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CASE REPORT

Nerve sheath myxoma in the upper extremity: a rare case report

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Abstract
Nerve sheath myxoma (NSM) is a rare benign tumor originating from peripheral nerves. We present a case of NSM in the upper extremity. A 67-year-old female presented with painful nodular bulging in the elbow. Magnetic resonance imaging and ultrasonography revealed diagnostic features. Surgical excision was performed and histopathological examination confirmed the diagnosis. Our case contributes to the understanding of NSM's clinical presentation, imaging characteristics, and management strategies of NSM.

Keywords Nerve sheath myxoma · Peripheral nerve tumor · Neurothekeoma · Benign tumor

Introduction

Nerve sheath myxoma (NSM) is a rare benign tumor originating from the perineural "Schwann" cells of peripheral nerves [1]. It commonly affects the lower limbs of young adults, with a peak incidence in their thirties [2, 3]. First described by Harkin and Reed in 1969 [1, 3], NSM has been a subject of controversy since its inception. The term neurothekeoma, introduced by Gallager and Helwig in 1980 [4], was initially considered a variant of NSM [2, 3]. However, recent studies by Sheth et al. distinguished between these two entities based on genetic expression and cell origin [5]. Debates have revolved around whether NSM is a true nerve sheath tumor, its specific cell of origin (e.g., perineurial cell, fibroblast, "fibrohistiocytic" cell, or Schwann cell), and its relationship to neurothekeoma, as described by Gallager and Helwig in 1980 [1–4].

In this study, we discuss the imaging and histopathological characteristics of this rare tumor.

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Case report

Patient information

A 67-year-old female presented with painful nodular bulging of the elbow.

Clinical presentation

The patient reported pain and progressive swelling of the arm and elbow.

Diagnostic assessment ultrasound analysis revealed a hypoechoic oval-shaped nodular lesion in continuity with the supinator muscle, close to the deep radial and interosseous nerves (Fig. 1). MRI analysis showed an oval-shaped mass, with intermediate signal in T1, prominent high signal in T2/STIR, with thin septae close to the radial nerve, which was thickened and had an increased T2 signal (Fig. 2).

After diagnostic ultrasonography and magnetic resonance imaging, the patient was referred for surgical removal of the mass (Fig. 3).

A histopathological study identified a mesenchymal lesion composed of spindle cells with eosinophilic and poorly delimited cytoplasm, presenting homogeneous chromatin nuclei elongated to wavy, forming arrangements in cellular bundles dispersed amidst a large amount of myxoid matrix—findings suggestive of nerve sheath myxoma (Fig. 4).

Management and treatment

Surgical excision was performed because of the characteristics observed on imaging.

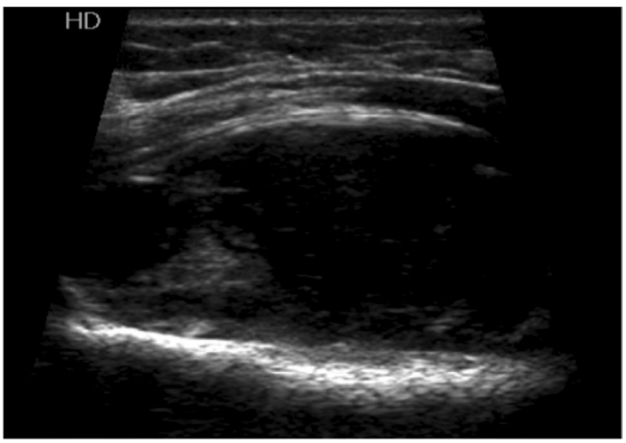

Fig. 1 Hypoechoic, heterogeneous oval-shaped mass, with partially defined limits, with no significant blood flow in Doppler analysis, in the middle of the supinator muscle fibers, close to the cortical bone of the radius and close to the radial nerve

Follow-up and outcome

The patient had an uneventful postoperative course.

Discussion

Benign nerve sheath myxoma (NSM) is a rare neuroectodermal tumor that was first described in 1969 [1]. It predominantly occurs in young adults with a peak incidence between 30 and 40 years of age and commonly affects the lower limbs [9]. In our case, the patient was 67 years old. Microscopically, NSM exhibits a distinctive, well-circumscribed, multi-lobulated growth pattern with loosely arranged spindle and stellate cells, often showing S-100 protein positivity within copious stromal mucin partitioned by collagen fibers [2, 4].

The diagnosis of NSM is often challenging due to its radiologic resemblance to other soft tissue lesions, including schwannomas and hemangiomas on ultrasound and MRI [15, 16]. The standard management continues to be total surgical excision followed by histopathological analysis, with complete resection being crucial to minimize recurrence risk [1, 2, 8, 9]. Nevertheless, conventional imaging modalities such as ultrasound and MRI frequently lack specificity, making histological and immunohistochemical evaluation essential, as they remain the diagnostic gold standard [4].

Nerve sheath myxomas exhibit characteristic immunohistochemical findings, confirming their status as peripheral nerve sheath tumors [3, 4, 7, 8]. These tumors commonly present as superficial, highly myxoid, multinodular masses with peripheral fibrous borders containing spindled, stellate-shaped, ring-shaped, and epithelioid Schwann cells [4, 15, 16]. Immunoreactivity for S-100 protein and GFAP, along with other neural markers, is typical [3, 4].

Neurothekeoma and nerve sheath myxoma (NSM) were historically considered related entities due to overlapping histologic characteristics and presumed neural origin. However, contemporary studies using immunohistochemistry and molecular markers have established that they are distinct lesions [4, 17]. Neurothekeomas, particularly the cellular subtype, tend to affect younger individuals, with a female predominance, and typically present as superficial dermal or subcutaneous nodules [18]. Unlike NSMs, neurothekeomas are characteristically negative for S-100 protein and may express markers such as NKI/C3, CD10, and MiTF, suggesting a fibro-histiocytic rather than Schwannian differentiation [4, 5]. Conversely, NSMs are typically S-100 positive, affect a slightly older demographic, and show a multinodular, myxoid architecture on histology,

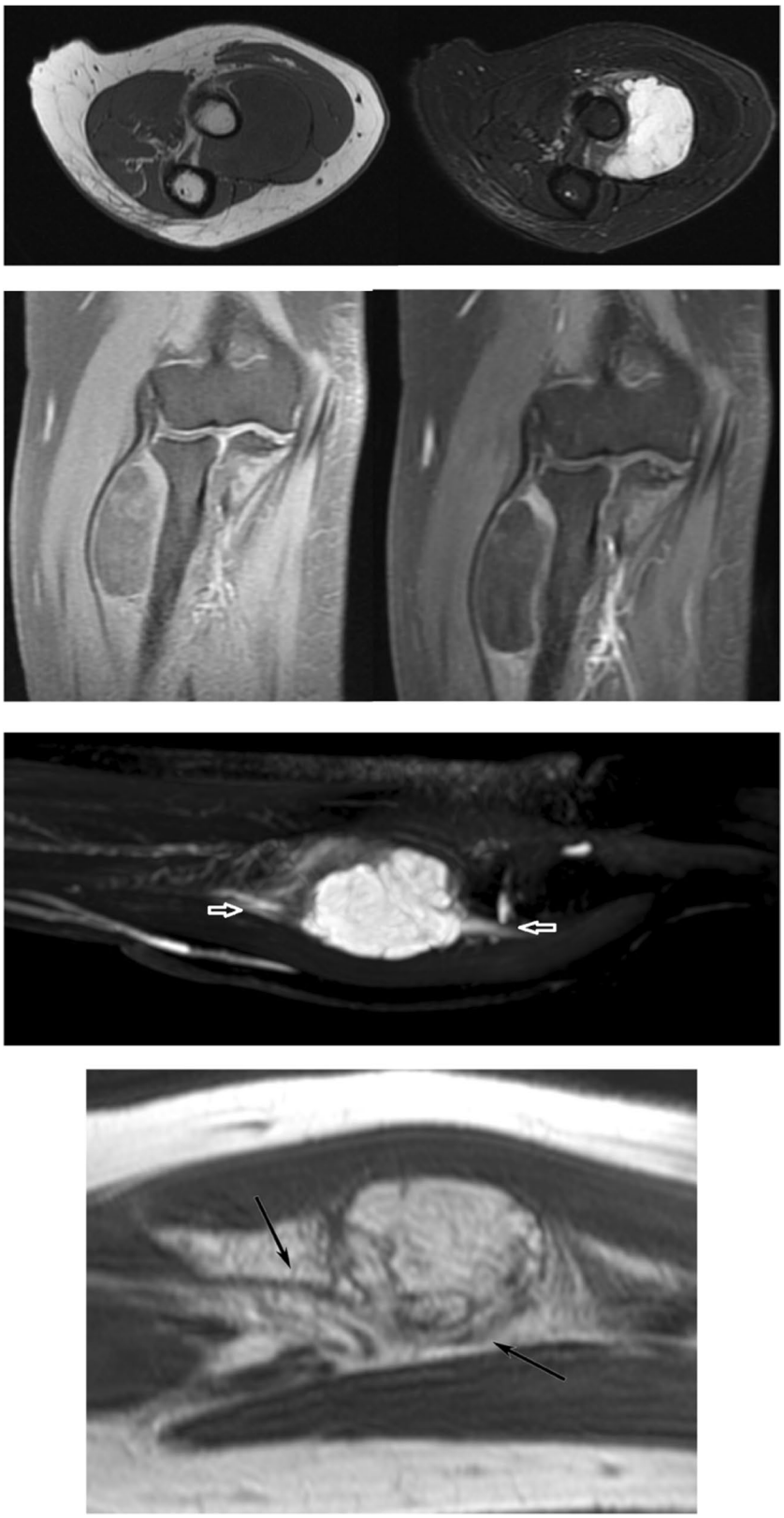

Fig. 2 MRI findings of nerve sheath myxoma of the radial nerve in the upper extremity. The MRI images were as follows: axial T1, axial STIR, coronal T1 fat sat sequences before and after contrast enhancement, and volumetric reformatting with inversion recovery. Expansive lobulated formation with a signal similar to the supinator muscle in T1, with a slightly heterogeneous hypersignal in STIR, and bands of low signal in between. Post-contrast sequences showed slight enhancement, especially internally and during septations. T1 and volumetric reformatting shows a close relationship with the radial nerve, especially with the deep branch

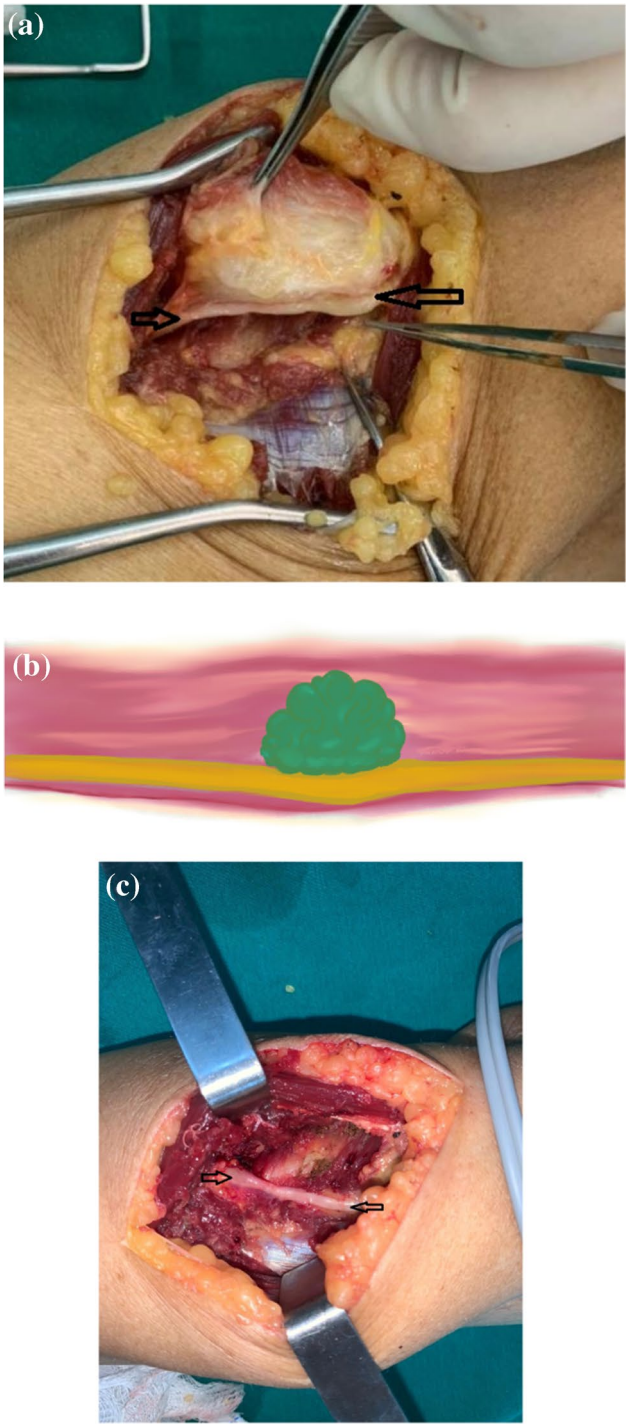

Fig. 3 Surgical piece identified next to the radial nerve, appointed with arrows (**a**). The illustration shows the proximity of the tumor lesion (green) to the adjacent nerve (yellow) (**b**). Finally, a photograph shows complete removal of the tumor at the end of the surgery, with preserved radial nerve signalized by the black arrows (**c**)

with abundant mucinous stroma and scattered spindle cells [17, 19]. Recognizing these histopathological and immunohistochemical distinctions is essential for accurate diagnosis and appropriate management.

The differential diagnosis of NSM includes various soft tissue tumors, including other types of neurothekeoma, superficial angiomyxoma, neurofibroma, schwannoma, perineural cell tumors, chondroma of the soft parts, and superficial acral fibromyxoma. NSM may ultimately be considered a distinct subtype of schwannoma or neurofibroma [1]. Although less frequently documented, the cytological features of NSM include spindle stellate cells and epithelioid cells arranged in loose clusters within a metachromatic myxoid background [5, 6]. Histopathological sections revealed well-defined multinodular tumors composed of myxoid nodules separated by fibrous septae, with nodules containing benign spindle stellate and epithelioid cells embedded in the myxoid stroma. Immunohistochemical markers such as S100, glial fibrillary acidic protein, vimentin, and collagen type IV are helpful for diagnosis, with negative EMA staining [4].

This study highlights the importance of understanding this rare tumor with an atypical presentation in the upper limb. The characteristics presented in this case are classic, both in imaging and histology, and help facilitate the recognition and evaluation of this tumor.

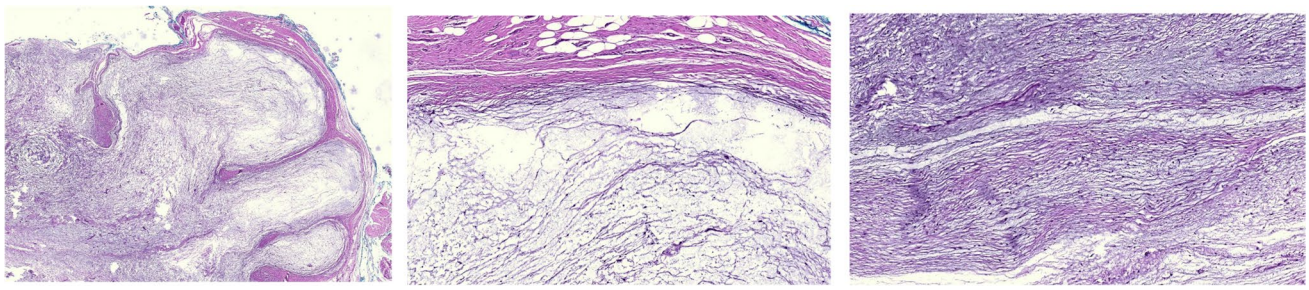

Fig. 4 The sections show a mesenchymal lesion composed of spindle cells with eosinophilic and poorly delimited cytoplasm, presenting homogeneous chromatin nuclei elongated to wavy, forming arrangements in cellular bundles dispersed amidst a large amount of myxoid matrix. The neoplasm forms nodular arrangements separated by fibrous bundles. There were some mast cells, and no mitotic images were observed

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Data availability Not applicable.

Declarations

Ethics approval and consent to participate Not applicable.

Competing interests The authors declare no competing interests.

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