose due to its histological variability. Accurate diagnosis is essential to distinguish it from other similar skin tumors, ensuring proper treatment. We present a case of a 40-years old female with cellular neurothekeoma and describe the diagnostic and therapeutic approach.

2. Case Report

A 40-years old female presented with an asymptomatic, slowly growing nodule on scalp. The patient reported that she had noticed it since childhood, but it had recently exhibited growth.

On physical examination, the lesion appeared as a well-circumscribed, violaceous-colored nodule of soft consistency (Fig.1a). No other lesions were found, and examination of her neck and supraclavicular lymph nodes revealed no abnormalities. The patient had no other medical conditions and her family history was negative.

Dermatoscopy examination revealed a non-specific pattern with a central purple structure, white structureless areas at the center, and linear irregular vessels (Fig.1b).

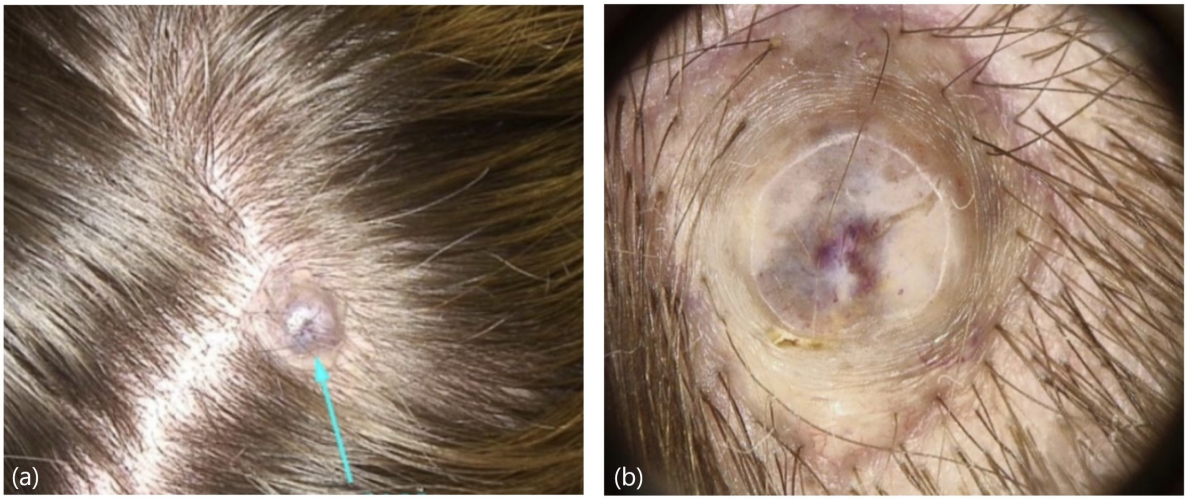

Fig. 1. *(a) Violaceous-coloured well-circumscribed nodule of soft consistency with a diameter of 7mm; (b) Dermatoscopic image showing linear vessels, whitish streak area, purple central structure, structureless white area.*

It was decided for an excisional biopsy with 1-mm margins around the lesion in the form of an ellipse with measures of 4x1.5, and the nodular lesion of 7mm (Fig. 2). The lesion was entirely excised and sent for histopathological examination.

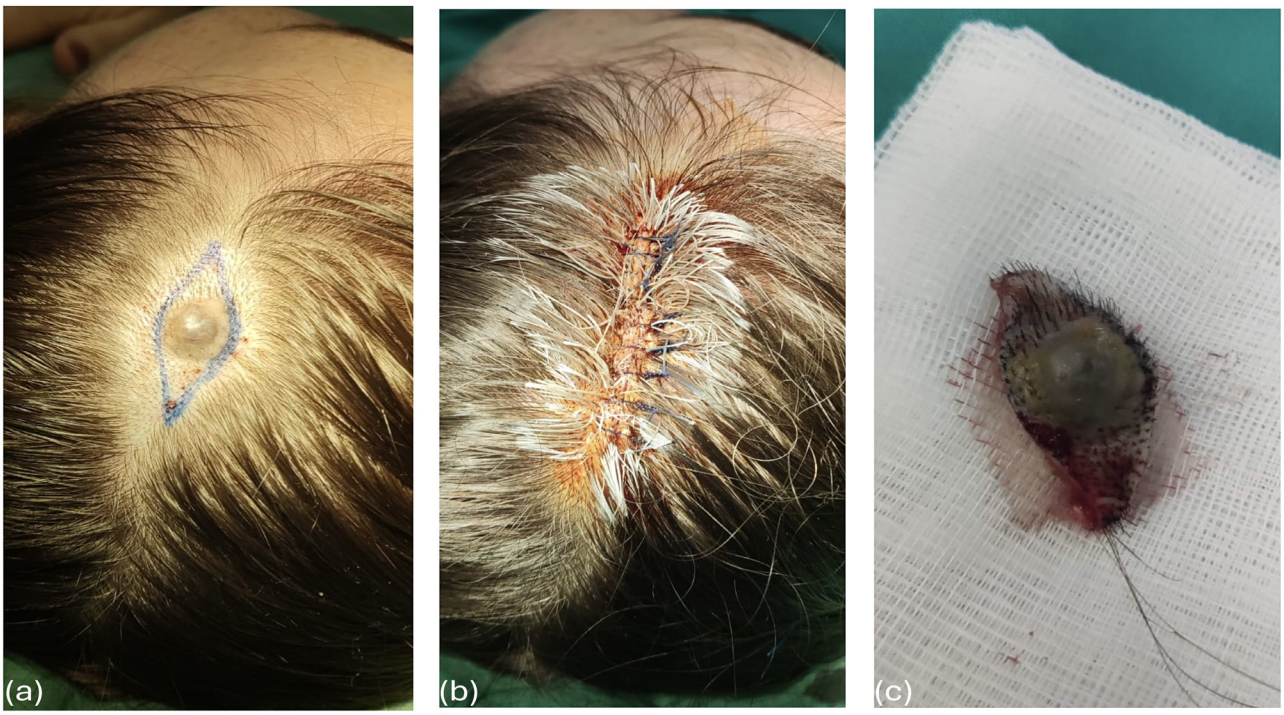

Fig. 2. *(a) Preoperative image of the lesion measuring 4 x 1.5 cm; (b) Postoperative image after complete excision; (c) The specimen.*

Histopathological analysis revealed a lobular growth pattern with infiltration of the subcutaneous tissue and chronic inflammation around histio-epithelioid lobules. The tumor consisted of fused cell neoplasia with a monomorphic appearance arranged in short bundles. No cytologic atypia or significant mitotic activity was observed. There were noticed focal deposits of melanin. Immunohistochemical reactions for protein S100 and CD34 were negative, ruling out nerve sheath tumors. The picture appears compatible with cellular neurothekeoma (Fig. 3).

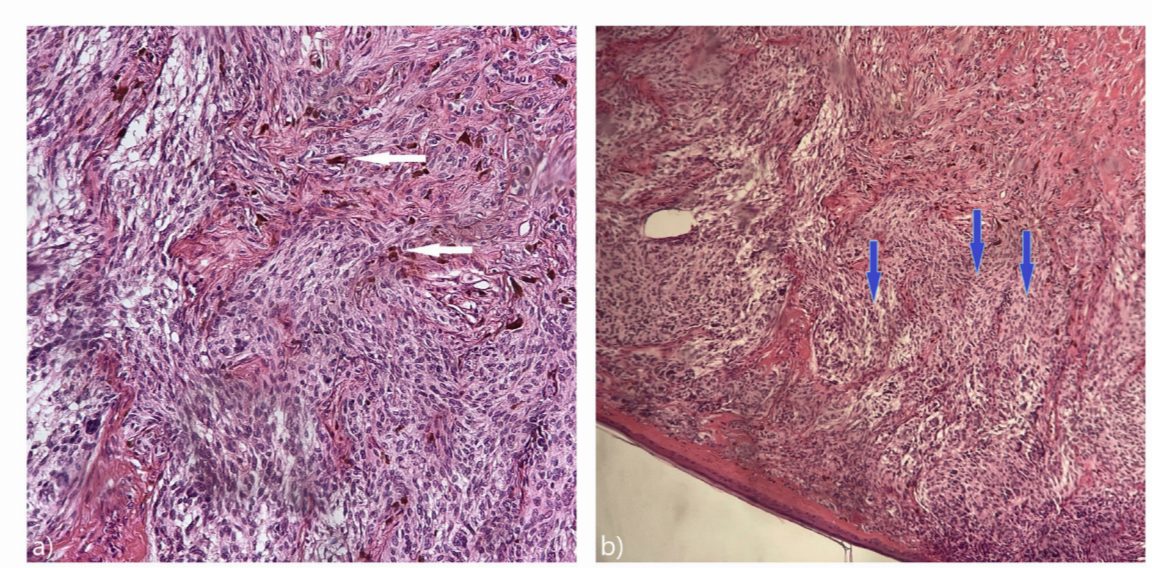

Fig. 3. *(a, b) Histopathology image of the lesion, H&E. Fascicular patterns of spindled and epithelioid mononuclear cells with abundant cytoplasm, indistinct cell borders (blue arrows). Focal deposits of melanin (white arrows).*

3. Discussion

Cellular neurothekeoma is a benign superficial skin tumor of uncertain origin, distinct from dermal nerve sheath myxomas. Previously classified as a subtype of nerve sheath myxomas, cellular neurothekeomas are now recognized as separate entities, with differences in immunohistochemical profiles and histogenesis. Cellular neurothekeomas do not express S100 protein, unlike dermal nerve sheath myxomas, suggesting a fibrohistiocytic origin. It usually occurs during the third and fourth decade of life, often affecting females more than males, with a predilection for the head and neck (1, 2). Neurothekeomas clinically appear as painless, slowly growing, red-colored cutaneous nodule with good mobility. They have a variety of histologic patterns, including myxoid, cellular, or mixed types depending on the amount of myxoid matrix (2, 3).

For many years, neurothekeomas were diagnosed and reported as one of the subtypes of dermal nerve sheath myxomas, because it was initially thought that they originated from Schwann cells (4).

Harkin and Reed described the first neurothekeoma in 1969 as a myxoma originating from the nerve sheath (5). It was only after 1980 that Gallager and Helwig gave this type of tumor its current definition of neurothekeoma (3). There were later two large case studies published in 2007 by Fetsch et al. and Hornick and Fletcher, establishing the immunoprofile of neurothekeomas and further distinguishing it from nerve sheath myxomas (2, 6). In contrast to dermal nerve sheath myxomas, neurothekeomas do not express the S-100 protein, according to recent studies (2, 7). Rather than emerging from peripheral nerve sheath, neurothekeomas have been proposed to originate from fibrohistiocytic cells (2, 3).

Most of these lesions are small and show no cytologic atypia and minimal invasion into other tissues. However, there have been studies that report atypical features of neurothekeomas, which include cytologic atypia, size greater than 1 cm, invasion into surrounding tissues and high mitotic index. The rate of recurrence remains low, in spite of their atypical features (8, 9).

The lack of clearly defined clinical symptoms and physical examination, makes them difficult to distinguish from other skin tumors. The differential diagnosis includes sebaceous cysts, fibrous histiocytoma, pilomatricoma, neurofibroma, basal cell carcinoma or other melanocytic lesions (2, 3). These diseases are clinically similar, but have different pathological features.

In this case, dermoscopy revealed a non-specific pattern that did not provide a definitive diagnosis but guided clinical suspicion for further investigation. Although dermoscopic features are not pathognomonic, these findings overlap with benign fibrohistiocytic tumors. Unlike melanocytic lesions, neurothekeomas typically lack a pigment network. In comparison, basal cell carcinoma may exhibit arborizing vessels, while pilomatricoma often shows a “rolled border” and central calcifications. Dermoscopy can occasionally aid in differentiating benign lesions from malignancies, but its utility in cellular neurothekeoma remains limited. Further studies may elucidate dermoscopic features specific to this tumor (10).

Thus, the confirmation of the diagnosis must be done by histopathological and immunohistochemical examination (7, 9).

Excisional biopsy remains the treatment of choice for cellular neurothekeomas. Most lesions show no cytologic atypia or significant invasion, and recurrence rates are low when margins are clear. However, atypical cases, characterized by cytologic atypia, deep tissue invasion, or increased mitotic activity, may have a higher recurrence risk. Literature suggests that follow-up for at least 12–24 months is beneficial in detecting recurrences, especially in cases with positive margins. While our case showed clear margins, periodic follow-up is recommended (11).

4. Conclusions

It is essential to diagnose neurothekeomas accurately since these lesions can be mistaken for malignancies, resulting in unnecessary treatment. The inclusion of dermoscopic features in diagnostic algorithms may aid in earlier recognition. While the prognosis is excellent, long-term follow-up is recommended for atypical cases to monitor recurrence risk. Dermatologists and pathologist should be aware of the clinical presentation of this rare tumor and the similar clinical and histologic features it shares with other benign and malignant tumors, in order to provide an accurate diagnosis, management and follow-up care.

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DISCLOSURE

All authors report no conflict of interest.

DATA AVAILABILITY STATEMENT

Data openly available in a public repository that issues datasets with DOIs.

ETHICS STATEMENT

The patients in this manuscript have given written informed consent to publication of their case details.

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