Supraclavicular Lump: Think Brachial Plexus Neurogenic Tumour

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Abstract

The diagnosis of a brachial plexus neurogenic tumour should be considered in patients presenting with a supraclavicular lump. Pre-operative diagnosis assists in optimal management. Five cases of neurogenic tumours of the brachial plexus presenting as a supraclavicular lump are reviewed. Excision or enucleation was performed. Only one case developed a permanent neurological deficit. Differences between neurofibroma and schwannoma are discussed.

INTRODUCTION

Neurogenic tumours of the brachial plexus are an important differential diagnosis for a supraclavicular lump. The majority present as a slow growing mass. The diagnosis may be suggested by pain or paraesthesia in the arm or shoulder particularly on palpation (Tinel's sign). A key element in management is a correct pre-operative diagnosis.

Our experience of five such tumours are reported with special emphasis on the micro-surgical technique and post-operative neurological status. Differences between neurofibromas and schwannomas are highlighted.

REVIEW OF CASES

We reviewed the cases of five brachial plexus neurogenic tumours presenting to one surgeon (RKM) in our hospital between 1987 and 1997. The details of the five cases are summarised in table 1. There were three females and two males whose ages ranged from 45 to 78 years old, with an average age of presentation at 62 years old. All five patients presented with a slow growing supraclavicular mass. Only one case complained of neurological symptoms pre – operatively. In this patient, Tinel's sign was positive to the infraclavicular and anterior deltoid region. The correct diagnosis of a neurogenic tumour was suspected in the latter three cases pre-operatively and confirmed on MRI imaging. All lesions were found to be arising from the brachial plexus at operation.

Figure 1

Table 1: Summary of five cases of brachial plexus neuroma

Age	Sex	Site of tumour	Surgical technique	Size	Diagnosis	Post-operative neurological deficit
78	Male	Supraclavicular	Excision	3x3x2cm	Neurofibroma	None
62	Female	Supraclavicular	Excision	4x4x4cm	Schwannoma	Transient hypoaesthesia of index finger
58	Male	Supraclavicular	Excision	2x2x2cm	Neurofibroma	Permanent hypoaesthesia ulnar aspect forearm
69	Female	Supraclavicular	Enucleation	3x2x2cm	Schwannoma	Transient hypoaesthesia of index finger and thumb
45	Female	Supraclavicular	Enucleation	1x1x1cm	Schwannoma	None

Total excision was carried out except for two cases where enucleation was performed on schwannomas in an effort to preserve the function of the nerve. Post-operative neurological status showed a permanent deficit in one case and temporary deficit in two cases and no deficit in two cases. All deficits noted were sensory. Histology of the lesions showed two neurofibromas and three schwannomas confirmed on immunostaining. There have been no recurrences to date.

Figure 2

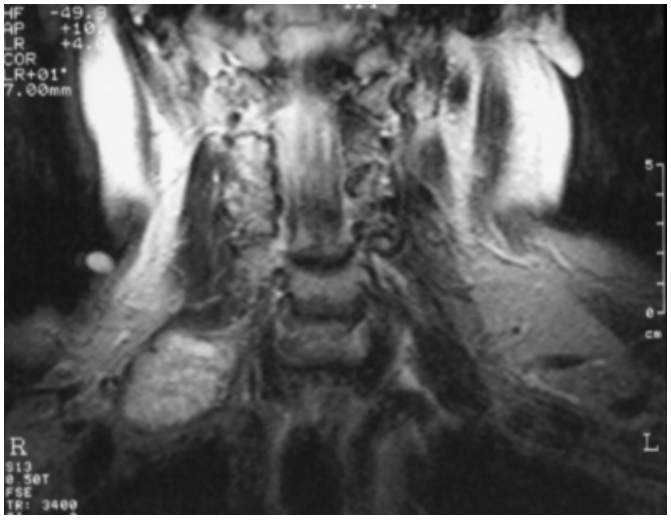
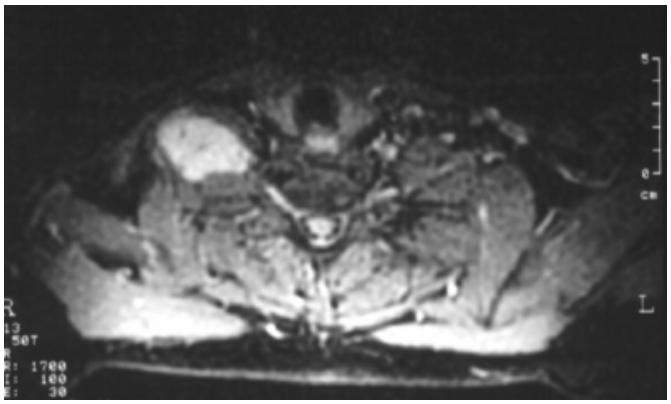


Figure 3



DISCUSSION

Neurogenic tumours of the head and neck are uncommon tumours that can arise from cranial, peripheral or autonomic nerves. The vast majority are benign and include neurofibromas and schwannomas. The malignant group comprises neurogenic sarcomas, malignant schwannomas, neuroeptheliomas and malignant melanomas.[1]

Most tumours originating from the glossopharyngeal, vagus, accessory and hypoglossal nerves and sympathetic chain are located in the medial aspect of the neck. Laterally, they arise from the cutaneous or muscular branches of the cervical plexus or from the brachial plexus.[2]

The commonest presentation is a slow growing mass. Upon palpation, they are slightly mobile except along the long axis of the nerve. Neurological symptoms are not usually seen.[3] The diagnosis relies on clinical suspicion.

PRE-OPERATIVE DIAGNOSIS

Pre-operative diagnosis is extremely important for two reasons. Firstly, if the nerve of origin can be determined by imaging, the patient can be warned about possible neurological sequelae post-excision.[4] Secondly, knowledge of the surgical techniques for excision of neurogenic tumours will give the best neurological outcomes. Confident pre-operative diagnosis may also avoid unnecessary panendoscopy and biopsy.

Fine needle aspirate may give a diagnosis in a quarter of cases. The predominant feature is the presence of spindle cells.[4] However it is new imaging techniques that have revolutionized the diagnosis of these tumours. CT has been superseded by MRI because of its better differentiation of soft tissues. T1-weighted images show the tumour to be of intermediate signal and T2-weighted images show a high signal with some heterogeneity. [5] These appearances are not specific to peripheral nerve tumours, although the diagnosis may be suggested if the lesion arises from a major nerve trunk.

MRI may also assist in pre-operative differentiation between schwannoma and neurofibroma. The nerve is shown to lie peripheral to the tumour in schwannomas while it is central or obliterated in neurofibromas. In addition, schwannomas often show cystic change where neurofibromas do not.[6]

HISTOLOGY

Neurilemomas are classically described as " well-circumscribed, encapsulated masses that are attached to the nerve but can be separated from it". Nerve filaments may be splayed over the surface of the tumour. Microscopically, the tumour shows cellular areas(AntoniA), including Verocay bodies, as well as looser, myxoid regions (Antoni B). Silver stains demonstrate that axons are excluded from the tumour, although they may become entrapped in the capsule.[7]

In contrast, neurofibromas are well – circumscribed but not encapsulated masses. It is not possible to separate the lesion from the nerve. Microscopically they show a haphazard arrangement of fibroblasts, Schwann cells and macrophages. Axons can be demonstrated within the tumour. Electronmicroscopy and immunohistochemical analysis (S-100, Leu-7) are often necessary to diagnose and accurately classify neurogenic tumours.[8]

However, the distinction between schwannomas and neurofibromas is not always clear cut as one recent report highlighted tumours with both components present within

the same specimen.[9]

SURGICAL TREATMENT

As the neurofibroma involves the main axon, excision always sacrifices the nerve. Depending upon the importance of the nerve it can be repaired by primary anastomosis or a nerve graft. The surgical management of schwannomas is less clear cut. As schwannomas arise from the side of the nerve, cautious surgical dissection, with extracapsular "peeling", or even enucleating, the tumour from the nerve of origin has been described in an effort to preserve function of the nerve.[10,11] The operating microscope should be employed to assist with dissection of nerve fascicles from the tumour.

However, in one study of six cervical schwannomas, only one was an eccentric mass pushing the undisturbed nerve aside. [12] In the five other cases, excision of the neurilemmoma required complete nerve excision. Neural elements travelling through the central portions of the tumour were clearly demonstrated histologically.

Hence, some authors feel that enucleation or partial excision is inadvisable for oncological reasons despite the appeal of functional preservation. [12,13] One literature review of 146 cervical schwannomas demonstrated a 4% incidence of malignant schwannomas.[12]

OUTCOME

Our neurologic outcomes for surgical treatment of brachial plexus neurogenic tumours are permanent deficit one out of five (20 per cent), transient deficit two out of five (40 per cent) and no deficit two out of five (40 per cent). These compare favourably with one review of the literature for surgical treatment of cervical schwannomas in which permanent deficits were reported to be two out of seven (29 per cent), transient deficits three out of seven (43 per cent) and no deficit two out of seven (29 per cent). [11] The number of cases in this study are too small to make any conclusions of differences in outcome between schwannomas and neurofibromas.

CONCLUSIONS

When confronted with a patient with a supraclavicular mass the differential diagnosis includes a brachial plexus neurogenic tumour. Neurological symptoms, clinical examination and FNA cytology may assist in the diagnosis. However, MRI imaging is crucial to diagnosis and management.

Knowledge of the nerve trunk involved assists in pre-operative counselling. If a diagnosis of schwannoma is suggested by imaging or the macroscopic appearances of the tumour enucleation may be attempted to preserve neural function.The malignant potential of these tumours is small and therefore conservative management is an option for selected patients.

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