ORIGINAL ARTICLE

Surgical excision for challenging upper limb nerve sheath tumours: a single centre retrospective review of treatment results

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Objective To review the accuracy of different investigation modalities for upper limb nerve sheath tumours and the resulting surgical outcomes, and propose a standard algorithm to deal with such tumours to minimise complications.

Design Retrospective review.

Setting Regional hospital, Hong Kong.

Patients All patients with upper limb nerve sheath tumours being excised in our hospital from 1999 to 2008.

Main outcome measures The accuracy rate of different investigations, as well as corresponding neurological deficits after excision and recurrence rates.

Results A total of 23 (10 male and 13 female) patients, aged between 28 and 72 (mean, 46) years, underwent excision of 25 lesions during the study period. The mean duration of symptom was 2.5 years and tumour size ranged from 1 to 10.5 cm (mean, 2.6 cm). A majority (80%) presented with a typical triad; only one had a true neurological deficit. Twenty-two ultrasonography and 20 magnetic resonance images were obtained, with a diagnostic accuracy of 77% and 100%, respectively. Eight fine-needle aspiration cytology examinations and two core biopsies were performed, which had respective accuracy rates of 13% and 100%. Fifteen patients experienced neurological deficits after the operation; three showed spontaneous recovery. Among 12 patients with long-term residual neurological sequelae, five had both motor and sensory deficits and four had moderate-to-severe disability. No recurrence was reported.

Conclusion Nerve sheath tumours in the hand need to be managed with care. Among the different investigation modalities, magnetic resonance imaging was considered to be the gold standard. Yet ultrasonography is still the most easily accessible and least invasive investigation in public hospital setting. Complications are liable to ensue even if patients are managed by hand specialists. Thus, well-planned operations and detailed discussions with the patient are important prerequisites before operation.

Key words
Nerve sheath neoplasms; Neurilemmoma; Treatment outcome; Upper extremity

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Introduction

Lumps and bumps in the hand and upper extremity are quite common presenting features in our daily practice. One of the most common causes is ganglion. Benign tumours involving peripheral nerves of the upper extremity are uncommon; however, they often pose diagnostic and treatment challenges. Understanding the patho-anatomy of such tumours is an important adjunct to successful treatment.

Nerve sheath tumour (NST), also known as schwannoma, is the most common benign neoplasm of peripheral nerve sheaths. These tumours account for 5% of all soft tissue tumours[1]; their incidence in the upper extremity varies as does their clinical presentation.[2]

The typical clinical presentation triad (mass, positive Tinel's sign, and differential motility) is seldom encountered in daily practice. In most patients the lesion is detected as a painless mass. Preoperative diagnosis is sometimes difficult, and the tumour is easily mistaken for a ganglion when it presents on the volar side near the wrist joint (Fig 1).

具挑戰性的上肢神經鞘瘤切除術：回顧一所中心的治療結果

目的 回顧本中心對於診斷上肢神經鞘瘤的準確度及治療結果，並提出處理上肢神經鞘瘤的標準方法以減低併發。

設計 回顧研究。

安排 香港一所分區醫院。

患者 1999至2008年間所有於本院進行上肢神經鞘瘤切除的病人。

主要結果測量 不同診斷方式的準確度、術後的神經缺損及復發率。

結果 研究期間23位（10男13女）病人進行共25次切除術。病人介乎28至72歲，平均年齡46歲，症狀持續時間平均有2.5年，腫瘤大小平均為2.6 cm（介乎1至10.5 cm）。大部份（80%）病發時有典形的三聯症；只有一位病人出現真正的神經缺損。分別進行了22次超聲波及20次磁共振成像，診斷準確率依次為77%及100%。8次的細針抽吸細胞學檢查和2次的穿刺活檢分別得出13%及100%準確率。術後15人感到有神經缺損；3人自然痊癒。出現長期神經缺損後遺症的12人中，5人有運動和感覺神經缺損，另外5人有中等至嚴重殘疾。沒有復發病例。

結論 要小心處理手部出現神經鞘瘤的病例。在眾多的診斷方式中，磁共振成像可以說是黃金標準治療方法。可是超聲波仍然是公立醫院中最方便和最不具侵略性的方法。即使由手部專科醫生處理這類腫瘤，仍容易出現併發。因此，計劃周詳的手術以及跟病人術前詳細討論病情是相當重要的。

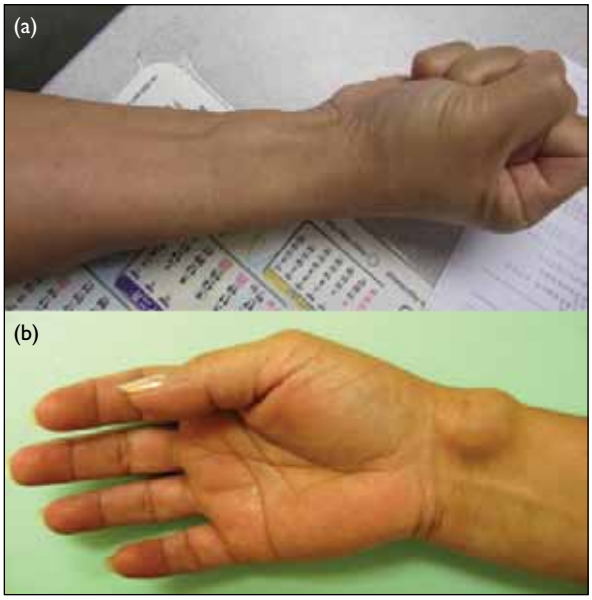

FIG 1. Clinical photos of volar wrist (a) nerve sheath tumour versus (b) ganglion

When the tumour arises from a major nerve trunk, careful planning is important to reduce the risk of neurological damage at operation. The risk is minimised when the diagnosis is made prior to surgery, so that the clinical team can select a surgeon with appropriate microsurgical expertise and equipment to perform the resection.[3]

This paper aimed to evaluate the accuracy of different methods (clinical, radiological, and histological) of making the diagnosis of NST and corresponding surgical outcomes after excision. Based on this experience in our unit, we set out to develop a standard algorithm to deal with upper limb tumours, in order to differentiate NSTs from other benign masses and thus minimise complications.

Methods

All patients who underwent excisional biopsy for upper limb NSTs from 1999 to 2008 at the Prince of Wales Hospital were included in this audit. The medical notes were reviewed retrospectively. We compared the accuracy of clinical diagnosis (typical triad), radiological investigation (ultrasonography [USG] and magnetic resonance imaging [MRI]), and histological assessment (fine-needle aspiration cytology [FNAC] and core biopsy). All excisions were performed directly by or under the supervision of orthopaedic specialists with microsurgical experience. Each procedure was designated as enucleation of the lesion without excision of normal nerve tissue. Complications specific to excision of NST (sensory and motor deficits and recurrent rates) were also recorded.

Results

A total of 23 patients with 25 lesions (one had three NSTs at different sites), aged between 28 and 72 (mean, 46) years, underwent excision during the study period. There were 10 males and 13 females. The mean duration of symptoms (a mass) at presentation was 2.5 years, and its size ranged from 1 to 10.5 cm (mean, 2.6 cm).

Regarding other clinical features upon presentation, 80% of the patients appeared to have the typical triad (mass, Tinel's sign, and differential motility), though only one had a true neurological deficit (sensory loss) [Table 1]. In all, 32% and 40% of the lesions were located in the neck region and along a major nerve trunk, respectively.

In our series of patients, 22 USGs and 20 MRIs were performed. The diagnostic accuracy of USG and MRI were 77% and 100%, respectively. One patient was misdiagnosed as having a lymph node, while in four no conclusive features were noted with USG

(Table 2).

We performed eight FNAC procedures and two core biopsies. In seven patients having the former, the lesions were not conclusively identified by histology (owing to inadequate amounts of tissue). By contrast, core biopsy yielded a correct histological diagnosis in all cases (Table 2). There were no complications in patients undergoing biopsy, except for pain during the procedure.

Concerning the postoperative complications, 15 patients had neurological deficits, three of whom showed spontaneous recovery. Among 12 patients with long-term residual neurological deficits, five had both motor and sensory deficits, and four had moderate-to-severe disability (Table 3). The latter four all had lesions in the neck or arm. No patient presented with a recurrence.

TABLE 1. Comparison of results in terms of clinical presentations

Symptom and sign	Results of present study	Results of Knight et al's study[2]
Tinel's sign	20 (80%)	155 (81%)
Mass (differential motility)	25 (100%)	187 (98%)
Sensory deficit	1 (4%)	9 (5%)
Motor deficit	0	3 (2%)

TABLE 2. Summary of different investigation results*

Results	USG	MRI	FNAC	Core biopsy
Correct diagnosis (accuracy)	17 (77%)	20 (100%)	1 (13%)	2 (100%)
Misdiagnosis	1 (lymph node)	0	0	0
No conclusion	4	0	7 (quantity insufficient)	0
Total	**22**	**20**	**8**	**2**

* USG denotes ultrasonography, MRI magnetic resonance imaging, and FNAC fine-needle aspiration cytology

TABLE 3. Summary of results in each patient

Case No.	Site of lesion*	Neurological deficit	Resolve	Disability
1	Forearm (volar)	-	-	-
2	Neck	Motor and sensation	-	Moderate
3	Neck C6	Motor and sensation	-	No
4	Neck C6	-	-	-
5	Neck C7	Sensation	-	No
6	Deltoid (cutaneous nerve)	Sensation	Yes	No
7	Forearm median	Sensation	-	-
8	Forearm ulnar	Sensation	-	Mild
9	Neck	-	-	-
10	Forearm ulnar	Sensation	Yes	-
11	Deltoid (cutaneous nerve)	-	-	-
12	Axillary NST	-	-	-
13	Elbow median	-	-	-
14	Neck	Motor and sensation	-	Moderate
15	Forearm volar AIN	-	-	-
16	Finger digital	Sensation	-	Mild
17	Neck C7	Motor and sensation	-	Moderate
18	Neck C8	Sensation	-	No
19	Forearm PIN	Motor	-	-
20	Forearm ulnar	-	-	-
21	Forearm median nerve	Sensation	-	Mild
22	Arm ulnar nerve	Motor and sensation	-	Severe
23	Dorsal cut nerve NST	Sensation	Yes	-
24	Axillary NST median	-	-	-
25	Anterior deltoid	-	-	-

* NST denotes nerve sheath tumour, AIN anterior interosseous nerve, and PIN posterior interosseous nerve

Discussion

Nerve sheath tumours often present as slowly growing painless masses while pain and neurological deficits are uncommon unless their size is relatively large in comparison to the nerve canals in which they are situated. This is rare in the upper extremity and Mackinnon and Dellon[1] reported that only 5% of such soft tissue tumours became compromised. The tumours could occur anywhere from the spinal nerve in the neck to the finger tip, but most commonly on the flexor surfaces of upper limbs, as these accommodate the major nerves.

Clinically, deep-seated masses that are present for a long time are located over the course of a nerve. Being transversely but not longitudinally mobile with respect to a nerve suggests schwannoma. In our study however, only 80% of the patients manifested such specific presentations. This was similar to the observation by Knight et al,[2] who reviewed 191 patients presenting with benign solitary schwannomas (Fig 1); their misdiagnosis rate therefore was up to 20%, which was comparable to the review by Rockwell et al.[4] True neurological deficits are rare.

Many diagnostic methods can facilitate an accurate diagnosis before excision. Ultrasonography is a relatively inexpensive and readily available investigation in public hospitals, but its accuracy is not as high as MRI. In our study, the accuracy of USG was about 77%. Höglund et al[3] reported a 59% accuracy rate for making a correct diagnosis of nerve tumours by USG, and they could not differentiate schwannomas from other nerve tumours. Both Fornage[5] in a study using high-resolution USG, and Hughes and Wilson[6] who examined three different neural tumours (neurofibroma, neurofibrosarcoma, and schwannoma) reported that distal sound enhancement is not specific to NST. Although USG is not 100% accurate for the diagnosis of NSTs, it is a

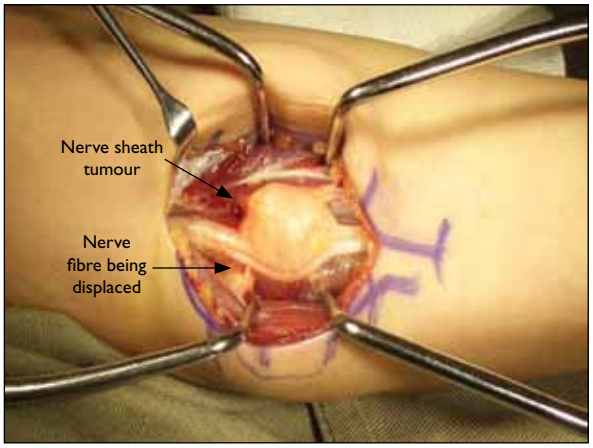

FIG 2. Intra-operative photo showing normal nerve fibre being displaced in the nerve sheath tumour

cheap and quick investigation that can be used as the initial imaging method to study a mass related to a nerve.

Among different diagnostic modalities, MRI is generally considered the gold standard[7]; useful to locate the tumour, define its origin, and reveal neurovascular structures. Such information helps proper preoperative diagnosis to facilitate the safest and most efficacious surgical approach. In T2-weighted MRI images, NST presents as a low central intensity with high peripheral intensity (the so-called 'target sign'). However, the target sign is not specific to NST; it is positive in 50 to 70% of neurofibromas and only 15 to 54% of NSTs.[8-10] In our study, although MRI gave 100% accuracy, there may have been several reasons for this phenomenon. First, our sample size was small (only 20 patients had an MRI). Second, our study was neither randomised nor controlled. Nevertheless, MRI is found to be a more accurate modality for diagnosing NSTs.[2]

Although imaging sometimes gives us a diagnosis, finally we still rely on histopathology and many patients prefer having a preoperative diagnosis before excision. However, FNAC was not particularly useful for that purpose, as in our series most NSTs were not correctly diagnosed, which was also the experience of Kitagawa et al.[11] Whereas, core biopsy can yield a much higher diagnostic rate; it is more invasive, and intolerable pain is always a major issue. Thus, excisional biopsy is only suggested if the diagnosis is in doubt.

Excision of the lesion is the treatment of choice for an NST, as it is well encapsulated and the nerve fibre is displaced, instead of being penetrated. Theoretically, it is possible to remove the lesion without significant nerve deficit (Fig 2). However, even under meticulous surgical dissection and use of microscopic techniques, complications still occur. In our study, almost 50% of patients showed different types of residual neurological deficits, and four showed moderate-to-severe disability. Oberle et al[12] also reported immediate postoperative sensory deficits in six out of 12 patients. Donner et al[13] reported that 13% of 85 patients with NSTs developed muscle weakness after the surgery.

Three reasons were postulated by Sawada et al[14] for nerve dysfunction occurring even when enucleation was possible. First, when an NST arises from the neural sheath, the fascicle surrounded by this sheath is always involved and may still have a function; in which case its resection results in neurological deficit. Second, longitudinal incisions along the sheath of the nerve may divide small numbers of fascicle held taut over the tumour mass. Third, intact fasciculi not involved in the tumour may be compressed or retracted during the operation, resulting in a neuropraxia. The recovery noted in some of our cases was probably due to the latter.

Many researchers are trying to correlate different features of NSTs with surgical outcomes, to anticipate which patient will have higher chances of complications, but this is a controversial area. In our study, review of MRIs revealed no specific feature associated with good surgical outcomes. So detailed informed consent should be obtained from all patients who proceed to surgery, to ensure that they understand the potential neurological risks.

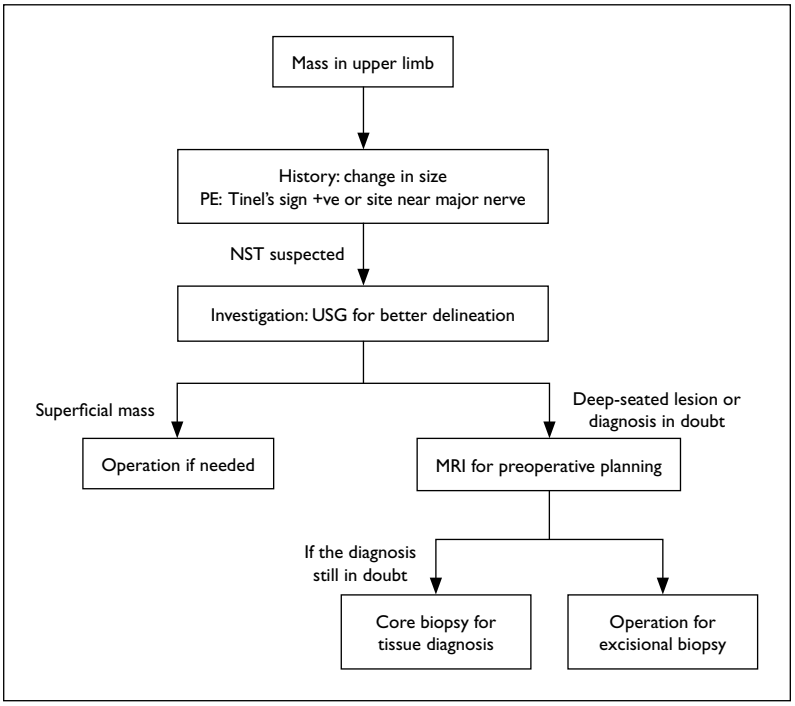

FIG 3. Algorithm when facing mass in upper limb
PE denotes physical examination, NST nerve sheath tumour, USG ultrasonography, and MRI magnetic resonance imaging

Conclusion

Lumps and bumps in the hand should be handled with care. Nerve sheath tumour should be one of the differential diagnoses. The paucity of symptoms and frequent absence of specific signs can pose a diagnostic challenge. Among different investigation modalities, MRI provides more information than USG for a preoperative diagnosis. It can also show exact anatomical relationships with surrounding tissues as a guide to the safest surgical approach. Yet USG is still the most readily available and least invasive investigation in public hospital settings. Complications cannot always be prevented, even if operations are performed by a specialist. Thus, a well-planned operation and detailed discussion with the patient is an important prerequisite to surgery. So as to reduce the complications, in the Prince of Wales Hospital we follow the algorithm shown in Figure 3, whenever we deal with a mass in the upper extremity.

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