

Brachial plexus tumors in a tertiary referral center: a case series and literature review

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ABSTRACT

Introduction: Brachial plexus (BP) tumors are very rare tumors, with less than 800 cases been described in the literature worldwide since 1970. These tumors often present as local or radicular pain, with scant or no neurological deficits. These symptoms are shared by many other more common rheumatologic diseases, thus making their diagnosis difficult in most cases. Additionally, these tumors often present as lumps and are therefore biopsied, which carries a significant risk of iatrogenic nerve injury.

Material and Methods: In this paper the authors describe their experience in the management of 5 patients with BP tumors, followed-up for at least 2 years. There were 4 males and 1 female. The median follow-up time was 41 ± 21 months. The average age at diagnosis was $40,0 \pm 19,9$ years. The most common complaints at presentation were pain and sensibility changes. All patients had a positive Tinel sign when the lesion was percussed. In all patients surgery was undertaken and the tumors removed. In 4 patients nerve integrity was maintained. In one patient with excruciating pain a segment of the nerve had to be excised and the nerve defect was bridged with sural nerve grafts.

Results: Pathology examination of the resected specimens revealed a Schwannoma in 4 cases and a neurofibroma in the patient submitted to segmental nerve resection. Two years postoperatively, no recurrences were observed. All patients revealed clinical improvement. The patient submitted to nerve resection had improvement in pain, but presented diminished strength and sensibility in the involved nerve territory.

Conclusion: Surgical excision of BP tumors is not a risk free procedure. Most authors suggest surgery if the lesion is symptomatic or progressing in size. If the tumor is stationary and not associated with neurological dysfunction a conservative approach should be taken.

Keywords: Brachial plexus; Schwannoma; Neurofibroma; Peripheral nerve sheath tumor; Upper Limb; Surgery.

INTRODUCTION

The first description of a brachial plexus (BP) tumor is attributed to Courvoisier who in 1886 published a paper on a patient with a tumor from the C5 nerve root^{1,2}. Unfortunately, his attempt to remove the tumor was marred by subsequent paralysis of the deltoid and biceps muscles^{1,2}. Since then several reports have been published on this subject, allowing a better prognosis than that described by Courvoisier in most cases^{2,4}. However, most of these reports are composed of case reports or small series of patients⁴. The largest series published is a 30 year retrospective study, in a large tertiary referral center in the United States of America, that identified 226 patients². The second largest study is a recent 10 year review of 115 BP tumors operated by a single surgeon in India³.

Upper limb peripheral nerve sheath tumors represent only 1 to 4,9% of all upper-extremity tumors⁵. BP tumors are even rarer⁵. Moreover, they often present as local or radicular pain, with scant or no neurological deficits^{2,4}. These symptoms are shared by many other more common rheumatologic diseases, thus making the diagnosis of BP tumors difficult in most cases^{2,4}. Additionally, these tumors often present as lumps and are therefore biopsied, which poses a significant risk of iatrogenic nerve injury^{2,4}.

In this paper the authors describe their experience

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TABLE I. SUMMARY OF THE PATIENTS WITH BRACHIAL PLEXUS TUMORS

Patient	Gender	Age at presentation (years)	Symptoms at presentation	Location	Surgical Procedure	Histology	Outcome 2 years after surgery
1	M	24	Hypoesthesia, intense pain, and diminished strength in the ulnar nerve territory	Origin of the ulnar nerve	Excision of the involved nerve segment, and sural nerve grafts to bridge the nerve defect	Neurofibroma	Partial recovery of strength and sensibility to the ulnar territory; no pain
2	F	42	Lump and pain in the supraclavicular region	Origin of the suprascapular nerve	Excision of the tumor preserving the nerve	Schwannoma	Normal function; no symptoms
3	M	51	Lump and pain in the supraclavicular region; hypoesthesia and paresthesia in the posterior aspect of the forearm	Posterior cord of the brachial plexus	Excision of the tumor preserving the nerve	Schwannoma	Normal function; residual hypoesthesia.
4	M	66	Pain and diminished strength in the territory of the radial nerve	Origin of the radial nerve	Excision of the tumor preserving the nerve	Schwannoma	Normal function; no symptoms
5	M	17	Pain and dysesthesia in the median nerve territory; lump in the arm	Median nerve	Excision of the tumor preserving the nerve	Schwannoma	Normal function; no symptoms

M: Male; F: Female.

with the management of 5 patients with BP tumors followed up for at least 2 years.

METHODS

The authors retrospectively reviewed the charts, and clinical images of 5 patients with brachial plexus tumors, referred to the Brachial Plexus and Peripheral Nerve Surgery Outpatient Clinic, at São José Hospital (Lisbon, Portugal). The basic demographic features, clinical picture and outcome, two years postoperatively of these 5 patients are described in Table I.

All patients but one were male (4M:1F). Median follow-up time was 41 ± 21 months. Average age at presentation was $40,0 \pm 19,9$ years, ranging from 17 to 66 years. All patients complained of pain in the territory of the involved nerve. This symptom was particularly

disabling in patient number 1, who was later found to have a neurofibroma. Two patients presented with diminished strength in the territory of the involved nerve. Two patients presented a palpable lump corresponding to the location of the tumor. In these two cases the mass had a greater side-to-side mobility than longitudinal mobility. Two patients complained of significant hypoesthesia. One of these patients (Patient 3) said that this symptom had occurred after a biopsy of the region in another institution prior to referral to the Brachial Plexus Clinic. One patient with a median nerve tumor referred crippling dysesthesia in the cutaneous territory of this nerve in the palm of the hand (Patient 5). All patients had a positive Tinel sign when the lesion was percussed.

All patients had imagiological evidence of the tumor either in computed tomography (CT) scan or magnetic resonance imaging (MRI). All patients with sensi-

bility changes had electroneuromyographic abnormalities. There was no history or evidence of neurofibromatosis in any of the patients.

The surgical procedure consisted of exposure of the involved nerve and surrounding structures. The tumor was then isolated using magnifying loupes. In 3 cases the tumor was partially encased in a fibrous sheath that facilitated intraneural dissection and removal of the tumor without significantly damaging the nerve fascicles (Figure 1). In one case, concerning a suprascapular nerve tumor, the lesion was highly adherent to the nerve fascicles and no fibrous sheath was found (Figure 2). However, after a laborious intraneural dissection, it was possible to isolate the tumor from the main nerve fascicles, maintaining nerve integrity. Finally, in one case of an ulnar nerve tumor, no dissection plane was found, as the tumor permeated all the thickness of the nerve (Patient 1). In this patient, it proved necessary to excise the involved segment of the nerve and bridge the defect with four cables of sural nerve graft (Figures 3 and 4).

RESULTS

No intra-operative or postoperative complications were noted, being the patients discharged home a few days after the surgery (3 to 5 days). All surgical wounds healed uneventfully. Patients were followed up for a minimum of two years after surgery. The pathology examination of the resected specimens revealed a schwannoma in 4 cases and a neurofibroma in one patient (Patient 1).

Two years postoperatively, no recurrences were observed. Additionally, all patients revealed clinical improvement. Three patients were symptom free. One patient, who had been subjected to nerve biopsy prior to referral, had normal motor function, but complained of residual paresthesia and hyposthesia in the dorsum of his forearm. Finally, the patient with the neurofibroma which had mandated resection of a segment of the ulnar nerve and reconstruction with sural nerve grafts showed improvement in pain, but presented diminished strength and sensibility in the ulnar nerve territory.

DISCUSSION

Carefully going through all the literature concerning BP tumors since 1970, less than 800 cases are found

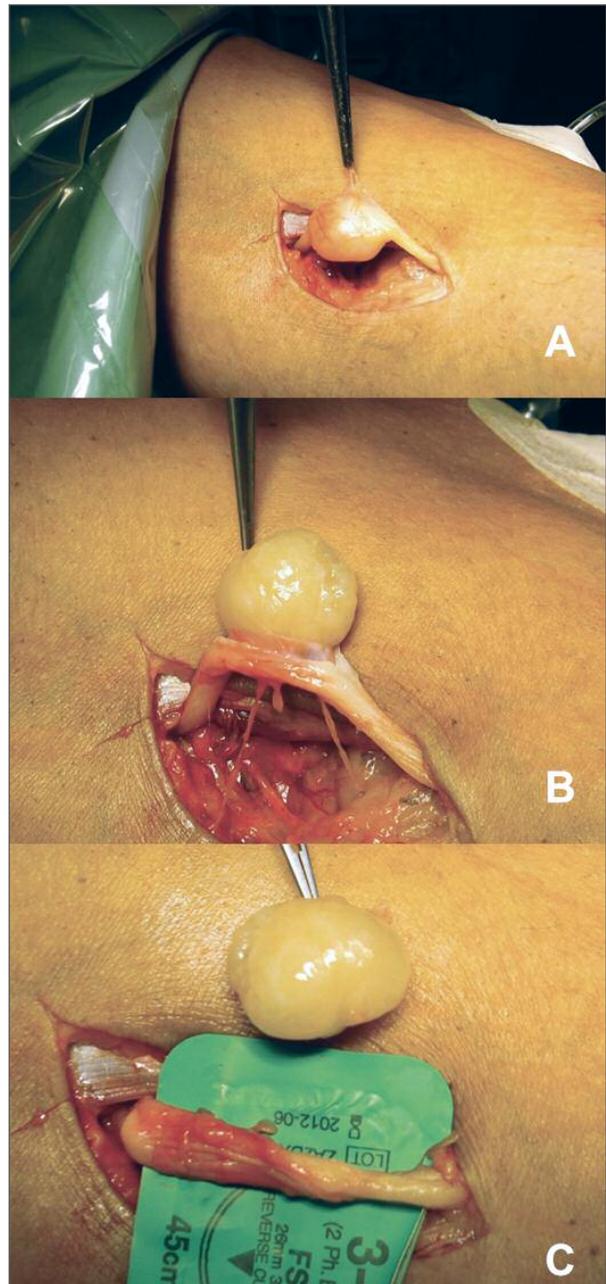


FIGURE 1. Intraoperative view of the extirpation of a peripheral nerve sheath tumor in the proximal portion of the radial nerve in the posterior aspect of the arm. **A)** On exposure of the nerve, the tumor was located peripherally. **B)** It was possible to isolate the peripheral nerve sheath tumor (later shown to be a Schwannoma) from the main nerve trunk, and a fibrous sheath resembling a fibrous capsule facilitated dissection. **C)** The tumor was dissected from the radial nerve leaving it intact

worldwide^{3,4}. These cases are mainly reported in single case reports or small series. There are only 11 papers with revisions of more than 15 patients^{3,4}. As these tu-

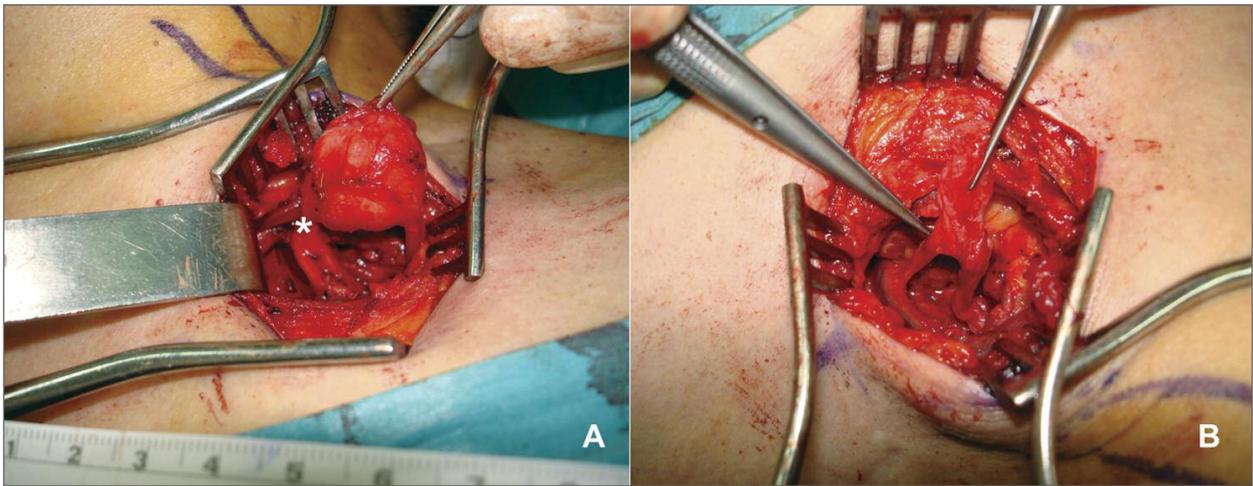


FIGURE 2. Intraoperative view of the excision of a Schwannoma of the supraclavicular nerve (marked with the asterisk) through a supraclavicular approach. A) The tumor was not eccentrically placed in the nerve. B) The tumor had to be dissected between the fascicles of the nerve under magnifying loupes, preserving nerve continuity

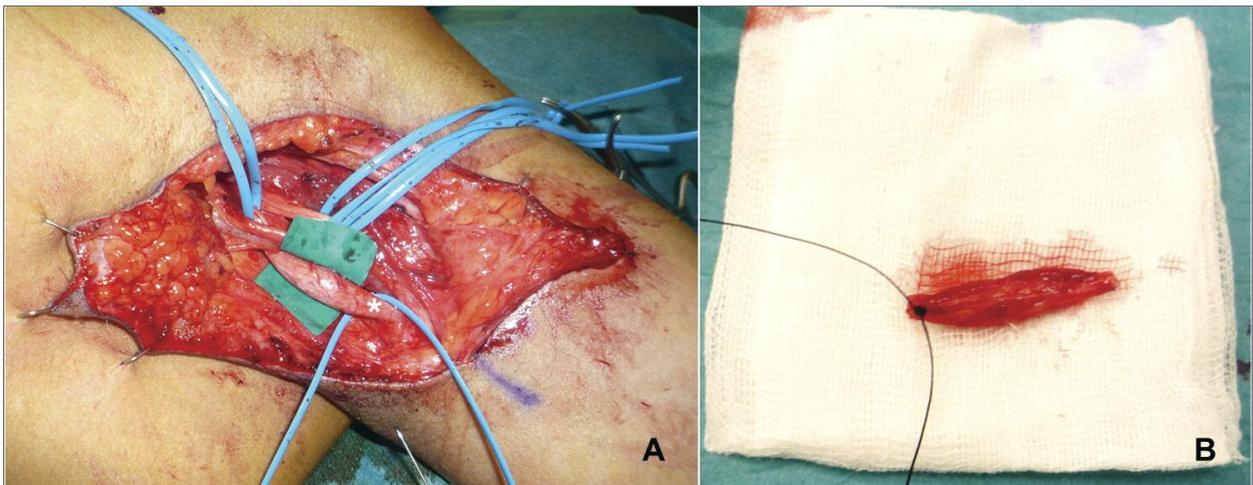


FIGURE 3. Intraoperative view of a fusiform neurofibroma located at the origin of the ulnar nerve (marked with an asterisk). A) The tumor was located inside the nerve, without any cleavage plane with the healthy nerve tissue. Intra-neural dissection proved to be impossible. B) The segment of the nerve involved by the tumor was excised

mors are so rare, it is unanimously accepted that these patients should be referred to specialized centers^{2,4-8}.

The mean age of our patient population at diagnosis (40 years) was in accordance with the largest series published, which presented an average value of 42 years². However, contrarily to most larger series, males were over-represented (4 males to one female)^{2,3}. This may be due to the relatively small number of patients in our series.

The most common symptoms presented by our patients were similar to what has generally been reported^{2,3,9}. In fact, most authors describe that most BP tu-

mors patients present with pain, sensibility changes, and a palpable mass^{2,3}. Tinel sign which is defined as an electric shock-like experience in the territory of the nerve on tapping the nerve is pathognomonic of nerve lesion and should be considered diagnostic of a nerve tumor, if no prior trauma occurred^{3,9-11}.

However, not always is it easy to establish this diagnosis, and BP tumors are frequently mistaken for other more common neoplasms in the BP region, namely lipomas, ganglion cysts, lymphadenopathies, brachial cleft cysts, desmoid tumors, cavernous angiomas, hemangiomas or lymphangiomas^{3,9}. In the

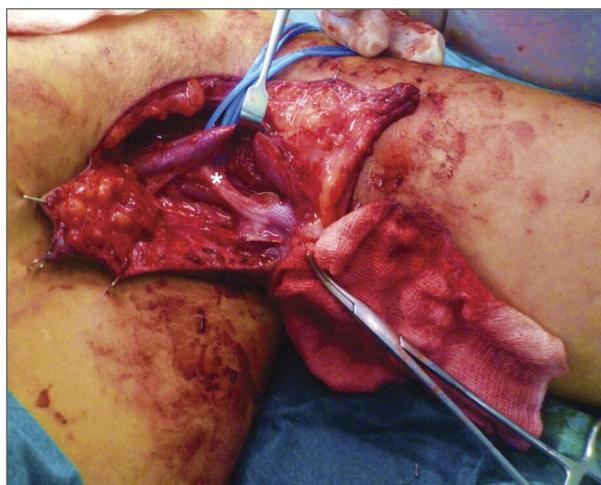


FIGURE 4. Intraoperative view of the patient presented in Figure 3. The nerve defect resulting from the nerve segment extirpation was reconstructed with 4 cables of autologous sural nerve graft.

work of Desai, 12% of patients had been initially misdiagnosed as having non-nerve tumors, and surgery with curative intent had been abandoned intra-operatively in favor of lesion biopsy by referring doctors³.

Most primary BP tumors are benign^{2,3,9}. More than 80% of these tumors are schwannomas or neurofibromas^{2,3,9}. Other examples of benign primary BP tumors are plexiform neurofibromas and ganglioneuromas^{2,3,9}. Primary malignant BP tumors correspond to less than 10% of all BP tumors, and are usually rapid growing, fixed lesions, associated with rapidly aggravating neurological deficits². They include neurogenic sarcomas and neurofibrosarcomas^{2,3,9}. Secondary BP tumors are more commonly Pancoast tumors or metastasis from the lung or breast^{2,3,9}.

In our series, 4 patients had schwannomas and one patient had a neurofibroma. Most authors report a larger proportion of neurofibromas over schwannomas^{2,5}. However, other series have also found a larger number of schwannomas, as we did^{3,4,9}. For example, Desai in a recent review of 115 benign brachial plexus tumors identified 70 schwannomas and 45 neurofibromas³.

Schwannomas are so named because they arise from Schwann cells within the endoneurium. As they grow, they progressively stretch the perineurium and epineurium that in turn gradually encapsulate the tumor. Hence, clinically, they tend to be tightly adherent to only one or two nerve fascicles sparing the remainder of the nerve^{2,3,9}. In contrast, the neurofibromas originate from cells of the epineurium and are there-

fore not encapsulated. As these latter tumors grow, they come to involve most of the nerve fascicles, resulting in frequent neurological deficits after tumor removal^{2,3,9}. Neurofibromas can be found to be solitary or multiple^{2,3,9}. The majority of patients with multiple neurofibromas have neurofibromatosis^{2,3,9}.

The preferred image test to visualize the tumor and its relations to neighborhood structures is MRI⁹. However, this test lacks accuracy in the distinction between primary benign and malignant BP tumors, and even in the distinction between schwannomas and neurofibromas⁹. If a malignancy is suspected, a positron emission tomography (PET) scan can be useful⁹. Recently, there has been great interest in MRI neurography, as it allows visualization of the entire course of nerves and its relations with the tumor^{9,12-16}. CT scan is the best method to assess bony involvement^{2,3,9}.

Regarding pre-operative electroneuromyography (EMG), most authors argue that it adds little to a complete medical history, physical examination and imagiological examination^{3,17}. However, other authors argue that EMG is useful in identifying subclinical deficits in involved BP elements and may even detect significant denervation usually associated with BP malignant tumors^{4,6}.

Fine-needle aspiration or core pre-therapeutic biopsy is generally discouraged because these methods lack accuracy, and can cause iatrogenic neurological deficits and/or vascular injuries. Furthermore, they are known to induce significant fibrosis that will make definitive surgical treatment more difficult and hazardous. Finally, there is the potential risk of tumor seeding along the needle track^{2,3,9}.

After a presumptive diagnosis of BP tumor is made, the main problem is to decide to keep the patient under close observation, with regular visits to the clinic, or, on the contrary, to operate¹⁸. It is generally accepted that this decision will be made based on the perceived risks and benefits of either option, according to patient's wishes¹⁸. Disfigurement associated with tumor growth, pain or progressive neurological deficits are usually considered indications for surgery. The latter two signs are associated with a higher risk of malignancy, particularly if a neurocutaneous syndrome is present^{2,3,18}. Most authors suggest surgery if the lesion is symptomatic or progressing in size^{2,3,12}. If the tumor is stationary and not associated with neurological dysfunction a conservative approach can be taken^{2,3,12}.

Most authors agree that complete extirpation of a schwannoma is more easily achieved than with neu-

rofibromas, because the latter tend to be more adherent to the core of the nerve, often times with a scant or absent cleavage plane^{3,18,19}. Most authors argue that in either case it is of utmost importance to preserve the largest number of functioning motor fibers, even if this means leaving small amounts of residual benign tumor or tumor capsule³.

Notwithstanding, it should be noted that surgery is far from being devoid of complications¹⁸. Besides anesthetic and wound healing problems, which are increasingly rare, 10 to 17% of all patients operated due to BP tumors present new neurological deficits of variable severity postoperatively^{4,18}. According to Kehoe *et al.*, for example, the number of neurological deficits doubled after surgery in a series of 15 patients²⁰. Other complication that has been described, although seldom encountered, is the intra-operative damage of major vascular structures, like the subclavian artery⁹. Overall, surgery benefits have been shown to outweigh risks, and surgery is consensually recommended in selected patients^{2,3,9}.

CONCLUSION

It is of utmost importance that every physician who deals with rheumatological patients is familiarized with BP tumors, in order to correctly diagnose them, through a targeted history and examination, combined with the use of appropriate ancillary tests. After establishing the diagnosis, the physician may opt to follow the patient or to refer the patient to a surgeon with experience in this field, in order to minimize operative risks and to increase the odds of complete resection of the tumor^{3,21,22}.

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