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The role of imaging in isolated benign peripheral nerve tumors: A practical review for surgeons

Rôle de l'imagerie dans les tumeurs nerveuses périphériques bénignes isolées : étude pratique pour les chirurgiens

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Abstract

The diagnosis of nerve tumor(s) must be suspected in all cases of tumefaction or pain on the path of a nerve exacerbated by percussion. Solitary nerve tumors are primarily schwannomas, but other rare tumors may be present such as intraneural ganglion cysts of controversial origin. Preservation of nerve continuity is the underlying goal for any surgical procedure, irrespective of the type of tumor. Therapeutic outcomes are closely linked to tumor resectability; in most patients, the resectability of the tumor, its type and benignity can be predicted based on medical imaging. Comparison with the clinical examination and case-based reasoning is crucial. Consequently, the aim of this review was to examine the role of imaging in isolated benign peripheral nerve tumors, and provide the surgeon with a practical guide for its application in predicting the nature and resectability of nerve tumors.

Keywords: Peripheral nerve tumors; Neurofibroma; Schwannoma; Intraneural mucoid pseudocyst; MR nerve imaging; Ultrasound

Résumé

Le diagnostic de tumeur nerveuse doit être évoqué en cas de tuméfaction ou de douleur sur le trajet d'un nerf que la percussion réveille. Les tumeurs nerveuses isolées sont en majorité des schwannomes ; les autres tumeurs sont beaucoup plus rares et d'une grande diversité histologique, comprenant les pseudo-kystes mucoïdes des nerfs dont l'origine est controversée. Quel que soit le type de tumeur, le traitement chirurgical vise à respecter la continuité nerveuse. Le pronostic est étroitement lié au caractère extirpable de la lésion. L'analyse radiologique permet, chez la plupart des patients, de prévoir l'extirpabilité, la nature et la bénignité des tumeurs. La confrontation avec l'examen clinique et la casuistique est essentielle. Le but de cette revue était d'analyser le rôle de l'imagerie en cas de tumeur nerveuse périphérique isolée et de faire une synthèse des critères cliniques et paracliniques qui permettront au chirurgien de prévoir au mieux le caractère extirpable et la nature des lésions.

Mots clés : Tumeurs des nerfs périphériques ; Neurofibrome ; Schwannome ; Pseudokyste mucoïde des nerfs ; IRM ; Échographie

1. Introduction

Benign tumors of the peripheral nerves (BTPN) are rare tumors that can occur either as isolated lesions [1] or in multiple forms [2], as in neurofibromatoses (not included in this review).

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Surgical treatment and prognosis vary greatly depending on whether the tumor is resectable, with the ultimate goal of the surgical procedure being the preservation of nerve continuity. Consequently, the aim of this review was to examine the role of imaging in isolated benign peripheral nerve tumors, and to provide the surgeon with a practical guide for its application in predicting the nature and resectability of nerve tumors.

The general characteristics of benign nerve tumors (including patient presentation and radiological aspect) and specific features of the various tumor types are successively reviewed.

2. General characteristics of benign tumors of peripheral nerves

BTPN represent approximately 1% of all soft tissue tumors [1]. While more frequent in the upper limb and mostly affecting large nerve trunks [3], ultimately any nerve can be affected. Eighty percent of BTPN are schwannomas [1,4]. BTPNs are rooted in either the nerve tissue or the non-neural elements of the nerve. From a practical and prognostic perspective, resectable tumors should be distinguished from non-resectable ones (Table 1). Essentially, resectable tumors grow eccentrically from the nerve, pushing back the fascicle groups without penetrating the perineurium and can be enucleated without disrupting nerve continuity. On the other hand, non-resectable tumors infiltrate all constituent elements of the nerve, with nerve fiber damage always occurring upon complete removal. Consequently, an epineurotomy for decompression is recommended in case of nerve compression symptoms [1,5].

3. Patient presentation

Most patients present with swelling over the course of a superficial nerve or, more rarely, localized pain and/or paresthesia revealed by percussion if a deeper nerve is involved [5,6]. BTPNs are solely responsible for neurological deficits by compressive intraneural damage (non-resectable tumors), development in a narrow anatomical groove or development spanning many years (resectable tumors). In some cases, its discovery is made during surgery or imaging for nerve entrapment syndrome in young adults. More rarely, the diagnosis is made secondary to nerve resection performed erroneously for a diagnosis of lymphadenopathy (especially in the supraclavicular fossa), synovial cyst or resectable tumor [7].

4. Radiological characteristics of BTPN

Ultrasound (US) and magnetic resonance imaging (MRI) [8,9] are the reference imaging modalities for the diagnosis of BTPN. Computed tomography (CT) is rarely performed because of its low resolution for soft tissues.

4.1. US vs. MRI

As a general rule, peripheral nerves are more visible when surrounded by tissues of a different echostructure. Peripheral nerves appear as tubular structures made of hypo-echoic nerve fascicles embedded in a hyperechoic connective tissue corresponding to the epineurium. Longitudinally, US images present a fascicular pattern, and transversely, fascicles appear rounded or oval in shape, giving the nerve the typical honeycomb appearance [10,11]. US diagnosis of a nerve tumor is based on the existence of a mass in continuity with the nerve at its proximal and distal poles [12].

While US eliminates the vast majority of false tumor lesions and allows for differentiation between lymphadenopathy, neuroma or liquid tumor (hematoma, abscess or thrombosed aneurysm), it is more operator-dependent than MRI with nerve lesions rarely being specific. Color Doppler ultrasound allows an accurate assessment of the presence of intralesional flow and helps in differentiating solid and cystic lesions [12]. Its dynamic method of study enables the physician to analyze the entire nerve course and look for other remote tumor [10]. US-guided pressure through the transducer on the mass can reproduce local pain and peripheral paresthesia (the so-called ‘‘ultrasonographic Tinel sign’’). This can be very helpful when assessing small peripheral lesions. But ultimately, US cannot replace MRI for determining the topography, the exact extent of tumor formation or the presence of invasion [13]. The boundary between the tumor and the nerve is sometimes difficult to

Table 1
Classification and origin of main single benign peripheral nerve tumors.

Benign peripheral neural sheath tumors (PNSTs)		Peripheral non-neural sheath tumors			
Schwannoma[a]	Neurofibroma	Intraneural hemangioma	Fibrolipomatous hamartoma	Intraneural lipoma[a]	Intraneural ganglion cyst
Neurinoma[b]		Perineural hemangioma[b]	Fatty infiltration[b]		Intraneural mucoid pseudocyst[b]
Neurilemmoma[b]		Schwann sheath hemangioma[b]	Lipofibroma[b]		Nerve sheath ganglion[b]
		Intrinsic hemangioma of the peripheral nerve[b]	Intraneural lipofibroma[b]		Intraneural synovial cyst[b]
			Neuro-fibrolipoma[b]		
		(Extrafascicular intraneural hemangioma)[a]	Fibro-fatty proliferation[b]		

Traumatic neuromas and pseudoneuromas (following nerve injury) are excluded. Desmoid tumors and neuromuscular choristoma (benign triton tumor) of controversial origin are excluded.

[a] Resectable tumors.

[b] Equivalent nerve tumor terms.