

Dermal nerve sheath myxoma of the toe and its management – A case report

*Mixoma dérmico da bainha nervosa do dedo do pé e seu manejo –
Relato de caso*

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ABSTRACT

Nerve sheath myxoma (NSM) is a rare benign tumor of peripheral nerve sheath origin that typically presents as a small nodule on the extremities of young adults. We report an unusual case of dermal NSM of the toe in a 40-year-old man, present since birth. Due to its nonspecific clinical features, NSM may be difficult to diagnose before biopsy and can resemble other benign soft-tissue lesions. Histopathologic and immunohistochemical evaluation is therefore essential for establishing accurate diagnosis. Awareness of this uncommon entity and its inclusion in the differential diagnosis of small, firm subcutaneous lesions may facilitate appropriate management and help prevent misdiagnosis.

Keywords: Myxoma; Neurothekeoma; Toes

RESUMO

O mixoma da bainha nervosa (MBN) é um tumor benigno raro originário da bainha dos nervos periféricos, que geralmente se apresenta como um pequeno nódulo nas extremidades de adultos jovens. Relatamos um caso incomum de MBN dérmico em dedo do pé de um homem de 40 anos, presente desde o nascimento. Devido às suas características clínicas inespecíficas, o MBN pode ser de difícil diagnóstico antes da biópsia e pode assemelhar-se a outras lesões benignas de partes moles. A avaliação histopatológica e imuno-histoquímica é, portanto, essencial para o estabelecimento de um diagnóstico preciso. O conhecimento dessa entidade incomum e sua inclusão no diagnóstico diferencial de pequenas lesões subcutâneas de consistência firme podem facilitar o manejo adequado e contribuir para evitar erros diagnósticos.

Palavras-chave: Mixoma; Neurotecoma; Dedos do pé

Case Report

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CASE REPORT

A 40-year-old man presented with a painless swelling over the dorsolateral aspect of the right fourth toe that had been present since birth and had progressively increased in size. The lesion had become painful approximately 1 year before presentation. There was no history of trauma, bleeding from the lesion, or similar swellings elsewhere on the body.

Physical examination revealed a tender, firm-to-hard, nonpulsatile, noncompressible, smooth, lobulated subcutaneous swelling measuring 2×1 cm over the dorsal aspect of the right fourth toe. The swelling was adherent to the overlying skin but did not restrict toe movement (Figure 1). No similar lesions were identified elsewhere, and there was no regional lymphadenopathy.

Plain radiography of the foot was unremarkable. Fine needle aspiration cytology indicated a benign spindle cell neoplasm. The lesion was completely excised on an outpatient basis under digital block anesthesia and sent for histopathologic examination. The resulting wound was primarily closed, with a small residual raw area left to heal by secondary intention.

Histopathologic examination of the excised tumor showed spindle-to-stellate cells with mild nuclear atypia, scant eosinophilic cytoplasm, and bipolar and multipolar processes. The cells were embedded in a fibrillary myxoid stroma, with no areas of necrosis or atypical mitoses (Figures 2 and 3).

At 2-year follow-up, there was no evidence of local recurrence (Figure 4).

DISCUSSION

Neurothekeomas were initially described as benign cutaneous tumors of nerve sheath differentiation; however, dermal

nerve sheath myxoma (NSM) was subsequently recognized as a distinct entity. Further studies demonstrated subtle morphologic differences, distinct immunohistochemical profiles, and differing ultrastructural features between these two tumor types. Reported cases of NSM have involved several anatomic sites, including the extremities (finger, knee, and pretibial region), neck,¹ and tongue. NSM most commonly occurs during the third and fourth decades of life, with a male-to-female ratio of approximately 1:1. In contrast, neurothekeomas predominantly affect the upper extremities and head and neck region, typically during the second and third decades of life, with a female predominance.²

Grossly, NSMs usually present as firm, slow-growing, painless nodules or masses measuring 0.4 to 4.5 cm. On cut section, they may contain multiple small, well-demarcated, translucent, whitish mucoid nodules. Histologically, these tumors have a characteristic multilobular structure, with individual lobules composed of spindle and stellate cells arranged in swirling, lamellar, and concentric patterns. The cells are embedded in an abundant myxoid stroma and generally exhibit mild nuclear pleomorphism and rare mitosis. Lesions may involve the epidermis, dermis, or subcutaneous tissue.³

Immunohistochemically, the tumor cells are usually strongly positive for S100 protein, glial fibrillary acidic protein (GFAP), and vimentin. Epithelial membrane antigen (EMA) is often absent or limited to a small number of perineural cells.⁴ In our patient, S100 staining was diffusely positive in spindle cells, whereas EMA was diffusely expressed in the perineural cells, which were more numerous at the periphery of the lesion, indicating bidirectional schwannomatous and perineural differentiation. A similar pattern of bidirectional differentiation was previously reported by Zamecnik et al.⁵ Electron microscopic



Figure 1: Dermal nerve sheath myxoma over the distal aspect of the right fourth toe

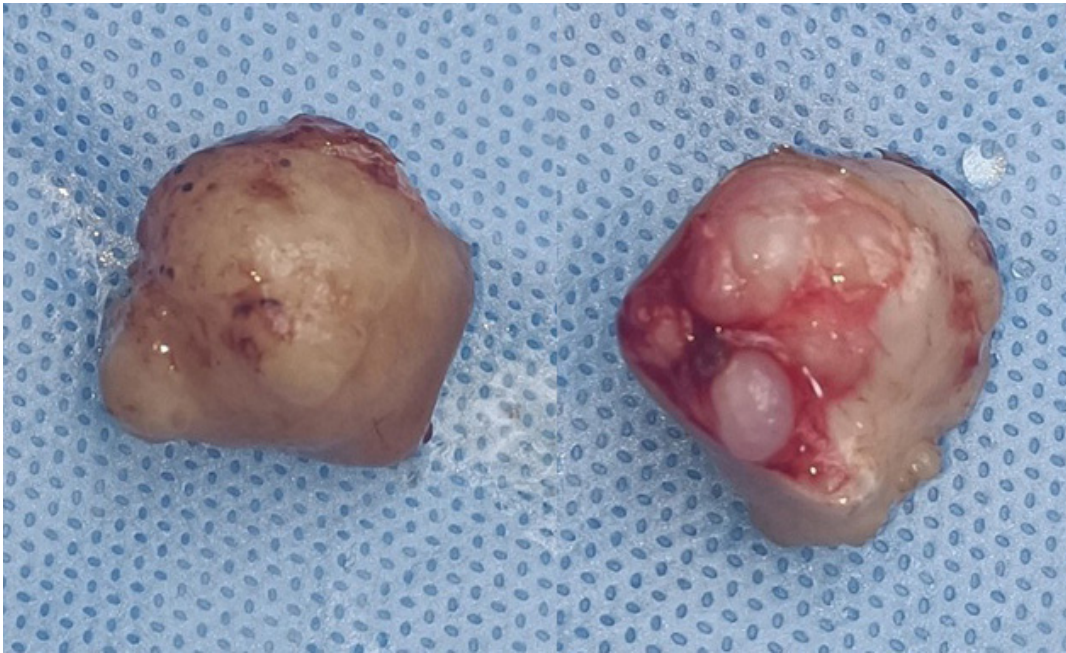


Figure 2: Gross appearance of the excised specimen

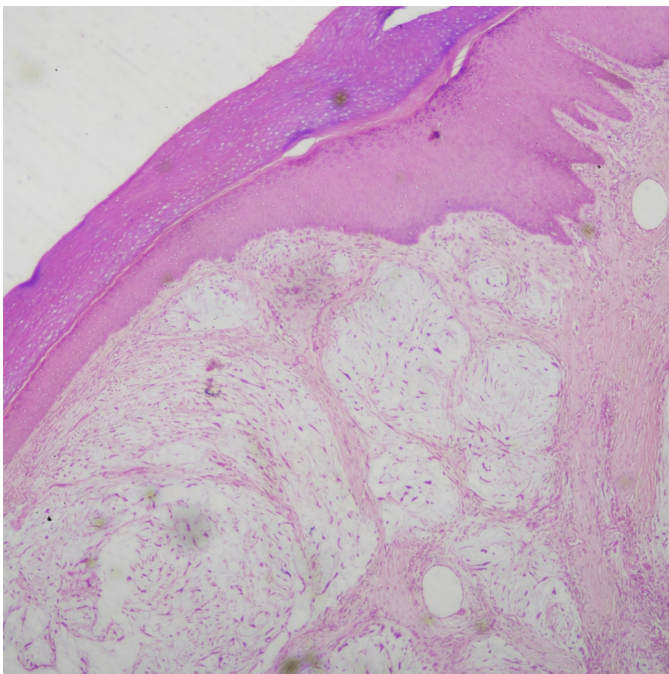


Figure 3: Histopathology showing hyperkeratotic stratified squamous epithelium with focal acanthosis and well-circumscribed myxoid lobules in the dermis containing fibrous tissue and spindle-to-stellate neoplastic cells (hematoxylin and eosin, ×10)

findings in NSM include single or duplicated external laminae surrounding the cells, desmosome-like junctions, cytoplasmic microfilaments, myelin figures, and interdigitating cytoplasmic processes.⁶



Figure 4: Clinical appearance at 2-year follow-up, with no evidence of local recurrence

More recent molecular evidence further supports the distinction between NSM and neurothekeoma. A gene expression study comparing the two tumors demonstrated that neurothekeoma expresses genes predominantly encoding glycoproteins and metalloproteinases associated with macrophages and fibroblasts. Differential expression of the S100B gene was also observed between NSM and neurothekeoma. These findings provide additional evidence that NSM is a neoplasm of peripheral nerve sheath origin and is distinct from neurothekeoma.³

Complete surgical excision is usually the treatment of choice for symptomatic lesions, as incomplete excision is associated with a high risk of local recurrence.⁵ The histopathologic differential diagnosis of NSM includes several myxoid tumors, particularly plexiform neurofibroma, myxoid neurofibroma, myxoid schwannoma, extraneural perineurioma, and cutaneous, juxta-articular, and intramuscular myxomas.⁷ Cutaneous myxomas involving the dermis and subcutis may similarly consist of multiple myxoid masses containing spindle-to-stellate neoplastic cells. However, cutaneous myxomas are positive for CD34 and negative for S100 protein, providing a useful immunohistochemical distinction from NSM.⁸

CONCLUSION

Dermal NSM is a benign myxoid neoplasm of peripheral nerve sheath origin. Its cytologic differential diagnosis includes

neurothekeoma, myxoid neurofibroma, plexiform neurofibroma, myxoid schwannoma, extraneural perineurioma, cutaneous myxoma, intramuscular and juxta-articular myxomas, cutaneous mucinosis, and hamartomas. These entities may share cytologic features, including myxoid material and benign spindle-to-stellate cells, making histopathologic and immunohistochemical evaluation essential for establishing an accurate diagnosis.⁹

Preoperative diagnosis can be challenging because of the lack of specific clinical or imaging features. This case highlights the importance of considering dermal NSM in the differential diagnosis of a long-standing myxoid soft-tissue lesion at an unusual site, such as the toe. Complete surgical excision is important because incomplete removal may result in local recurrence. Therefore, clinical follow-up is warranted, particularly after incomplete excision or when there is concern for residual disease.¹⁰ ●

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Statistical analysis, Approval of the final version of the manuscript, Conception and design of the study, Preparation and writing of the manuscript, Acquisition, analysis and interpretation of data, Effective participation in the conduct of the study, Intellectual participation in the propaedeutic and/or therapeutic approach to the cases studied, Critical review of the literature, Critical revision of the manuscript.

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