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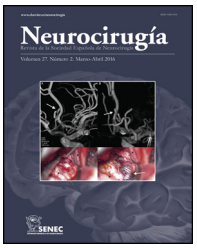

Case Report

Removal of a giant musculocutaneous nerve schwannoma under intraoperative neurophysiological monitoring: Case report video and review of the literature

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ABSTRACT

Schwannomas of the musculocutaneous nerve (MCN) are rare benign tumors of the peripheral nerve sheath. Due to their slow growth, they are often diagnosed late. In the upper limbs, schwannomas typically affect longer peripheral nerves at a distal level, making MCN cases uncommon. Ultrasound (US) and magnetic resonance imaging (MRI) are essential tools for early detection. While most schwannomas can be surgically removed without damaging the nerve, intraoperative neurophysiological monitoring (IONM) is critical if fascicular involvement exists.

We present a 73-year-old right-handed Jehovah's Witness with a history of mild polio affecting the right arm. MRI showed a slow-growing, cystic mass in the right biceps, originating from the MCN and suggestive of schwannoma. Surgery achieved gross total resection without nerve damage. Histopathology confirmed a cystic schwannoma.

In atypical proximal upper limb tumors, MCN schwannoma should be considered, with US/MRI crucial for diagnosis. IONM-assisted removal can minimize postoperative complications.

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Exéresis de un schwannoma gigante del nervio musculocutáneo bajo monitorización neurofisiológica intraoperatoria: comunicación de un caso en vídeo y revisión de la literatura

Palabras clave:
Musculocutáneo
Schwannoma
Monitoreo neurofisiológico

RESUMEN

Los schwannomas del nervio musculocutáneo son tumores benignos poco comunes de la vaina del nervio periférico. Debido a su crecimiento lento, suelen diagnosticarse tardíamente. En las extremidades superiores, los schwannomas típicamente afectan a los nervios periféricos más largos a nivel distal, lo que hace que los casos del nervio musculocutáneo sean infrecuentes. La ecografía (US) y la resonancia magnética (RM) son herramientas esenciales para su detección precoz. Aunque la mayoría de los schwannomas pueden ser resecados quirúrgicamente sin dañar el nervio, la monitorización neurofisiológica intraoperatoria (MNI) es fundamental si existe afectación fascicular. Presentamos el caso de un hombre de 73 años, diestro y Testigo de Jehová, con antecedentes de poliomielitis leve que afectaba la extremidad superior derecha. La RM evidenció una masa quística en el bíceps derecho, originada en el nervio musculocutáneo y sugestiva de schwannoma. La cirugía permitió una resección macroscópica completa sin daño al nervio. La histopatología confirmó el diagnóstico un schwannoma quístico. En los tumores proximales atípicos de la extremidad superior, debe considerarse el schwannoma del nervio musculocutáneo, como posibilidad diagnóstica, siendo fundamental la ECO/RM para su diagnóstico. La exéresis asistida por MNI puede minimizar las complicaciones derivadas de la cirugía.

Introduction

Schwannomas of the MNC are a rare but well-described entity impairing one of the shorter peripheral nerves of the upper limb, and therefore being less frequent than those originated in the median, ulnar or radial nerves which have longer trajectories.[1,2] The deep course of the musculocutaneous nerve[3,4] piercing the coracobraquialis muscle near to his point of insertion to the humerus running deep to the biceps brachii muscle, added to the benign slow-growing nature of the schwannoma allowing nerve fascicles adaptation, can drive to a big-size presentation[5,6] with mass-only symptoms.[7,8] Outside the setting of neurofibromatosis, most schwannomas of the upper limb are solitary.

These benign peripheral nerve sheath tumors exclusively composed by Schwann cells are usually suspected according to MRI scan[9,10] or ultrasonography,[11] showing tumor location, margins, and relationship to adjacent structures, and even specific features such as the target sign or the fascicular sign.

Gross total resection (GTR) of the tumor is normally feasible as schwannomas of the MCN are well encapsulated and surgical enucleation followed by careful dissection of the tumor capsule from the nerve is believed to be routinely possible without causing neurological deficit.[1,2,8,12]

However, a small percentage of these tumors have neural fascicular involvement and GTR cannot be achieved without adjuvant IONM, an effective tool that allows monitoring of MCN function and identification of neural tissue for a complete and safe resection.[13] GTR precludes relapse of benign sporadic upper limb schwannomas according to long-term treatment and follow-up data.[1,2]

Case report

A 73 year-old right-handed Jehova's witness was referred to our clinic with a chronic history through the years of a slowly growing, painless mass in the front aspect of the right proximal upper arm. As past medical history he suffered mild poliomyelitis sequelae from childhood in the right arm, and his wife was also operated on in our Department because of L4L5 spondylolisthesis. Physical examination revealed an elastic-hard, poorly mobile, non-tender 8 cms mass under the skin of the right arm (Fig. 1a). Apart from the second motoneuron paresis' signs with motor balance 4/5 impairing the right deltoid and biceps muscles lasting from polio, the rest of the neurovascular examinations, including Tinel's sign, were normal.

Laboratory data were within normal limits. MRI demonstrated an oval-shaped soft tissue mass measuring $8 \times 6 \times 5.5$ cm in the right biceps muscle, arising from the right MCN with depiction of the nerve entering and exiting the tumor. The mass appeared isointense to muscles on T1-weighted images and hyperintense on T2-weighted images[9] (Fig. 1b,c) with a rim of fat surrounding the mass being also observed (split fat sign on T1-weighted MR images oriented on the long axis of the affected limb, Fig. 1c).

Based on these findings, a preoperative diagnosis of MCN schwannoma was made.

The uneventful surgical excision was performed under general anesthesia and IONM of motor potentials evoked by transcranial stimulation (tcEMP), with biceps muscle recording and motor mapping of nervous structures performed using a bipolar stimulator.

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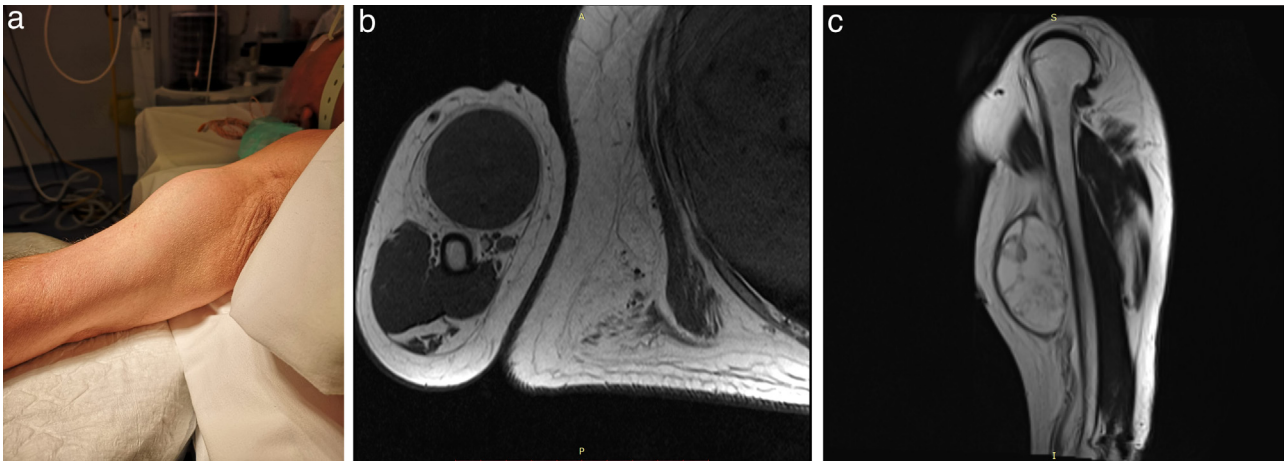

Figure 1 – (a) Eight centimeters mass in the right arm. (b) T1-weighted axial image of the isointense oval-shaped soft tissue mass. (c) T2-weighted sagital image showing an hyperintense cystic mass in the anterior and middle aspect of the right arm with inner walls and a rim of fat surrounding the mass (split fat sign oriented on the long axis of the affected limb).

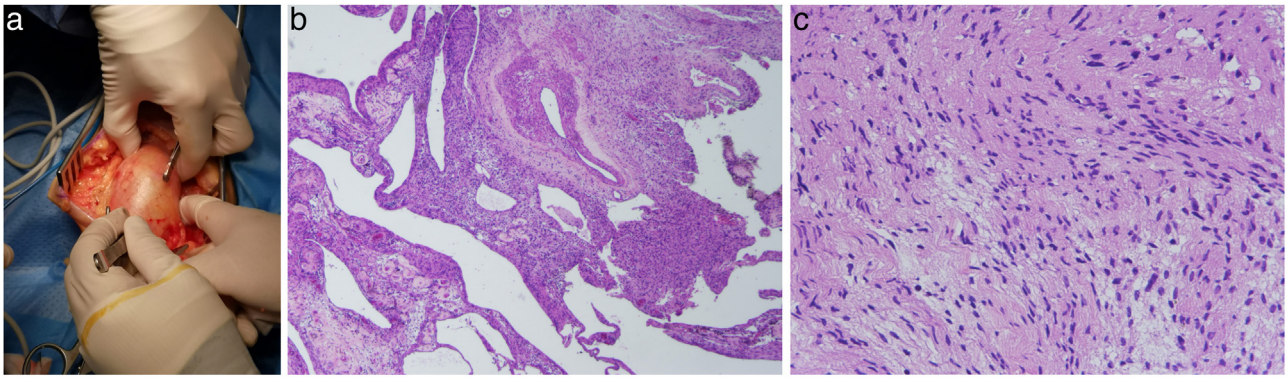

Figure 2 – (a) Intraoperative macroscopic picture of the tumor arising from musculocutaneous nerve. (b) Tumor well delimited by thick fibrous capsule containing cystic spaces of different sizes separated by septa of different thickness. (H-E × 4). (c) In the thicker septa, proliferation of spindle cells is identified without cellular atypia or mitosis of elongated nuclei and poorly defined, eosinophilic, fibrillar cytoplasm. (H-E × 20). (d) IHC study with S-100. Extensive positivity of the cellularity of the tumor that is identified as neural type. The negative areas correspond to connective tissue and abundant blood vessels with thickened and hyalinized walls. (IHQ S-100 × 4).

The tumor and the proximal and distal portions of the affected nerve were exposed under magnification glasses. The epineurial layers were gently peeled away until the shiny surface of the tumor was exposed (Fig. 2a). A longitudinal incision was carefully made in the epineurium far away from the fascicles. Once the tumor capsule was incised, a significant debulking of the cystic part of the mass was performed. The mapping of the tumor capsule and adjacent structures was negative both distally and proximally during resection, allowing careful dissection of the tumor from the MCN. A complete resection of the lesion was carried without damage to the fascicles nor compromising nerve function (Video 1 in Supplementary material). After surgical closure, there were no changes in the amplitude of the tcEMP. According to patient's beliefs and his distinctive interpretations of specific passages from the Bible, no allogeneic blood products were used.

Diagnosis of cystic schwannoma was confirmed by histopathology. Intraoperative findings were consistent with the diagnosis of schwannoma. Grossly, the smooth-surfaced tumor was yellow-whitish (Fig. 2a). Microscopically, the tumor demonstrated a proliferation of spindle-shaped cells arranged in short bundles or interlacing fascicles in Antoni A areas with edematous, hypocellular areas known as Antoni B areas (Fig. 2b and 2c). Neither nuclear atypia nor mitotic figures were observed. The immunohistochemical (IHC) study for the S-100 protein showed extensive positivity for tumor cellularity compatible with the diagnosis of schwannoma. The negative areas for immunohistochemistry corresponded to connective tissue and abundant blood vessels with thickened and hyalinized walls[14] (Fig. 2d).

There was no immediate postoperative neurological deficit following surgery. At eighteen months of follow-up, the patient has no evidence of recurrence and no added neurological impairment.

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Discussion

In different series of peripheral nerve schwannomas of the upper limb,[7,8] these occurred mainly at the level of the elbow or distal to it, and to a lesser extent in its proximal aspect. This may be due to the fact that masses in the distal region of upper extremities are easier to notice than those in the proximal one, and that these tumors are more common in peripheral nerves of greater length and range. The nerves that are most frequently affected in the upper limbs by schwannomas are the median, ulnar and radial, while the MCN is rarely involved.

The exceptionality of this type of tumor in this location, with only three publications as case reports in the literature[4–6] make schwannomas of the MCN a rare but well-described entity. In Guha's paper from 2017,[1] anatomical location on MCN is not reported, and in most extensive series of peripheral neural sheath tumors collected to date by Kim,[2] only 1.7% of cases involving peripheral nerves of the upper extremities arised from MCN.

According to their pathological appearance, schwannomas present a solid pattern,[14] and may have cystic areas of hemorrhagic degeneration and necrosis, although this occurs very rarely. The term ancient schwannoma[9] describes long-standing tumors that are usually relatively large and have undergone degenerative change with areas of cyst formation, calcification, hyalinization, and hemorrhage might be evident on histological examinations. Nuclear atypia has been described to occur in these tumors on the basis of the degenerative process. It is typically not associated with mitotic activity and therefore should not be misinterpreted as an indicator of malignancy.

Total excision of MCN schwannomas was achieved in the three specific case reports in

the literature without recording recurrence after resection, being hypoesthesia and paresthesia in the distribution territory of the MCN the most frequent immediate sequelae after surgery, with practically complete recovery of this impairment after one year, and as it occurs in the majority of schwannomas operated on in the upper extremity.[7,8] Despite the conventional description that most schwannomas can be completely excised without damaging the nerve because the nerve fibers are displaced and do not penetrate the tumor, the study by Chris Tang[15] showed that the vast majority of them had fascicular involvement. This fact points towards a growing interest in the use of IONM, confirming through direct stimulation of neural structures the functionality of the main affected nerve branches and the presence or absence of action potentials in the distal nerve impaired by the lesion, anticipating postsurgical prognosis.[13]

Therefore, this case report describes several strong points for those interested in schwannomas of the upper limb, an unusual presentation impairing the musculocutaneous nerve, infrequent pseudocystic architectural pattern hisyologically confirmed, and the use of IONM to improve the functional outcome after the surgical resection considering the clinical situation of a patient who presented motor sequelae to his childhood poliomyelitis in the right upper extremity. Not to mention the inherent difficulties of approaching surgery where the patient refuses a blood transfusion.

Conclusion

In cases of atypical proximal tumors in the upper limb, MCN schwannoma should be considered as a diagnostic possibility with US and MRI playing a crucial role in its detection. Although infrequent, a pseudocystic pattern can be observed in this type of tumor. Surgical removal of musculocutaneous nerve schwannoma can be achieved under IONM minimizing postoperative complications derived from neural involvement.

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Declaration of competing interest

The authors declare that they have no conflicts of interest.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi: https://doi.org/10.1016/j.neucie.2025.500667.

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