Benign Peripheral Nerve Sheath Tumors: A 10 Year Hospital Based Study

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Abstract

Background: Benign nerve sheath tumours present as isolated soft tissue masses and are classified as solitary neurofibroma and schwannoma. They cause virtually identical symptoms and signs, and there are no clinically distinct pathognomonic features. Failure to appreciate the possibility and the nature of these tumours at the time of surgery may lead to a catastrophic loss of function for the patient. Aim of the study is to analyse peripheral nerve sheath tumours in reference to; Hospital based incidence, age and sex distribution peripheral nerve involved most preoperative diagnostic tool efficacy histopathological variations and surgical outcome. **Materials and Methods:** the study was carried out at SKIMS Srinagar from January 2005 to October 2014.Study included 49 patients, with an average age of 38 years (range 7-65 years). The diagnostic tools used were FNAC, NCV, USG, & MRI for location & provisional diagnosis, surgical excision was performed and final diagnosis achieved by HPE. **Results**: The incidence of isolated peripheral nerve sheath tumors in our study was 4.9 per year. In case of schwannomas upper limbs were involved more frequently than lower limbs (2:1), while neurofibromas were more evenly distributed (1.16:1). Presenting symptoms were mass, pain, weakness, dysesthesia an average follow-up of 6 months after surgery pain syndromes had complete resolution in 66% vs 44%, paraesthesia 80% vs 50% and motor strength restoration to normal in 45.5% vs 6.25% patients in schwannomas and neurofibromas respectively. **Conclusion:** Meticulous dissection with Intracapsular tumour removal under magnification can achieve complete tumour removal without neurological loss or recurrence.

Key Words: schwannoma, neurofibroma, microsurgical excision, paraesthesia, mass

Introduction

Tumors arising from peripheral nerves are a rare cause of lumps in the limbs. Some are easily diagnosed from characteristic symptoms, an isolated mass and distal neurological sign, but in others the diagnosis is more difficult. Benign nerve sheath tumors are classified as, solitary neurofibroma and schwannoma (neurilemmoma). Solitary neurofibromas and schwannomas cause virtually identical symptoms and

signs, and there are no clinically distinct pathognomonic features. Either may present as an isolated soft-tissue mass of uncertain origin and duration. Failure to appreciate the possibility and the nature of these tumours at the time of surgery may lead to a catastrophic loss of function for the patient. Such tumours can present to many different medical and surgical specialties. We have reviewed only tumours of the peripheral nerves referred to neurosurgeon.[1] The purpose of the study was to analyse benign peripheral nerve sheath tumours (schwannomas and neurofibromas) in reference to hospital-based incidence and prevalence, age of presentation, peripheral nerve involved most, management options, histopathological variations and surgical outcome.

Materials and Method

The patients of peripheral nerve sheath tumours (C1-S5) who have been treated from January 2005 to May 2012 were analysed in the Department of Medical Records retrospectively, while those patients who were admitted from June, 2012 to October 2014, were analysed with respect to hospital-based incidence and prevalence, age and sex distribution, peripheral nerve involved most, preoperative diagnostic tool efficacy, histopathological variations and surgical outcome. The diagnostic tools like FNAC, NCV, USG & MRI were used in provisional diagnosis and location. All tumours were excised completely with intracapsular excision using loupe magnification and microsurgical dissection techniques and final diagnosis achieved by histopathological examination. Malignant peripheral nerve sheath tumors are excluded from the study.

Results

The patients of peripheral nerve sheath tumors (schwannoma and neurofibroma) who have been treated from January 2005 to December 2012 were analysed in the Department of Medical Records retrospectively, while those patients who were admitted from January, 2013 to October 2014, were analysed in our hospital i.e. SKIMS, which is the apex institute of the J & K State and following observations were made.

The incidence of isolated peripheral-nerve sheath tumors is low, in our study 4.9 per years. Among schwannomas eighteen patients were male and 15 females (M: F ratio, 1.2:1) with an average age of 38.03 years (range 15-65 years). While among neurofibromas eight patients were male and 8 females (M: F ratio, 1: 1) with an average age of 39.1 years (range 7-60 years). The age at presentation did not correlate with type of tumour with 60% of schwannomas (range 15-65 years). and 63% of solitary neurofibromas (range 7-60 years), presenting in patients between 30 and 59 years of age. In case of schwannomas upper limbs were involved more frequently than lower limbs (2:1), while the neurofibromas were more evenly distributed (1.16:1). Most common nerve involved in upper limbs was median nerve (10), followed by ulnar (7), radial (6), brachial plexus (5), supraclavicular (2), lateral cutaneous nerve of arm (2). while in lower limbs superficial peroneal nerve (4) and medial cutaneous nerve of thigh (4) were affected equally followed by lateral cutaneous nerve of thigh (3), deep peroneal nerve (3), anterior tibial nerve (2), and sural nerve (1). In case of schwannomas average duration of symptoms was 5.1 years (range 6 months to 11 years). All patients presented with a palpable mass, 15 (45.45%) with pain syndromes. Ten patients (30.30%) had refered dysaesthesia on percussion over the mass.

Pre-operative motor deficit was reported 11 (33.33%) patients, with G IV power in 6 and G III in 5 patients. While as the average duration of symptoms in neurofibromas was 3.1 years (range 6 months to 8 years). In all patients the presenting symptom was mass. Of the 16 patients, 9 (56.25%) presented with pain, six patients (37.5%) reported dysaesthesia on percussion over the mass. Pre-operative motor deficit was reported in 7 (43.75%), with G IV power in 4 and G III in 3 patients as shown in Table 1 & 2.

High-resolution ultrasound was done in 40 patients which showed masses of low echogenicity with distal enhancement. FNAC was done in 20 patients, especially those with diagnostic uncertainty,14 retrospectively and 6 prospectively out of which 11 were Schwannomas, 6 Neurofibromas and 3 were

labelled as inconclusive with diagnostic efficacy of 85%.NCV test was done in 38 patients, with 24 patients (18 schwannomas & 6 neurofibromas) showing delayed sensory nerve conduction velocity while 8 patients, (5 schwannomas & 3 neurofibromas) had delayed motor nerve conduction velocity .MRI is capable of imaging the tumour, its capsule and the nerve from which the tumour arises. The signal on T1-weighted images has usually been homogeneously iso-intense, on T2-weighted images homogeneously hyperintense. Contrast enhancement is usually strong or inhomogeneous in the center of the lesion (Figure A-I showing pre operative, MRI and intraoperative pictures of the lesions). In our 31 cases (10 with Neurofibroma), we did not find any differences related to the frequency of each type of enhancement among patients. Out of 31 patients scanned four had lipoma and two turned out to be synovial cysts with the diagnostic efficacy of MRI being 80.6%. Using the above diagnostic modalities together preoperative diagnosis was possible in 69.79% of cases.

Tumor size ranged from 1 to 6 cm. One of our patients required re-operation along the same nerve but at a different level (ulnar) after four years of initial operation. The immediate post operative problems although transient were noted in 6of 49 (12.2%) patients these include new onset of pain in two patients among schwanomas while among neurofibromas there was worsening of pain in one, paresthesia in one, new onset motor weakness in one patient and mixed-nerve deficit in one.

At an average follow-up of 6 months Pre and Post Operative Status of symptoms and Motor power Grade is depicted in table 1 and table 2. Figure A-I showing pre operative, MRI and intraoperative pictures of the lesions

Table 1. Pre and Post Operative Status of symptoms

SCHWANNOMA					
	Preoperative	Postoperative			
Symptom	Total No.	Resolved	Improved	Unchanged	Worsened
Pain	15	10(66.6%)	4(26.6%)	1(6.6%)	0
Paresthesia	10	8(80%)	2(20%)	0	0
NEUROFIBROMA					
Pain	9	4(44.4%)	2(22.2%)	3(33.3%)	0
Paresthesia	6	3(50%)	2(33.3%)	1(16.6%)	0

Table 2. Pre- and Post-operative Status of Motor Grade

SCHWANNOMA							NEUROFIBROMA						
	Pre-operative Motor Grade					Total		Pre-operative Motor Grade					Total
Post-operative Motor Grade	5	4	3	2	1		Post-operative Motor Grade	5	4	3	2	1	
5	22	4	1	0	0	27	5	9	1	0	0	0	10
4	0	2	1	0	0	3	4	0	3	1	0	0	4
3	0	0	3	0	0	3	3	0	0	2	0	0	2
2	0	0	0	0	0	0	2	0	0	0	0	0	0
1	0	0	0	0	0	0	1	0	0	0	0	0	0
Total	22	6	5	0	0	33	Total	9	4	3	0	0	16

Discussion

The incidence of isolated peripheral-nerve sheath tumors is low, in our study 4.9 per year, which is consistent with other studies as Jenkins (I952) [2] described only three seen in 12 years at the Hammersmith Hospital.In our study in case of schwannomas upper limbs were involved more frequently than lower limbs (2:1), while the neurofibromas were more evenly distributed (1.16:1) which is

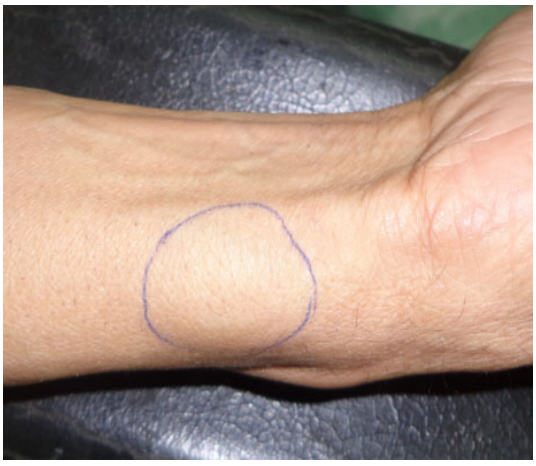
consistent with the study done by Das Gupta et al (1969)[3]. The neurofibromas were more evenly distributed. This imbalance contrasts with the incidence of soft-tissue tumors of the musculoskeletal system in general; for these the lower limb is the more common site. The imbalance may relate to the more prominent function of the nerves in the upper limb, but both Stack (1960)[4] and Strickland and Steichen (1977) [5] found that less than 5% of hand tumors had arisen from nerves. In our study the age at presentation did not correlate with tumor type, 60% of schwannomas and 63% of solitary neurofibromas were in patients presenting between 30 and 60 years of age. which is consistent with the study done by Das Gupta et al (1969)[3].In our study tumors were particularly less common in children with only three patients under 16 years of age, youngest being 7 years old, two with schwannoma and one with neurofibroma which is consistent with the study done by Ruda et al (1991)[6] who reported only one schwannoma in a 12-year-old child and Das Gupta et al (1969)[3] who reported only four patients under 18 years of age, two with schwannoma and two with neurofibroma. Anatomical considerations suggest that a nerve-related mass should be in the line of a nerve trunk, mobile in the transverse plane, but tethered in the longitudinal axis of the limb. Such clinical findings, however, do not lead to a diagnosis more specific than 'peripheral-nerve tumor'.Most of our patients presented after a long delay, All patients presented with a palpable mass , with a palpable mass as their only symptom in nearly half of the patients, In case of schwannomas average duration of symptoms was 5.1 years (range 6 months to 11 years), While as the average duration of symptoms in neurofibromas was 3.1 years (range 6 months to 8 years), as reported for other series (Holdsworth 1985)[7] .Spiegl et al (1986)[8] .Nerve irritation is usually apparent as paraesthesia with a positive Tinel sign, which was positive in 81.5% of cases in our study. Spiegl et al (1986)[8] reported that 63 of 76 patients (83%) with a schwannoma in the lower limb complained of pain. In our study 48.9% of the palpable masses were associated with pain, or tenderness, 32.6% with paresthesia and weakness in 36.7%. Using the below mentioned diagnostic modalities together preoperative diagnosis was possible in 69.79% of cases. Poor diagnostic accuracy increases operative uncertainty and poor outcome. Most patients in our study were evalvated using sophisticated imaging techniques and FNAC,which is in against to study by Nicholas J.S.Kehoe et al.[1] in which preoperative diagnosis of a nerve tumor was made in only one quarter of cases due to lack of sophisticated imaging techniques .New imaging techniques give an indication of the precise pathology Ultrasound is well established but little has been published in relation to peripheral nerves and their small tumors (Hughes and Wilson 1986)[9]. In our study high-resolution ultrasound was done in 40 patients which showed masses of low echogenicity with distal enhancement which is consistent with study by Obayashi, Itoh and Nakano 1987 [10] Fornage 1988)[11]. FNAC was done in 20 patients, especially those with diagnostic uncertainty,14 retrospectively and 6 prospectively out of which 11 were Schwannomas,6 Neurofibromas and 3 were labeled as inconclusive, with diagnostic efficacy of 85%. No new neurological deficit was reported in any of the patients subjected to FNAC, which is consistent with study by Domanski HA, Akerman M, Engellau J, Gustafson P, Mertens F, Rydholm A[12].NCV test was done in 38 patients, with 24 patients (18 schwannomas & 6 neurofibromas) showing delayed sensory nerve conduction velocity while 8 (5 schwannomas & 3 neurofibromas) had delayed motor nerve conduction velocity. Nerve-conduction studies are useful to exclude other pathology such as compression syndromes, but the function of nerve fibrils is rarely disrupted by the presence of a discrete, benign schwannoma or neurofibroma[1].MRI is the best imaging method since it is non-invasive and provides both longitudinal and transverse sections.MRI is capable of imaging the tumor, its capsule and the nerve from which the tumor arises. When the parent nerve is identified, an eccentrically positioned lesion (in relation to the nerve) suggests a schwannoma whereas a centrally located mass suggests a neurofibroma. The signal on T1-weighted images is usually homogeneously iso-intense, on T2-weighted images homogeneously hyperintense.Contrast enhancement is usually strong or inhomogeneous in the center of the lesion[13].In our 31 cases (10 with Neurofibroma), we did not find any differences related to the frequency of each type of enhancement among patients.Out of 31 patients scanned four had lipoma and two turned out to be synovial cysts with the diagnostic efficacy of MRI being 80.6%.The excision of a solitary nerve tumor is the only way to obtain exact tissue diagnosis. The characteristic appearance of

a schwannoma is of an ovoid mass, often smooth, shiny and greyish in colour, with nerve fibrils spread thinly and diffusely over its surface. A neurofibroma, by contrast. is more intimately involved with the nerve fibrils and any attempt at removal must be balanced against the danger of structural damage to the nerve. This must be considered before the surgical exploration of a nerve and particularly for proximally positioned tumors. Bonney (1986)[14] and Birch et al (1991)[15] have stressed the hazard of limited exposures near important axial structures and especially the brachial plexus. They condemn the practice of biopsying nerves and paraneural structures. In our study all tumors were excised completely with intracapsular excision using loupe magnification and microsurgical dissection techniques . Tumor size ranged from 1 to 6 cm. The immediate post operative problems although transient were noted in 6 of 49 (12.2%) patients these include new onset of pain in two patients among schwannomas while among neurofibromas there was worsening of pain in one, paresthesia in one, new onset motor weakness in one patient and mixed-nerve deficit in one.Which is consistent with the study done by Nicholas j. s. kehoe, et al[1].

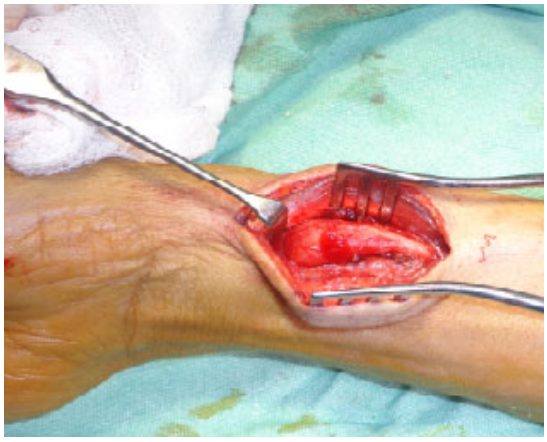
As seen in table 1 and 2 no patients in our series had an increase in nerve dysfunction after surgery.Pain was graded using Visual Analog Scale ,strength and sensation were graded from 0 to 5 using the grading scale reported by Donner[16].This is comparable to the study done by M. J. Park,et al who evaluated neurological deficit after surgical enucleation of schwannomas of the upper limb[17].Another study with similar postoperative results was reported by Kim SM, Seo SW, Lee JY, Sung KS but with a mean follow up of 58.8 months (32-79 months)[18].In our study we encountered three patients showing predominant features of neurofibroma with distinct, often nodular regions of classical schwannomatous differentiation ,two patients being male and one female, Cavallazzi et al (1988)[19] has also reported the combination of a schwannoma and neurofibroma on the same peripheral nerve trunk (median).One of our patients required re-operation along the same nerve but at a different level (ulnar) after four years of initial operation. Lewis, Nannini and Cocke (1981) [20] reported a patient with multiple schwannoma in hand and forearm. Multiple tumors however are rare except in von Recklinghausen's disease.

Conclusion

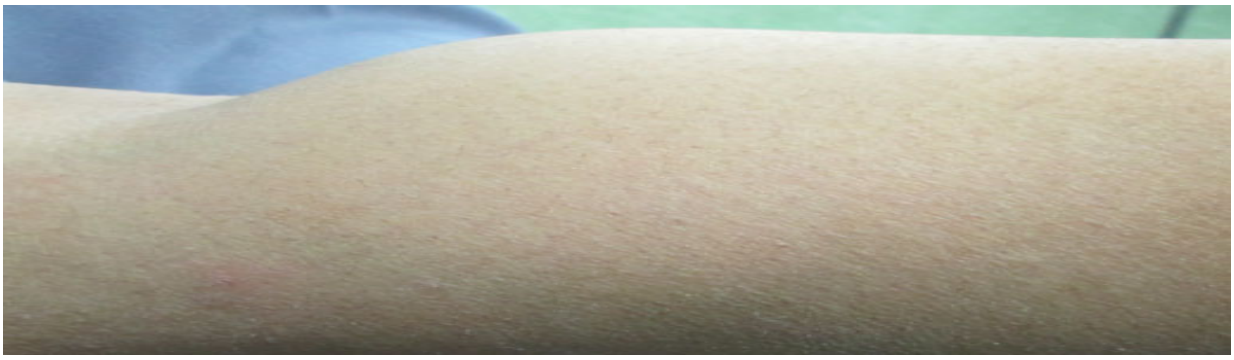
Benign nerve sheath tumors (Schwannoma & neurofibromas), are the most frequent neural tumor of the peripheral nerves usually presenting as an innocuous mass with or without pain, paresthesia or weakness. The lesions when present along the distribution of peripheral nerves should be carefully evalvated to prevent any catostrphic nerve injury. Meticulous dissection with micro surgical intracapsular excision of the lesion helps in preserving maximum nerve fibers and thereby preventing/limiting postoperative neurodeficit and decreasing the chances of recurrence.



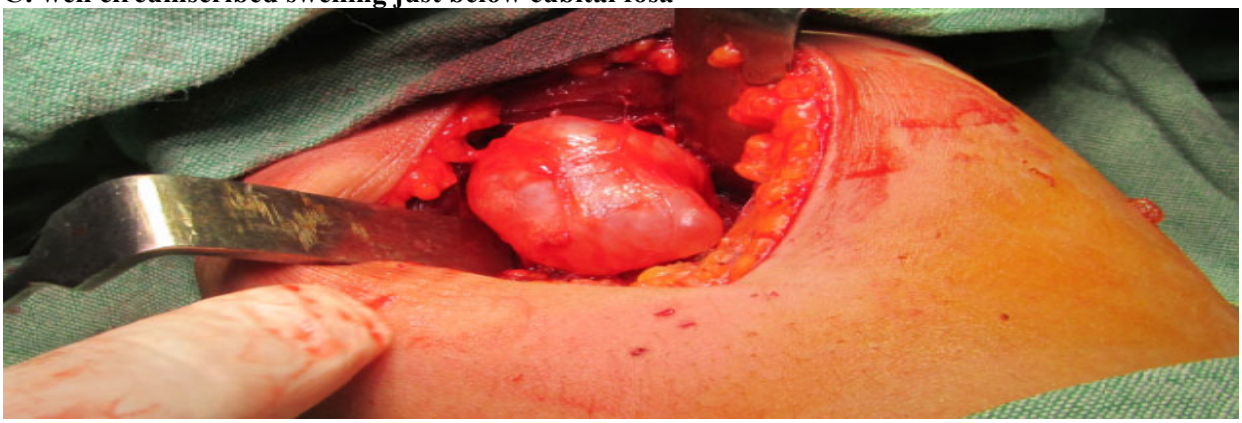
A:well circumscribed swelling on ulner border of left hand of ulner nerve



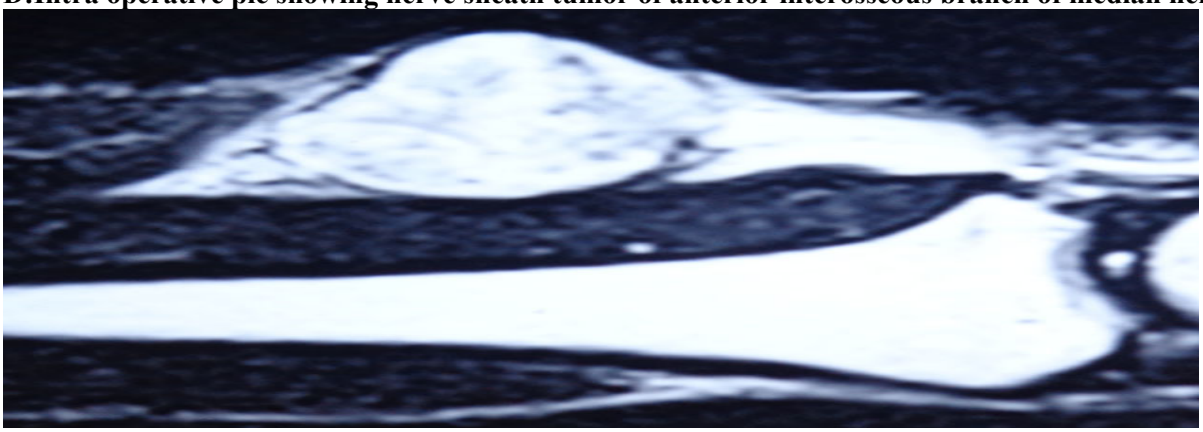
B: Intra operative pic showing nerve sheath tumor



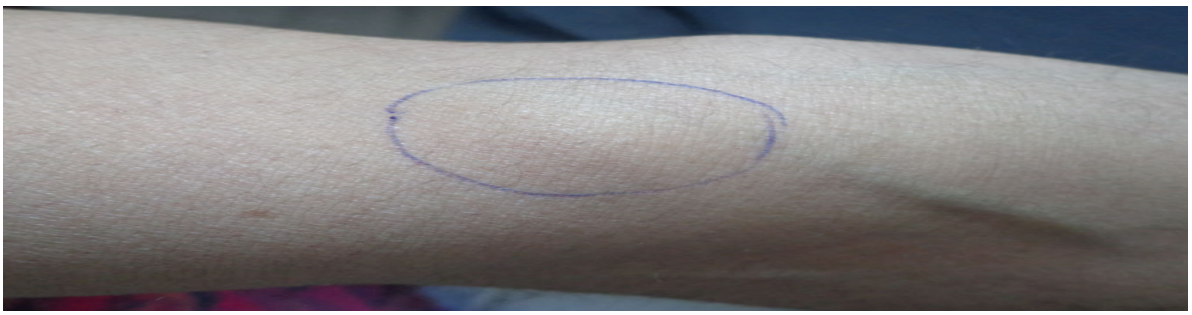
C: well circumscribed swelling just below cubital fosa



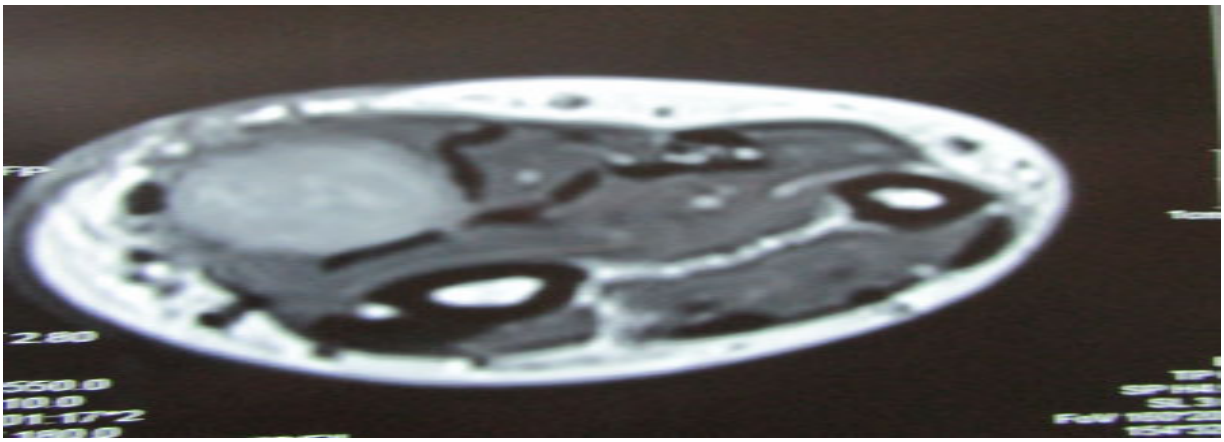
D:Intra operative pic showing nerve sheath tumor of anterior interosseous branch of median nerve



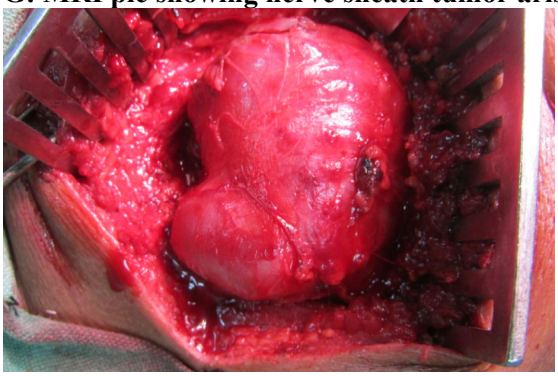
E:MRI showing nerve sheath tumor arising from anterior interosseous branch of median nerve



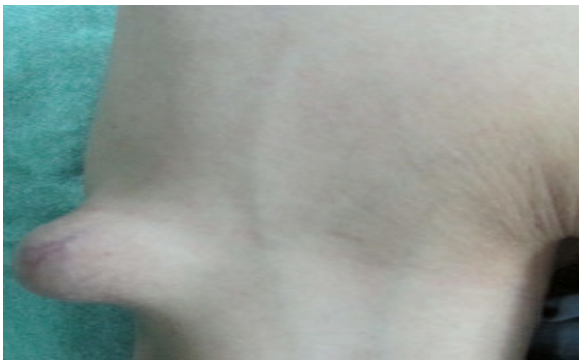
F:Ill defined swelling just above cubital fosa laterally



G: MRI pic showing nerve sheath tumor arising from Radial nerve



H: Intra operative pic showing nerve sheath tumor of radial nerve



I: Nerve sheath tumor of cutaneous branch of Axillary nerve

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