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ORIGINAL ARTICLE

Approach to Brachial Plexus Tumors - Our Experience

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Abstract

Aims of this study: Our aim is to define a rationale approach in dealing with brachial plexus tumors, the investigations required and the best surgical approach to excise the tumour with least morbidity and to provide early functional recovery.

Methods and materials: We retrospectively reviewed the medical records of the nine patients with primary brachial plexus tumour treated in our institution between 2008 and 2015. Mean age was 34 (range 13 - 44) years at the time of presentation. All patients had preoperative MRI scans .

Results: All patients were evaluated clinically. Radiographs of the chest and MRI were taken to identify the lesion and the nerves involved. All the patients presented with pain as the principal symptom followed by swelling. All but one patient had single stage resection of the tumor. Three Patients had clavicular osteotomy. Microscopic resection was possible in eight out of the nine patients.

Conclusions: Benign lesions in which excision of tumour had done, needs no further intervention except for conditions in which nerve fascicles had been removed need secondary nerve transfers, nerve grafting or tendon transfers. Malignant tumours need subsequent radiotherapy with or without chemo therapy & once they were clinically recovered may need secondary procedures for their functional impairment.

Keywords: Brachial plexus tumour, Schwannoma, Neurofibroma

Introduction

Tumors involving the brachial plexus are rare and constitute about 5% of primary upper limb tumors. These tumors could be of nerve sheath origin (schwannoma), non-nerve sheath origin (neurofibroma). Benign schwannomas are the commonest peripheral nerve tumors and malignant transformation is extremely rare. Solitary neurofibromas are seen in about 10% of patients with neurofibromatosis.

Most cases of brachial plexus tumors reported in literature have been managed by neurosurgeons with all large series reported from neurosurgical specialty centres.[1,2,3] Schwannomas are derived from the myelinating cell of the peripheral nervous system and are composed almost entirely of Schwann cells. Schwannomas typically grow within a capsule that remains peripherally attached to the parent nerve. The tumor subsequently grows, eccentrically compressing the normal adjacent axons. Antoni A and B tissue types represent distinct histologic architectural patterns that aid in the pathologic diagnosis of schwannomas and may influence their imaging characteristics.[4] Type A tissue is highly cellular and demonstrates nuclear palisading and associated Verocay bodies, which may reflect their prominent extracellular matrix and secretion of laminin. The adhesive properties of laminin are thought to explain the tight organization within Antoni A tissue. Type B tissue is loosely organized with myxomatous and cystic changes and may represent degenerated Antoni A tissue (Fig 1). By contrast, neurofibromas appear to contain all the cellular elements of a peripheral nerve, including Schwann cells, fibroblasts, perineurial cells, and axons. The tumor cells grow diffusely within and along nerves, causing the nerves to expand radially while entrapping native neural elements within the substance of the tumor This intraneural growth pattern, with its entrapped axons, provides a key feature to histologically distinguish neurofibroma from schwannoma. Pain, swelling and loss of function are the reasons why patients seek treatment and the diagnosis though can be made clinically, if facilitated by the widespread use of MRI.[5]

We present nine cases of tumors involving the brachial plexus, all of which were surgically treated,to define a rationale approach in dealing with brachial plexus tumors, the investigations that are needed and the best surgical approach to excise the tumor with least morbidity and to provide early functional recovery.

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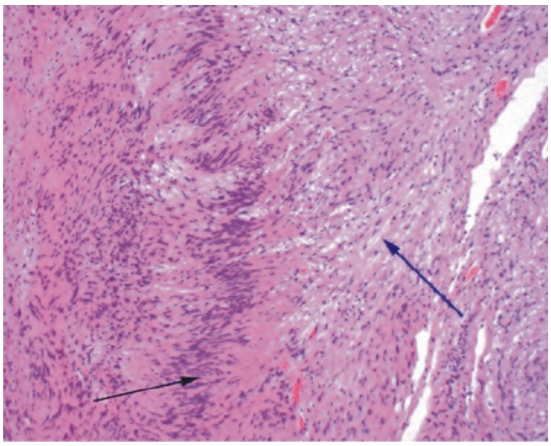


Fig. 1 Photomicrograph of tissue within a schwannoma

Patients and Methods

We retrospectively reviewed the medical records of the nine patients with primary brachial plexus tumor treated in our institution between 2008 and 2015.

These patients included six females and three males. The mean age was 34 (range 13-44) years at the time of presentation. All patients had preoperative MRI scans which localized the tumors to the brachial plexus

Results

All the patients presented with pain as the principal symptom followed by swelling. One patient had ulnar clawing of the hand on the affected side. Only one patient had cutaneous stigmata of neurofibromatosis. The duration of symptoms ranged between 20 days to 1 year. All but one patient has had single stage resection of the tumor. The supraclavicular approach was used in eight of the nine patients except for one patient who had an additional posterior neck incision to access the intrathecal extension of the tumor. This was done as a staged procedure one week apart. Three Patients had clavicular osteotomy to better access the tumor. The clavicle was preplated and osteotomized then replated after the tumor was microsurgically dissected and excised. Intraoperative nerve stimulation was used in all patients. One patient had primary sural nerve grafting after the entire C7-T1 roots were excised with the tumor. One patient had developed shoulder weakness following surgical excision of neurofibroma for which she had underwent nerve transfer using motor branch of long head of triceps to axillary nerve.

Gross and microscopic resection was possible in eight out of the nine patients. All surviving patients over the follow up period, have complete resolution of symptoms with no residual motor or sensory deficits.

Schwannoma was histologically confirmed in six patients; neurofibroma in two patients and neurofibrosarcoma in the remaining one patient who succumbed after chemoradiation therapy. Table:1

Case 1

A 32 year old female presented with neck pain with radiation to left upper limb for four months duration, increased in intensity for 1 month duration. Neck pain was an intermittent dull ache aggravated by neck movements. She also had numbness and paresthesia in left upper limb. There was no history of trauma, fever, systemic illness or bladder and bowel disturbances. She had fullness in the posterior triangle of neck with no spinal tenderness. Her range of neck movements were good, but associated with pain. Sensory evaluation showed decreased sensation over left C6,C7,C8 dermatome. MRI neck showed a neurofibroma in left supra clavicular region of left brachial plexus with intra spinal extension through left neural foramina at C5-C6, C6-C7 and C7-T1 causing significant cord compression and edema at C6-C7 level (Fig 2,3).The surgery was performed in two stages; stage I - excision of intradural extramedullary tumour of C6,C7 level, and stage II - excision of Brachial plexus tumor.

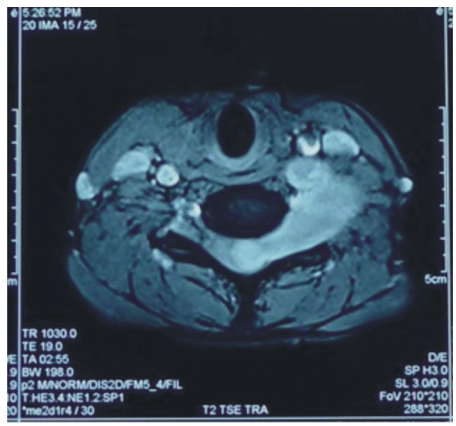


Fig. 2 MRI shows intra thecal extension of brachial plexus tumour

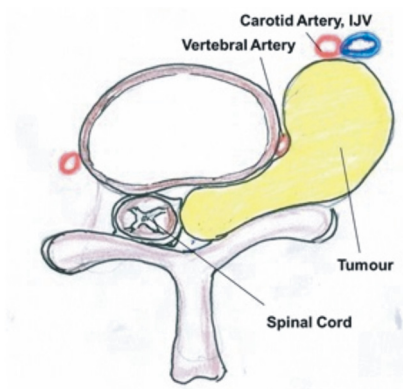


Fig. 3 shows schematic representation

Table:1

S no	Age	Sex	Clinical features	Duration	Side	Location of tumour	Size of tumour	Excision	Histology	Comments
1	21	M	1.swelling 2. radiating pain 3. ulnar clawing	20 days	R	Lower trunk	3x2x2cm	Total	Schwannoma	Supraclavicular approach 30/09/2008
2	18	F	1.Weakness 2.pain	5 months	R	C7,C8,T1 ROOTS	5x4x3cm	? total	Neurofibro sarcoma	Sural nerve grafting Clavicle osteotomy (EXPIRED) 11/05/2010
3	44	M	1.pain 2.paresthesia	8 months	R	C7 ROOT	2cmx3cm	Total	Schwannoma	Supraclavicular approach 01/08/2012
4	35	F	1.pain 2.recurrent neck swelling	12 months	L	Lower trunk	6x4x3cm	Total	Schwannoma	Clavicle osteotomy
5	32	F	1.pain 2.swelling	1 months	L	C4 ROOT	3.4X3X2.9cm	Total	Schwannoma	Supraclavicular approach 02/02/2010
6	24	F	1.pain 2.swelling 3.paresthesia 4.weakness	3 months	R	C7 ROOT	4.2x2.3x2.2cm	Total	Schwannoma	04/05/10 : Exploration and excision of entire tumor along with c7 root and its division and repair of lateral cord. Clavicle osteotomy 04/05/2010
7	30	F	1.pain 2.swelling	4 months	L	Intrathecal C5-T1 mainly C7	4x3cm	Total	Neurofibroma	Two stage excision 16.04.2013 : posterior and postero-lateral excision of C6-C7 intra-dural neurofibroma (left side). 23.04.2013 : Supraclavicular approach
8	44	M	1.pain	5 months	L	ulnar nerve in axilla	5x3cm	total	Schwannoma	Infraclavicular approach
9	37	F	Pain , numbness in right shoulder	3 months	R	Posterior division of the upper trunk	8.5x4.5x3.5cm	total	Neurofibroma	02.04.2015 : Supraclavicular approach 24.11.2015- Nerve transfer from motor branch of long head of triceps to axillary nerve.

Stage I: Under general anaesthesia, patient was positioned prone over bolster with Mayfield skull traction. Using posterior midline 'T' shaped incision, posterior elements of C5 to C7 exposed laminectomy of C6 and C7 done. C6-C7 facetectomy and excision of C6 lateral mass done. Durotomy done at C6-C7 level. Neurofibroma mass was separated from the cord and nerve root ;and excised in piecemeal. C7 nerve root on left side was sacrificed. Debulking of the tumor mass done in sequential manner anteriorly up to apex of left lung.

Stage II: (1 week later) - Skin incision made over the left supra clavicular fossa. Incision deepened to the platysma. omohyoid muscles and external jugular veins were divided. C5, C6 nerve roots were found to be free. Tumor measuring 4x3 cms was found deep to C5, C6 nerve roots. Previously excised segment margins found at the proximal aspect of C7 nerve root. Tumor removed from C7 root. (Fig 4,5).

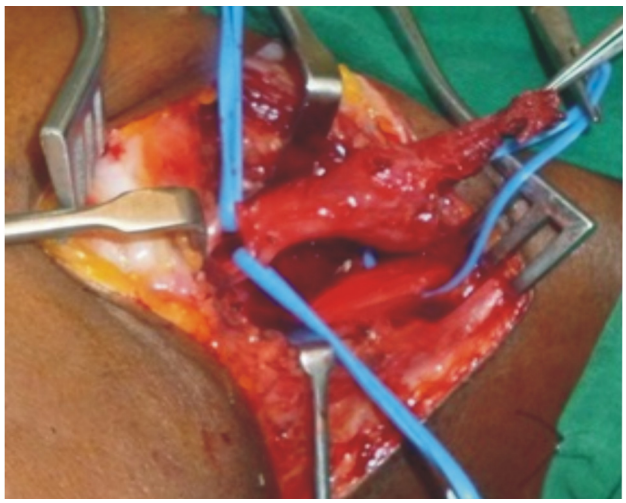


Fig. 4 Tumour mass from C7 root

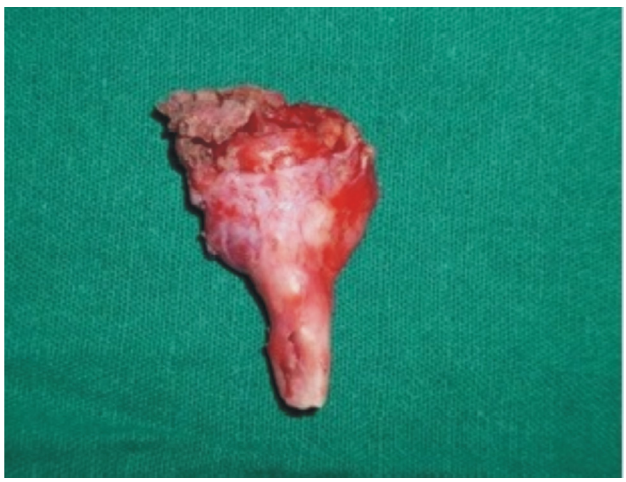


Fig. 5 Tumour mass removed

Histopathology of the tumor showed a benign lesion consistent with neurofibroma composed of proliferating slender spindle cells arranged in sheets with slender and wavy nuclei. No pleomorphism or increased mitosis was seen. Collagen bundles of varying thickness were noted.

Case 2

A 35 year old doctor had presented with the complaints of pain and swelling over the left side of the base of neck. It was a recurrent lesion at the site of previous operation at her native place, now associated with mild pain in the neck and deltoid region (Fig 6). She had no weakness or numbness in the left upper limb. On examination there was a 5x5 cms firm swelling in the left clavicular region with extension behind the left clavicle and firm in consistency. Except for the paresthesia in the deltoid region she had no neurological deficit. MRI showed schwannoma at of the left brachial plexus behind the mid third of clavicle.

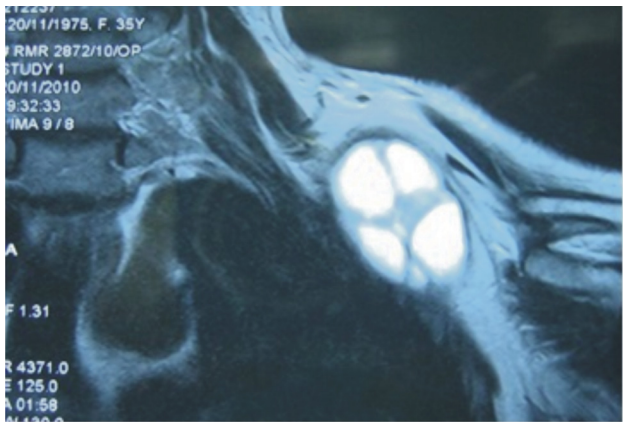


Fig. 6 MRI showing well circumscribed mass, with lobulation

Under general anaesthesia,in supine position, a transverse incision was made above and parallel to the clavicle and extended in a 'T' shaped fashion to the infraclavicular region (Fig-7). Clavicle osteotomy was required for adequate exposure of the tumour. Preplating of clavicle was done and it was osteotimized. The tumour was found to be superior and anterior to the brachial plexus pushing the plexus posterio inferiorly and anterior to the spinal accessory nerve. With careful dissection, the tumour was dissected from the spinal accessory nerve and brachial plexus and it was completely excised enmass (Fig-8,9). Clavicle osteotomy site was fixed with 3.5 mm dynamic compression plate (Fig-10) and the wound closed over a drain. Histopathology of the tumour show a benign tumour consistent with cystic schwannoma.

Implant removal from the left clavicle was done 27 months later and she has no neurological deficit.

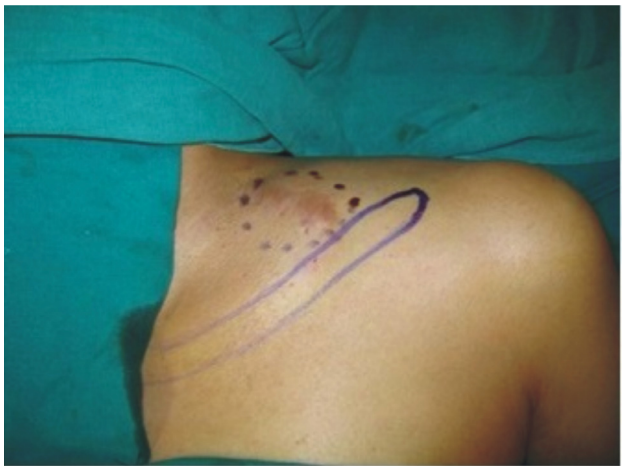


Fig. 7 Surface marking of left clavicle showing the mass in supraclavicular region with previous surgical scar.

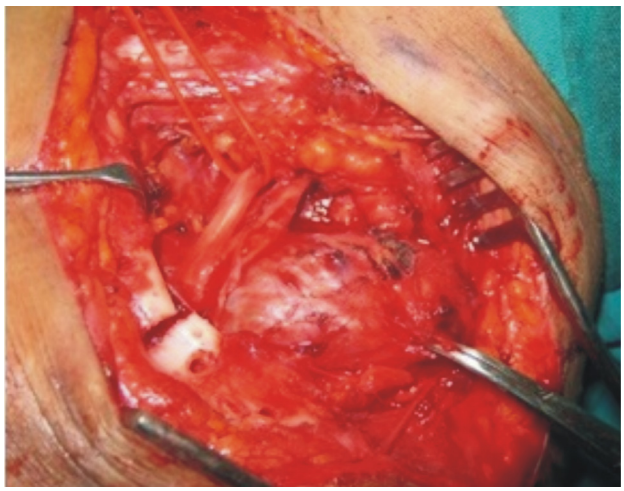


Fig. 8 Intra operative picture shows clavicular obsteotomy with tumour mass

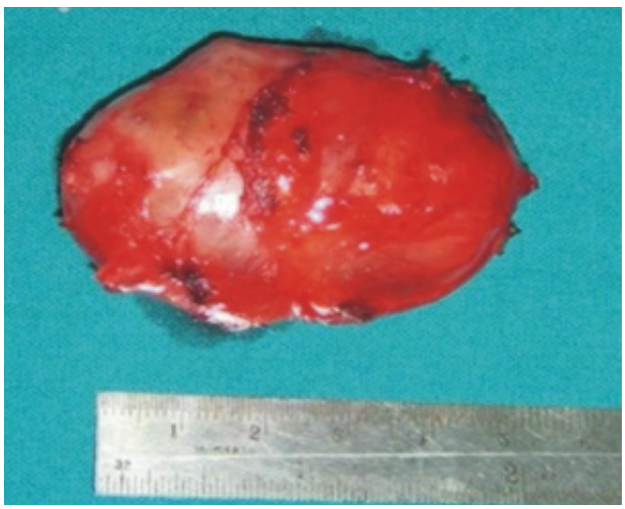


Fig. 9 Tumour mass removed

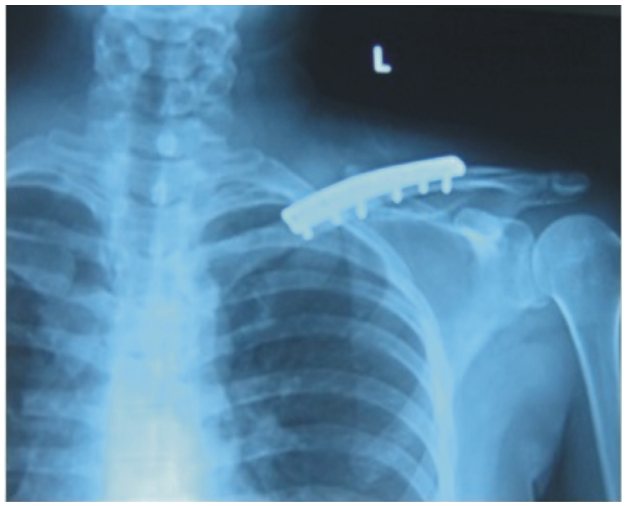


Fig. 10 Post operative radiographs showing clavicular plate fixation following osteotomy.

Discussion

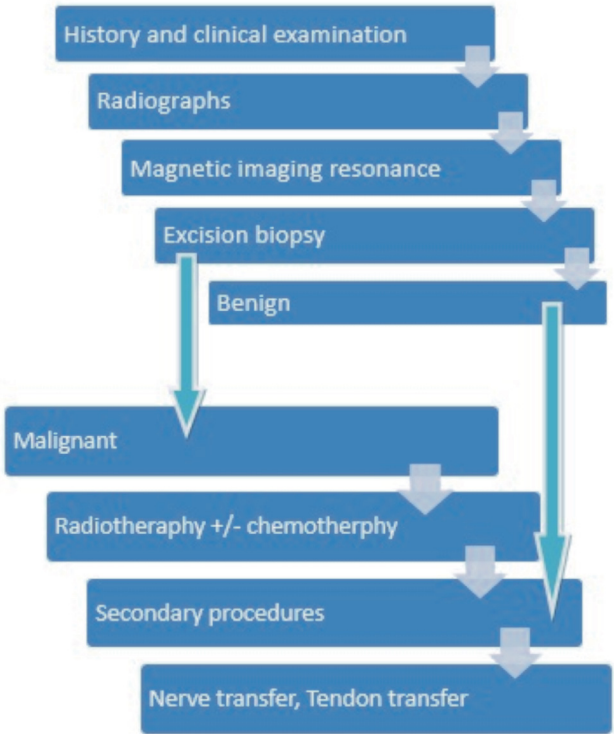
Patients with tumours arising from, or involving the brachial plexus commonly present with pain and a mass on the affected side with varying degrees of paresthesia and uncommonly with other neurologic symptoms. In patients with soft tissue masses, MRI may contribute to recognition of nerve sheath tumors by showing the nerve of origin and typical signal hyperintensity on T2-weighted images[5].In schwannomas the nerve was is usually situated at the periphery of the lesion. In neurofibromas the nerve was is either visible in a central position within the mass or no longer visible. It can also be helpful in distinguishing between schwannomas and neurofibromas by the location of the nerve of origin and the presence of a capsule.

In patients presenting with schwannomas, it is possible to achieve complete surgical excision with no residual deficits. Schwannomas typically have a well delineated capsule which makes resection relatively easier and total excision possible in almost all cases whereas in neurofibromas, the fascicles are intimately involved in the tumour and some nerve fascicles must necessarily be sacrificed.

Numerous surgical approaches have been described in literature including the trans clavicular approach which was used in three patients in our series. This approach is infrequently used but it has the advantage of wide exposure. Zadnik et al[6] have reported the use of the trans clavicular approach in twenty patients for exploration of the brachial plexus after acute injury. In their series, no complication relating to the hardware

was reported and we have had similar experience in outpatients who have tolerated the implant. Malignancy involving the brachial plexus poses a challenge, and we managed our index case with primary nerve grafting of the brachial plexus with the view that radiation to the affected side will make future reconstruction difficult. Satisfactory outcomes have been reported in all case series of brachial plexus tumours. Donner et al reported improved or unchanged motor function in 87% of patients with schwannoma and 85% had resolution of pain.Ganju et al[3] in a large series, reported improved or unchanged pain in 78% of patients with schwannomas and 70 % with improved or unchanged motor function.

Treatment algorithm



All patients were initially evaluated clinically to localize the lesion with detailed neurological examination. Radiographs of the chest with cervical spine were done to rule out osseous erosions in the vertebral bodies and or any apical pulmonary tumour of the lung. MRI helps in will help in identifying the site, size of the lesion along with the nerves involved and in identifying schwannoma and neurofibroma as also malignant lesions. Fine needle aspiration will not provide adequate tissue samples, and trucut biopsy can cause iatrogenic damage to brachial plexus hence was not done in any of our patients. Supra clavicular approach was used in eight cases and decision to do clavicular osteotomy was decided based on the extent of lesion. Tumours with infraclavicular extension needed clavicular osteotomy for better exposure and surgical excision. Following excision biopsy of the lesion further management depends on the histo pathological identification of the specimen. Tumour involving C7 root or the middle trunk can be excised without any significant neurological deficit. Benign lesions in which excision of tumour has been done need no further intervention except for conditions in which nerve fascicles had been removed with need of secondary nerve transfers, nerve grafting or tendon transfers. Malignant tumours need subsequent radiotheraphy with or without chemo therapy. Patients with recovery may need secondary procedures for their functional impairment.

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