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CASE REPORT

Multiple schwannomas of the upper limb related exclusively to the ulnar nerve in a patient with segmental schwannomatosis

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KEYWORDS

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Summary Schwannomas are benign encapsulated tumours arising from the sheaths of peripheral nerves. They present as slowly enlarging solitary lumps, which may cause neurological defects. Multiple lesions are rare, but occur in patients with neurofibromatosis type 2 or schwannomatosis. Positive outcomes have been reported for surgical excision in solitary schwannomas. However, the role of surgery in patients with multiple lesions is less clear. The risk of complications such as iatrogenic nerve injury and the high likelihood of disease recurrence mean that surgical intervention should be limited to the prevention of progressive neurological deficit.

We report a case of a 45 year old male who presented with multiple enlarging masses in the upper limb and sensory deficit in the distribution of the ulnar nerve. The tumours were found to be related exclusively to the ulnar nerve during surgical exploration and excision, a rare phenomenon. The masses were diagnosed as schwannomas following histopathological analysis, allowing our patient to be diagnosed with the rare entity segmental schwannomatosis. One year post-operatively motor function was normal, but intermittent numbness still occurred. Two further asymptomatic schwannomas developed subsequently and were managed conservatively.

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Introduction

Schwannomas are benign encapsulated tumours arising from the sheaths of peripheral nerves. They present as slowly enlarging solitary lumps, with or without associated neurological defects. Multiple schwannomas are less common, but may occur in the context of neurofibromatosis type 2 (NF2) or schwannomatosis.¹ We report an unusual case of multiple schwannomas confined exclusively to the ulnar nerve, in a patient with segmental schwannomatosis.

Case report

A 45 year old right-hand dominant catering manager presented with two masses in his right forearm, the largest of which measured 2.5 cm in diameter. These had been slowly increasing in size and becoming more painful over a three-year period. He complained of associated sensory symptoms in the distribution of the ulnar nerve, but there was no motor deficit. The patient was normally fit and well, with no significant past medical history. Magnetic resonance imaging (MRI) revealed two further tumours, one just proximal to the cubital tunnel and the other 9 cm distal to the olecranon along the course of the ulnar nerve. The median and radial nerves were normal both clinically and radiologically. In light of the patient's progressive pain and neurological symptoms, the decision was made to perform an elective exploration and excision of the lesions.

Surgical technique

At elective exploration incisions were made directly over the lesions, and four intraneural tumours were identified and excised (Figure 1). All nerve dissection was carried out under the microscope. The lesion was injected with saline to facilitate hydro-dissection. An ophthalmic crescent knife (Beaver Mini-Blade™, Beaver-Visitec International, Inc., Waltham, MA, USA) was used to dissect and tease the tumour from its fascicles, as these tumours usually shell-out. One lesion was intimately involved with the dorsal branch of the ulnar nerve, corresponding to the majority of the patient's sensory deficit. As a very long length of nerve was involved, this branch was sacrificed in order to permit tumour resection. The three remaining lesions were successfully dissected free of the ulnar nerve.

Post-operative course

Histopathological examination of the excised lesions confirmed multiple benign schwannomas of the ulnar nerve. With this information, it was possible to diagnose the patient with segmental schwannomatosis. Post-operatively the patient preserved full motor function but was troubled with sensory symptoms including burning pain in the dorsal branch territory. One year later, the sensory symptoms had reduced to intermittent numbness only. The patient has also developed two further proximal schwannomas in his arm, which are currently being managed conservatively as they are asymptomatic at present.

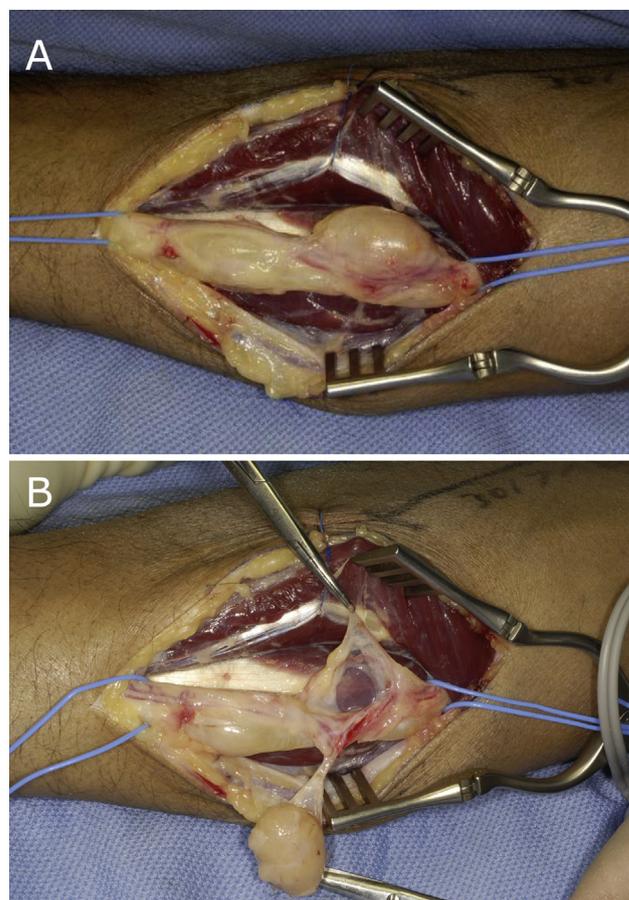


Figure 1 Intra-operative photograph of a schwannoma along the course of the right ulnar nerve with the nerve proximal and distal to the tumour supported by blue slings (A), and a photograph showing the "shelled out" schwannoma (B). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Discussion

Tumours of the peripheral nerves may be either benign or malignant. The benign variants include schwannomas (also called neurilemmomas), neurofibromas and the rarer

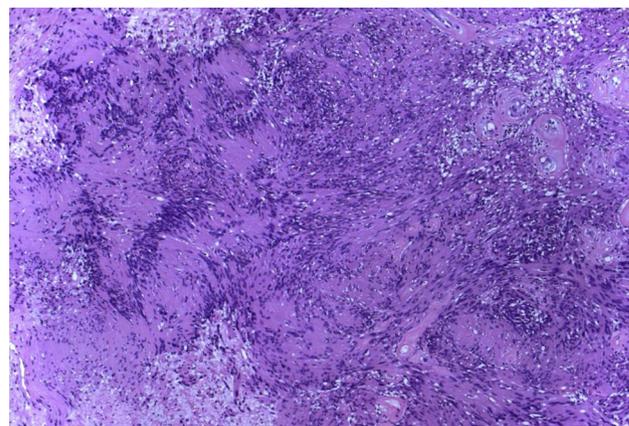


Figure 2 Selected histology of the excised lesions showing the characteristic Antoni A and B patterns seen in schwannomas.

Table 1 Criteria for diagnosis of neurofibromatosis type 2 and schwannomatosis.³**Neurofibromatosis 2**

Any one of the following:

- Bilateral vestibular schwannomas
- First degree relative with NF2 plus unilateral vestibular schwannoma or two of; meningioma, schwannoma, glioma, neurofibroma, posterior lens opacity
- Unilateral vestibular schwannoma plus two of; meningioma, schwannoma, glioma, neurofibroma, posterior lens opacity
- Multiple meningiomas plus unilateral vestibular schwannoma or two of; schwannoma, glioma, neurofibroma, posterior subcapsular lens opacity

Schwannomatosis

Individuals must not fulfil the criteria for NF2 or have any of the following:

- Vestibular schwannoma on MRI, constitutional NF2 mutation, or a first degree relative with NF2

Definite schwannomatosis

- Age >30 years and two or more non-intradermal schwannomas (at least one with histological confirmation)
- One schwannoma confirmed with histology and a first-degree relative who meets the above criteria

Possible schwannomatosis

- Age <30 years and two or more non-intradermal schwannomas (at least one with histological confirmation)
- Age >45 years and no symptoms of eighth cranial nerve dysfunction and two or more non-intradermal schwannomas (at least one with histological confirmation)
- Radiographic evidence of a schwannoma and a first-degree relative who meets the criteria for definite schwannomatosis

Segmental schwannomatosis

- Meets criteria for definite or possible schwannomatosis but limited to one limb or five or fewer contiguous segments of the spine

perineuriomas. Schwannomas are encapsulated spindle cell tumours with cells showing strong immunopositivity for S100 protein.² Characteristic histopathologic features include cytological atypia and the identification of characteristic Antoni A and B patterns, as seen in our case (Figure 2). Longstanding "ancient" schwannomas show cystic degeneration and hyalinisation, and can be more easily confused with malignant peripheral nerve sheath tumours.

Schwannomas typically present as solitary palpable lumps with or without symptoms of nerve compression.¹ As these lesions enlarge they may cause progressive and permanent neurological deficit. Multiple schwannomas occur in the context of NF2, or more rarely without the other hallmarks of NF2, in schwannomatosis. This rare clinical entity has a reported annual incidence of one per 1,700,000, and our patient meets the diagnostic criteria for segmental schwannomatosis (Table 1).³ In addition, cases in which multiple schwannomas are localised to a single peripheral nerve are very rare, and we can find only two reported cases of multiple schwannomas isolated to the ulnar nerve.^{4,5}

The evaluation of schwannomas involves careful clinical assessment of the patient in conjunction with imaging and histopathological analysis. The differential diagnosis for a subcutaneous lump is wide, but the presence of neurological deficits in conjunction with a positive tinell's sign over the lump should alert the clinician to the possibility of a schwannoma. Risk factors such as NF2 should also raise clinical suspicion. MRI is diagnostic and useful in finding occult lesions or new growths in follow-up patients. The diagnostic features of schwannomas on MRI scans include low T1 signal with high T2 signal and uniform enhancement with gadolinium.⁶

In solitary schwannomas surgery has proven to be successful with minimal resulting morbidity.⁷ However, the

management of patients with multiple tumours should be considered an evolving process, with the aim of controlling symptoms and disease progression, as patients inevitably develop further tumours.⁸ Surgical intervention should therefore be limited to the prevention of progressive neurological deficit, for example when sensory deterioration or loss of motor function occurs. Additionally, patients should be counselled regarding the potential for iatrogenic nerve injury during surgery and the high likelihood of disease recurrence. Appropriate rehabilitation regimes should be in place and the possibility of future tendon transfers should be explained.

Conclusion

Schwannomas are tumours of the peripheral nerve sheaths that present as slowly enlarging lumps, with or without neurological deficit. In patients with progressive neurological symptoms and risk factors the threshold for referral to a specialist and further investigation with MRI should be low. The decision to operate on patients with multiple symptomatic lesions is difficult, and the benefits of surgery must be weighed against the potential for iatrogenic nerve injury and likelihood of disease recurrence.

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None.

Conflicts of interest

None.

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