High-resolution ultrasonography and shear-wave sonoelastography of a cystic radial nerve Schwannoma

Patrick J. Battaglia¹ · Vivian Carbone-Hobbs² · Gary M. Guebert¹ · Susan E. Mackinnon³ · Norman W. Kettner¹

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Abstract Peripheral nerve tumors are often evaluated with magnetic resonance imaging (MRI), although there are many advantages offered with high-resolution ultrasonography (HRUS). This case report emphasizes the value of HRUS in the diagnosis and management of a patient with a cystic radial nerve Schwannoma. In addition, information on tumor stiffness, obtained with shear-wave sonoelastography (SWE), is presented.

Keywords Schwannoma · Neurilemmoma · Diagnostic ultrasound · Sonoelastography

Sommario I tumori dei nervi periferici vengono spesso studiati con la risonanza magnetica (MRI), anche se l'ecografia ad alta risoluzione (HRUS) presenta molti vantaggi. Questo case report sottolinea il valore della HRUS nella diagnosi e nella gestione di un paziente con uno Schwannoma cistico del nervo radiale. Vengono inoltre riferite le informazioni sulla stiffness del tumore ottenute con la shear wave elastosonografia.

Introduction

Benign peripheral nerve sheath tumors include Schwannoma (neurilemmoma) and neurofibroma. These tumors have equal prevalence each comprising approximately 5% of benign soft-tissue tumors [1]. In a review of upper extremity benign peripheral nerve sheath tumors at one institution ($n = 110$), Schwannomas were less common than neurofibromas (29 vs 71%, respectively) [2]. Schwannomas of the upper extremity occur most frequently in the ulnar nerve followed by the median nerve [3, 4]. Schwannoma of the radial nerve or its terminal branches is rare compared to these other locations. If a peripheral nerve Schwannoma evades early diagnosis, it may undergo cystic degeneration and mimic a malignancy at imaging [4, 5]. The term ancient Schwannoma has been used in some instances to describe a Schwannoma that has undergone degeneration and is characterized histopathologically by cyst formation, calcification, hemorrhage, and fibrosis [6].

While magnetic resonance imaging (MRI) is often utilized for the evaluation of soft-tissue masses, high-resolution ultrasonography (HRUS) is being increasingly employed to study lesions of the peripheral nervous system [3, 7, 8]. Of note, sensitivity for detecting peripheral nerve pathology is greater with HRUS compared to routine MRI, while specificity is equally high for both modalities [9]. Shear-wave sonoelastography (SWE) is an ultrasound function that permits quantification of tissue stiffness and is currently used clinically in various body systems to differentiate benign and malignant entities. For example, SWE stiffness values discriminate between benign and malignant thyroid nodules, with malignancy demonstrating significantly stiffer tissue properties [10, 11]. Although many imaging features exist which may differentiate benign and malignant peripheral nerve sheath tumors,

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✉ Patrick J. Battaglia
patrick.battaglia@logan.edu

¹ Department of Radiology, Logan University, 1851 Schoettler Road, Chesterfield, MO 63017, USA

² Private Practice, Arnold, MO 63010, USA

³ Shoenberg Professor and Chief Division of Plastic and Reconstructive Surgery, Washington University School of Medicine, 660 S Euclid Ave, St. Louis, MO 63110, USA

when a tumor has undergone degenerative change or is atypical in appearance (e.g., cystic degeneration), this distinction becomes less clear. Therefore, additional information, such as tissue stiffness with SWE, may have clinical value. This case report emphasizes the value of HRUS in the diagnosis of a cystic radial nerve Schwannoma. In addition, this is the first report to describe SWE stiffness values of a peripheral nerve sheath tumor.

Case description

Patient consent for publication was obtained. A 47-year-old female presented with a palpable right lateral elbow mass of greater than 1-year duration. The mass was firm and non-mobile and the patient also had pain and paresthesia that radiated distally along the dorsum of the forearm and hand. At examination, there was no atrophy of the forearm muscles and no strength deficit. The patient reported receiving a diagnosis of a subcutaneous lipoma 1 year prior from another provider. A non-contrast MRI (1.5 Tesla, GE Healthcare, Wauwatosa, WI) of the right elbow demonstrated a mixed signal intensity soft-tissue mass located between the brachioradialis and brachialis muscles at the level of the elbow. Subsequently, an HRUS examination of the lateral elbow was performed with a GE Logiq E9 system (GE Healthcare, Wauwatosa, WI) using a linear transducer (ML6-15) operating at 15 MHz. The HRUS clearly displayed that the mass originated from the periphery of the radial nerve (Figures 1, 2). The mass measured 1.49×1.95 cm in long axis and 2.26×1.50 cm in short axis. There was a prominent internal hypoechic cyst and multiple smaller cystic foci. External transducer force demonstrated that the mass was minimally compressible. Cine imaging verified the specific origin of the mass arising from the deep branch of the radial nerve just caudal to the radial nerve bifurcation (Supplemental media). Power Doppler imaging displayed surrounding peripheral and internal vascularity. Examination with SWE revealed the internal elastic properties of

the mass to be soft with a stiffness value ranging from 24 to 30 kPa (kilopascals) (Fig. 3). There was no observable difference in tissue stiffness between long- and short-axis imaging. In addition, the prominent internal cystic areas were void of SWE information. Based on the imaging findings, the likely diagnostic consideration was a benign peripheral nerve sheath Schwannoma and the patient was referred for surgical management. Surgical intervention confirmed the mass originated from the deep branch of the radial nerve just proximal to the arcade of Frohse. Microsurgical instrumentation and magnification were used to excise the tumor and decompress the deep branch of the radial nerve (Fig. 4). In addition, tenotomy of the extensor carpi radialis brevis was performed for further nerve decompression. Pathologic analysis of the specimen was consistent with a benign peripheral nerve Schwannoma (Figs. 5, 6). The patient tolerated the surgical procedure well and there was no damage to the radial nerve.

Discussion

This case highlights the value of HRUS in evaluation of peripheral nerve tumors. In addition, this is the first report to present tissue stiffness values of a benign peripheral nerve Schwannoma.

The value of HRUS in peripheral nerve pathology evaluation has been well described [3, 7–9]. Zaidman et al. demonstrated that HRUS had greater sensitivity (93 vs 67%) for detection of peripheral nerve lesions compared to MRI [9]. In that same study, both HRUS and MRI had equal specificities (86%), but HRUS had greater accuracy (93 vs 70%) [9]. Interestingly, these authors described that some peripheral nerve lesions not detected on the MRI examinations were outside of its field of view. This seems counterintuitive in that HRUS would offer a broader field of view compared to MRI. However, an operator conducting an HRUS examination has the freedom to image structures across many body regions (e.g., neck, axilla, arm, and forearm) with ease. In addition, the operator is not

Fig. 1 **a** Coronal T2-weighted MRI and **b** long-axis HRUS of a right radial nerve Schwannoma (arrows). The MRI has been rotated to align with the HRUS anatomy and the *left side* of each image in

cranial. Note how the HRUS better displays continuity with the nerve (arrowheads). This is an important feature when establishing benignity of a peripheral nerve tumor

Fig. 2 **a** Axial T2-weighted MRI and **b** short-axis HRUS of a right radial nerve Schwannoma (arrows). Note how only the HRUS displays the eccentric origin of the tumor from the nerve (arrowheads). This is an important feature when differentiating between

Schwannoma and neurofibroma. In addition, note the prominent internal cyst (asterisk) that is hyperintense with T2-weighting and hypoechoic at HRUS

Fig. 3 **a** Long-axis and **b** short-axis SWE images of a right radial nerve Schwannoma (arrows on the gray-scale images in the left panel). Note the tumor stiffness to be soft, with representative images showing approximate kPa values of 24 in the *long-axis* and 27 in the *short-axis*

confined to the traditional anatomic planes. This is especially beneficial as peripheral nerves have nonlinear courses throughout the extremities. In support of the work of Zaidman et al., Toia et al. examined a cohort of patients with various peripheral nerve pathologies (e.g., tumor, entrapment, and traumatic injury) and found that HRUS had a key diagnostic role in the presence of equivocal electrodiagnostic findings in 16% of cases studied. These authors, therefore, concluded that HRUS should be the first-line imaging modality for masses of suspected nerve

origin [12]. Conversely, specific to peripheral nerve tumors of the upper extremity, Nilsson et al. found that MRI could only localize and diagnose a nerve tumor in 75% of cases [13]. In our case, the patient's MRI demonstrated a non-specific soft-tissue mass without a discrete neurogenic origin, whereas the HRUS accurately demonstrated communication of the mass with the radial nerve. There are many other advantages of HRUS compared to routine MRI in the evaluation of peripheral nerves that have been nicely summarized in a recent review by Ali et al. [8]. These

Fig. 4 **a** Pre-excision and **b** post-excision intraoperative photographs. In **a**, note the relationship of the Schwannoma (arrow) to the radial nerve (arrowheads). In **b**, the Schwannoma (arrow) has been removed, while its capsule (dashed arrow) can be seen adjacent to the radial nerve (arrowheads)

Fig. 5 **a** Photomicrograph at $\times 0.25$ magnification and **b** short-axis SWE image of a right radial nerve Schwannoma. In **a**, the organized elements typical of an Antoni A pattern are stained with hematoxylin and eosin. A large unstained cystic/mucoid area (asterisk) is also seen consistent with an Antoni B pattern and typical of degenerative change. Note that in **b**, the area of cystic degeneration (asterisk) is void of SWE information

advantages include: greater axial in-plane resolution, ability to image near metal implants, portable examination (e.g., for use in clinic and at operation, ease of contralateral comparison), more time and cost efficient, no contraindications (e.g., cardiac pacemakers), improved patient satisfaction, and ability for the examiner to obtain immediate feedback during the examination [8]. Specific to tumor imaging, the ability for HRUS to establish the origin of a mass from a nerve is paramount, and the ability to determine its precise origin (eccentric vs central) may help

differentiate between Schwannoma (eccentric) and neurofibroma (central) [6, 8]. In addition, HRUS in the perioperative and intraoperative settings may help avoid iatrogenic nerve injury [14], and HRUS is well suited for routine postoperative evaluation [8]. An additional advantage of HRUS that we utilized in this case was with dynamic scanning in a craniocaudal direction to permit exact determination of the nerve of origin being the deep branch of the radial nerve. Importantly, there are more sophisticated MRI sequences that may be used that are

Fig. 6 Photomicrograph at $\times 100$ magnification of a right radial nerve Schwannoma with hematoxylin and eosin staining demonstrates elongated cells with a fascicular arrangement typical of an Antoni A pattern. Note unstained areas of tissue consistent with cystic/mucoid change (asterisk)

better than routine MRI to evaluate peripheral nerves. Perhaps, the most utilized of these sequences, diffusion tensor imaging with tractography, has been shown to demonstrate native nerve fascicles surrounding a peripheral nerve tumor [14, 15]. This may prove especially important in tumors of the brachial or lumbosacral plexus or other, deeper located peripheral nerves that are not as easily resolved with HRUS. The addition of intravenous gadolinium contrast to routine MRI sequences may also be performed. Both benign and malignant peripheral nerve sheath tumors enhance avidly after contrast administration, whereas the presence of a central non-enhancing focus (“target sign”) suggests benignity, and perilesional soft-tissue enhancement suggests malignancy [4, 16, 17]. These findings, however, are not pathognomonic and certain risks must be considered when intravenous gadolinium is utilized [18].

At HRUS imaging, Schwannomas are typically well-defined, homogeneous hypoechoic oval masses with poor vascularity [3]. With MRI, they are a homogenous mass with low signal on T1-weighted and high signal on T2-weighted sequences [4]. However, when degeneration of a Schwannoma has occurred, as seen in our case, the HRUS and MRI appearance is more heterogeneous with areas of

cystic degeneration, calcification, hemorrhage, and fibrosis [4, 6]. Therefore, the imaging appearance of a degenerative Schwannoma is striking and concerns for malignancy may arise [5]. However, certain imaging features exist which may help differentiate a benign Schwannoma that has undergone cystic degeneration from a malignant nerve sheath tumor. Imaging features suggesting benignity include: eccentric origin of the mass relative to the peripheral nerve, contiguity of the tumor with the nerve of origin, well-defined margins, and tumor size (benign tumor size 3.4 ± 2.5 cm; malignant tumor size 8.2 ± 3.1 cm) [5, 17]. At MR imaging, surrounding edema in the soft tissues is more suggestive of a malignant peripheral nerve sheath tumor [16, 17]. Since HRUS is likely to become the first-line imaging of extremity peripheral nerve tumors, continued knowledge of the HRUS appearances and properties of benign cystic Schwannomas would be valuable. In our case of a pathologically confirmed cystic Schwannoma, there were many HRUS features suggesting benignity including an eccentric origin, continuity with the peripheral nerve, and small tumor size. In addition, the stiffness of the tumor was soft, with a kPa range from 24 to 30. Although there is no literature regarding tissue stiffness values for peripheral nerve tumors, in other organs, malignancy presents as hardened tissue. For example, invasive ductal carcinoma in situ of the breast has SWE properties of approximately 70 kPa [19]. In addition, a soft SWE value (<50 kPa) combined with benign gray-scale US features may obviate need for biopsy and follow-up with a palpable breast mass in women less than 40 years [20]. In thyroid nodules, using 65 kPa as a cut-off value yields 76% sensitivity and 64% specificity in diagnosing malignancy (i.e., papillary thyroid carcinoma) [10]. In a recent pilot study of 15 malignant and 35 benign soft-tissue masses, Pass et al. found that tissue stiffness determined with SWE was lower in malignant masses [21]. This did not reach significance, however, and the authors also noted a few important limitations in their work, namely, sample size and incomplete SWE examination in certain malignant masses [21]. These authors also did not provide specific data on individual tumor type and SWE properties. However, one of their figures was of a cystic nerve Schwannoma, with soft SWE values displayed (1.08 m/s vs a mean of 2.20 m/s for benign lesions) [21]. The only report specific to US elastography of peripheral nerves is a case using qualitative strain-based elastography of a common fibular nerve Schwannoma [22]. In this report, elastography demonstrated that the Schwannoma was harder compared to surrounding muscle. Utilizing SWE would be preferred for clinical decision making as precise cut-off values for tissue stiffness, such as those used in the breast and thyroid, can be adhered to.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest to disclose.

Ethical approval All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000. All ethical standards were upheld during both the imaging and surgical procedures inherent in this case.

Informed consent All patients provided written informed consent to enrollment in the study and to the inclusion in this article of information that could potentially lead to their identification. Patient informed consent was obtained prior to publication.

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