Surgical Management of Peripheral Nerve Tumors: Experience at a Tertiary Care Centre

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Abstract

Benign and malignant peripheral nerve tumors are rare. In this study, the authors present their experience in operative management of various peripheral nerve tumors and analyze the results of treatment.

This study was conducted on patients with peripheral nerve tumors admitted to a tertiary care hospital.

Twelve patients with 13 tumors were admitted, of which 9 were benign and 4 were malignant. All the patients underwent surgical excision of the tumors. Benign tumors had a good outcome and malignant tumors had to undergo tumor excision, but no nerve reconstruction was possible due to surrounding tissue infiltration.

Treatment for schwannoma is complete removal, whereas a malignant peripheral nerve sheath tumor requires a wide resection to obtain a favorable prognosis.

Keywords
- peripheral nerve
- tumor
- histopathology
- good outcome
- benign
- malignant

Introduction

Nerve tumors represent uncommon extremity tumors and include benign peripheral nerve sheath tumors (BPNSTs) and malignant peripheral nerve sheath tumors (MPNSTs). Benign nerve tumors are slow growing and are minimally symptomatic, whereas malignant masses are known for a more aggressive growth pattern.[1] In this study, the authors present their experience in the operative management of various peripheral nerve tumors and analyze the results of the treatment.

Materials and Methods

This is a cross-sectional study of patients who underwent surgery for peripheral nerve tumors in the department of neurosurgery of a tertiary care hospital between 2017 and 2024. Data were collected from the patients' hospital records.

Results

This study included 12 patients, in whom 13 tumors were excised from extremities between 2017 and 2024. Eleven tumors from the upper limbs and two tumors from lower limbs were excised. Nine were benign mases, of which six were schwannomas and three were neurofibromas. Four were malignant masses, of which two were parachordoma and two were MPNSTs. Thorough clinical evaluation and imaging were done for all the cases. All the patients had satisfactory outcome after surgery. ►**Table 1** presents the characteristics of the patients included in this series.

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Table 1 Patient's characteristics

Case no.	Age (y)/sex	Location	Originating nerve	Surgery	Histopathology
1	46/male	Right axilla	Axillary nerve	Enucleation	Neurofibroma
2	57/male	Left axilla	Radial nerve	Enucleation	Schwannoma
3	55/male	Left axilla	Radial nerve	Tumor excision	MPNST
4	61/male	Left axilla	Ulnar nerve	Enucleation	Schwannoma
5	55/male	Right shoulder	Suprascapular nerve	Enucleation	Neurofibroma
6	45/male	Left arm	Median nerve	Tumor excision with sural nerve grafting	Schwannoma
7	9/female	Right forearm	Ulnar nerve	Enucleation	Schwannoma
8	35/male	Left forearm	Unnamed nerve	Enucleation	Neurofibroma
9	62/female	Right forearm	Median nerve	Enucleation	Parachordoma
10	15/male	Left wrist	Ulnar nerve	Enucleation	Schwannoma
11	54/male	Right leg	Superficial peroneal nerve	Enucleation	Schwannoma
12	36/female	Left ankle	Tibial nerve	Tumor excision	MPNST

Abbreviation: MPNMST, malignant peripheral nerve sheath tumor.

Illustrated Cases

Tumors in the Axilla

Case 1 (Axillary Nerve Neurofibroma with Neurofibromatosis 1)
A 46-year-old man presented with a history of tightness of all four limbs (right > left) associated with a history of decreased hand grip in the right hand when compared with the left side (►**Fig. 1**). The patient had an associated history of weakness in the right upper and lower limbs with a history of right axillary swelling for 3 months. Multiple small subcutaneous neurofibromas were noted over the body. Magnetic resonance imaging (MRI) showed an enhancing extra axial soft tissue lesion at the C1–C2 level filling both neural foramina compressing and insinuating the adjacent spinal cord. An enhancing large nodular lesion was seen in the right axillary region measuring $60 \times 35 \times 30$ mm with a central nonenhancing area, suggestive of a nerve sheath tumor. Right axillary exploration and excision of the axillary tumor was done. The tumor was bilobed and abutting the chest wall in the right axilla. The tumor was grayish yellow, mildly vascular, soft to firm in consistency, and arising from the right axillary nerve. Gross total excision of the tumor was done. Biopsy of the lesion confirmed neurofibroma.

Case 2 (Radial Nerve Schwannoma)
A 57-year-old man presented with left axillary swelling for 10 years and an electric shock–like sensation over the left forearm and hand for 4 years (►**Fig. 2**). There was a 4×4 cm globular, firm, nontender swelling over the left axillary region with restricted mobility along the direction of the brachial plexus with an electric shock–like sensation present over the distribution of the radial nerve over the thumb and the dorsum of the left hand. MRI showed a well-defined T1-weighted (T1W) isointense, T2W-hyperintense, heterogeneously contrast-enhancing lesion in the left axilla arising from the brachial plexus, abutting the left axillary artery anteriorly with maintained fat planes, compressing the axillary vein posteroinferiorly. Left axilla exploration and intracapsular enucleation of the tumor was done. A well-circumscribed globular, firm tumor was found with attachment to a nerve on its medial and lateral ends. The tumor was arising from a branch of the posterior cord of the brachial plexus. The median and ulnar nerves were identified at the superolateral aspect of the tumor and the radial nerve was identified at the medial aspect of the tumor with intraoperative direct nerve stimulation. The axillary artery and vein were found at the posterosuperior aspect of the tumor with good fat planes. Some flattened nerve fibers of the radial nerve were splayed over the medial surface of the tumor. Intracapsular enucleation of the tumor was done and thinned-out nerve fibers were preserved. Median, ulnar, and radial nerve functions were found preserved with nerve stimulation after tumor excision. Postoperatively, he developed weakness in left wrist extension with no other deficit in the left upper limb. A biopsy of the lesion confirmed schwannoma. The patient is currently under follow-up.

Case 3 (Radial Nerve MPNST)
A 55-year-old man presented with a swelling in the inner aspect of the left axilla for 1 year, which was progressively increasing in size and was associated with radiating pain to the thumb and the index finger (►**Fig. 3**). There was a swelling measuring approximately 10×4 cm in the left axilla. It was tender, slightly mobile on side to side with normal overlying skin. Left wrist drop was present. MRI showed an elongated well-defined fusiform-shaped lesion in the left axilla extending in the line of the radial nerve of the adjacent arm suggestive of a nerve sheath tumor. Computed tomography (CT) angiography of the left upper limb showed a well-defined fusiform lesion in the left axilla extending in the line of the radial nerve of the adjacent arm, with the lesion compressing the brachiocephalic vein and abutting

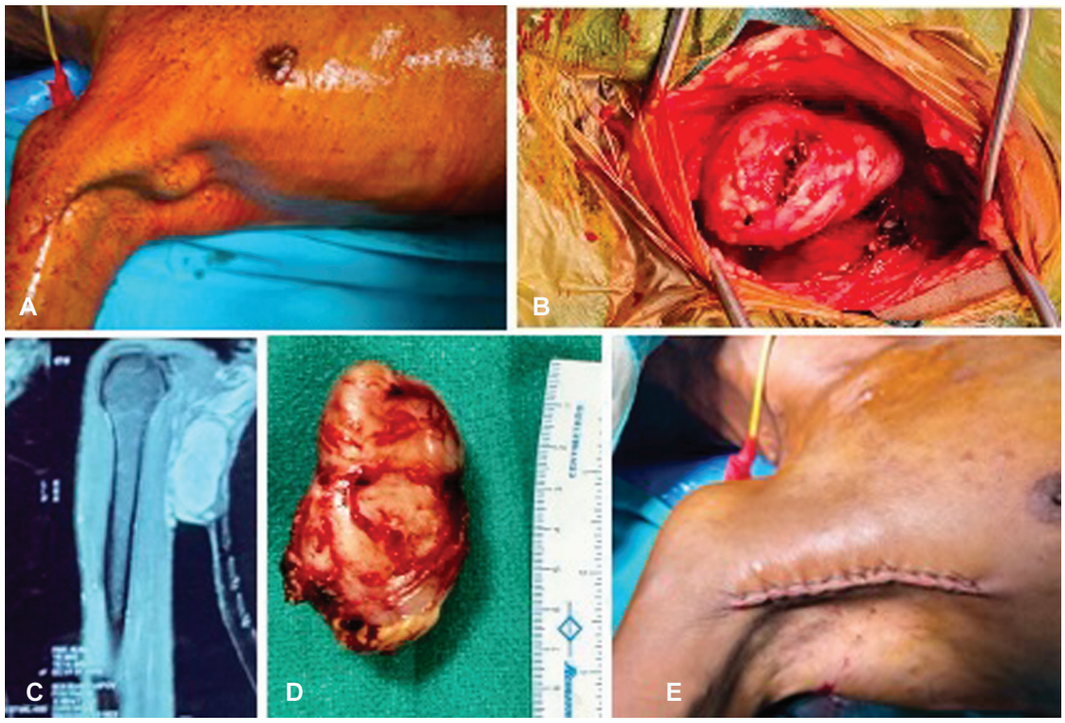

Fig. 1 Right axillary nerve schwannoma. (**A**) Preoperative clinical image. (**B**) Intraoperative image showing tumor arising from the right axillary nerve. (**C**) Magnetic resonance imaging of the axilla showing an enhancing tumor along the right axillary nerve. (**D**) Excised tumor specimen. (**E**) Postoperative image.

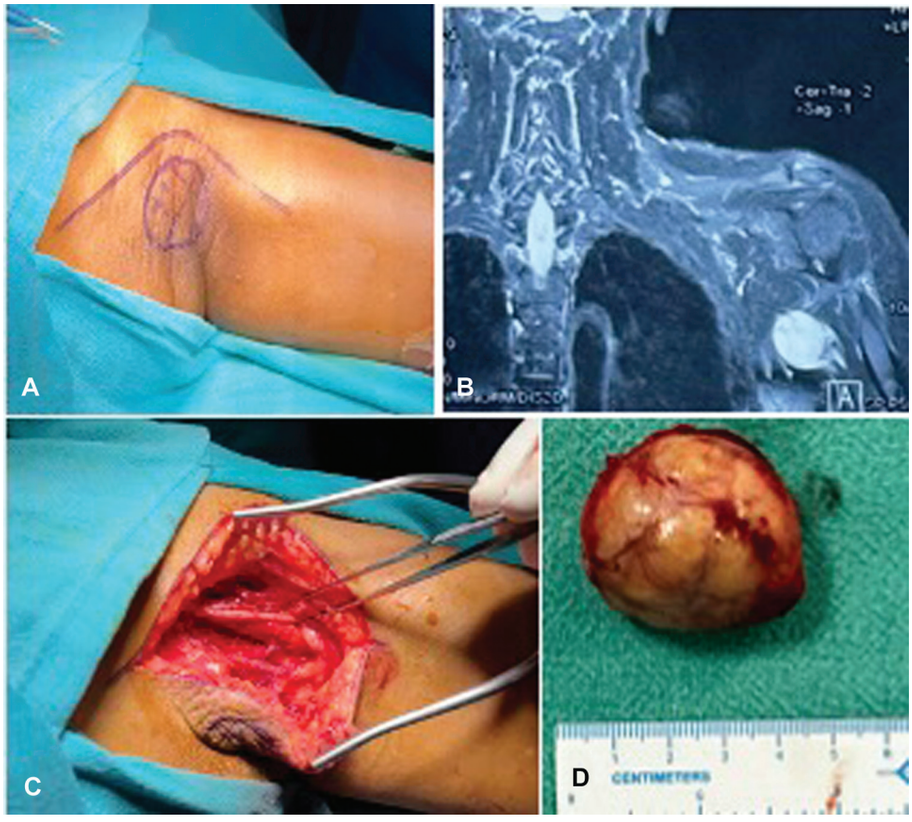

Fig. 2 Left radial nerve schwannoma. (**A**) Preoperative clinical image. (**B**) Magnetic resonance imaging of the axilla showing an enhancing tumor along the left radial nerve. (**C**) Intraoperative image showing a tumor arising from the left radial nerve. (**D**) Excised tumor specimen.

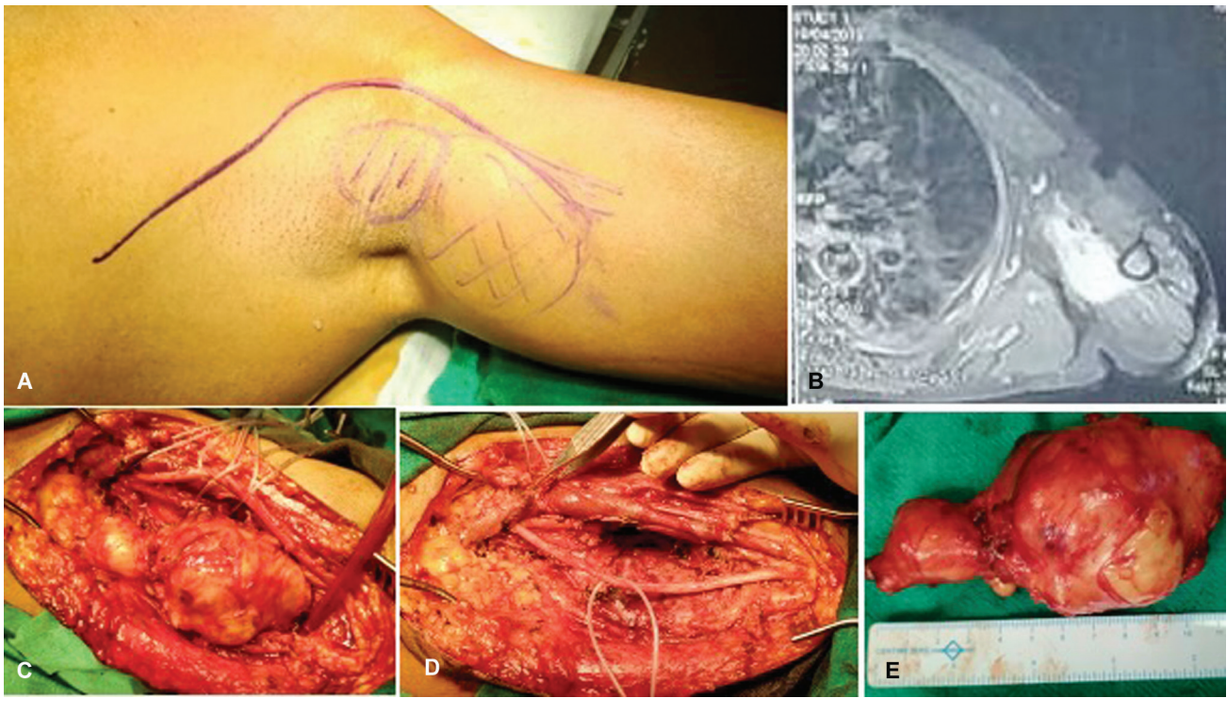

Fig. 3 Left radial nerve malignant peripheral nerve sheath tumor. (**A**) Preoperative clinical image. (**B**) Magnetic resonance imaging of the axilla showing enhancing tumor along the left radial nerve. (**C**) Intraoperative image showing a tumor arising from the left radial nerve. (**D**) Intraoperative image after excision of the left radial nerve tumor. (**E**) Excised tumor specimen.

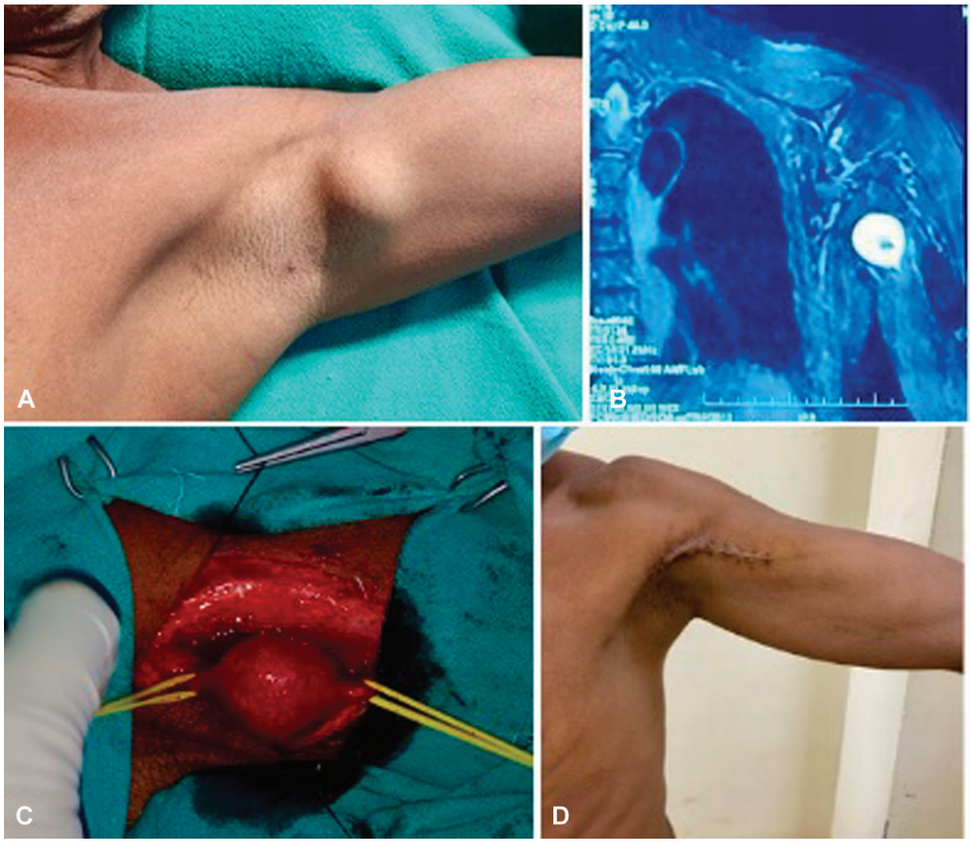

Fig. 4 Left ulnar nerve schwannoma. (**A**) Preoperative clinical image. (**B**) Magnetic resonance imaging of the axilla showing an enhancing tumor along left ulnar nerve. (**C**) Intraoperative image showing a tumor arising from the left ulnar nerve. (**D**) Postoperative image.

the circumflex humeral branch and the muscular branch of the left axillary artery. An exploration of the left axilla and excision of the tumor were done. The tumor was located in the left axilla and extended to the medial aspect of the left arm. The normal lateral cord and medial cord were identified lateral to the tumor. The axillary artery and vein along with the nerve to the biceps were identified and retracted from the surgical field by an infant feeding tube. The tumor was arising from the posterior cord. The tumor was bilobed, fusiform in shape, approximately 12 × 5 cm, and encompassing the whole circumference of the radial nerve. It was moderately vascular, firm with poor plane of demarcation from other neurovascular structures. Both proximal and distal ends of the tumor were identified and transfixed with Prolene 6–0 and excision of the tumor was done. Nerve reconstruction was not performed due to large nerve defect after tumor excision. Nerve monitoring was used to confirm the medial and lateral cords and preserved functionally. The postoperative period was uneventful. A biopsy of the lesion confirmed MPNST. Motor power remained the same as preop with no new deficit. The patient was followed up in radiation oncology for adjuvant radiotherapy.

Case 4 (Ulnar Nerve Schwannoma)
A 61-year-old man presented with a swelling in the inner aspect of the left axilla for the past 1 year, which was progressively increasing in size (►**Fig. 4**). Exploration and enucleation of the tumor were done. The tumor was arising from the ulnar nerve. A biopsy of the lesion confirmed schwannoma. The postoperative period was uneventful.

Tumors in the Shoulder

Case 5 (Suprascapular Nerve Neurofibroma)
A 55-year-old man presented with pain in the right shoulder for 4 months with no neurological deficit (►**Fig. 5**). MRI showed a well-defined enhancing soft tissue mass (29 mm × 16 mm) at the right coracoclavicular interval, suggestive of peripheral nerve sheath tumor. Ultrasonography (USG) guided fine needle aspiration cytology (FNAC) suggested benign spindle cell lesion. Right infraclavicular exploration and excision of the tumor were done using intraoperative USG. A linear incision was given 2 cm below the right middle clavicle and extending laterally up to the medial border of the shoulder joint. The fibers of the pectoralis major and deltoid attached to the inferior border of fine needle aspiration cytology clavicle were detached. A deep dissection was done in the gap between the acromion, coracoid process, and lateral end of the clavicle. A grayish pink, well-encapsulated, and circumscribed firm tumor was identified above the supraspinatus muscle in the gap. The tumor was dissected from surrounding muscles and excised in toto. The suprascapular nerve could not be identified separately. A biopsy of the lesion confirmed neurofibroma. The postoperative period was uneventful.

Tumors in Arm

Case 6 (Median Nerve Schwannoma)
A 45-year-old man presented with complaints of progressively increasing swelling in the inner aspect of the left arm for the past 5 years and associated with radiating pain to the left thumb and the index and middle fingers, and a tingling sensation of the forearm and hand. He also complained of slight weakness in the left thumb and the index finger for the past 5 years. A 5 × 4 cm swelling in the medial aspect of the left arm, approximately 5 cm from the medial epicondyle, was observed. It was tender and mobile side to side with normal overlying skin. Tinel's sign was positive along the left median nerve. Reduced sensation (90%) was present along the median nerve supply in the left hand (first three fingers). MRI showed a well-defined fusiform-shaped lesion along the median nerve, suggestive of peripheral nerve sheath tumor. Left median nerve tumor excision and reconstruction with sural nerve graft were done. Intraoperatively, the normal median nerve was identified proximal and distal to the tumor. The tumor was fusiform in shape, encompassing the whole circumference of the median nerve. It was

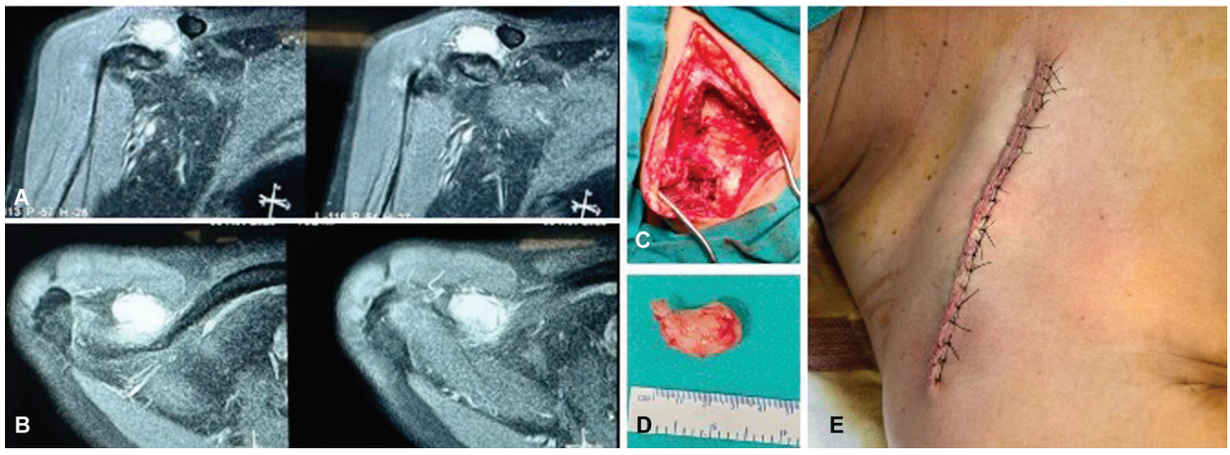

Fig. 5 Right suprascapular nerve neurofibroma. Magnetic resonance imaging of the shoulder showing an enhancing tumor along the right suprascapular nerve in (**A**) coronal and (**B**) axial views. (**C**) Intraoperative image showing a tumor arising from the right suprascapular nerve. (**D**) Excised tumor specimen. (**E**) Postoperative image.

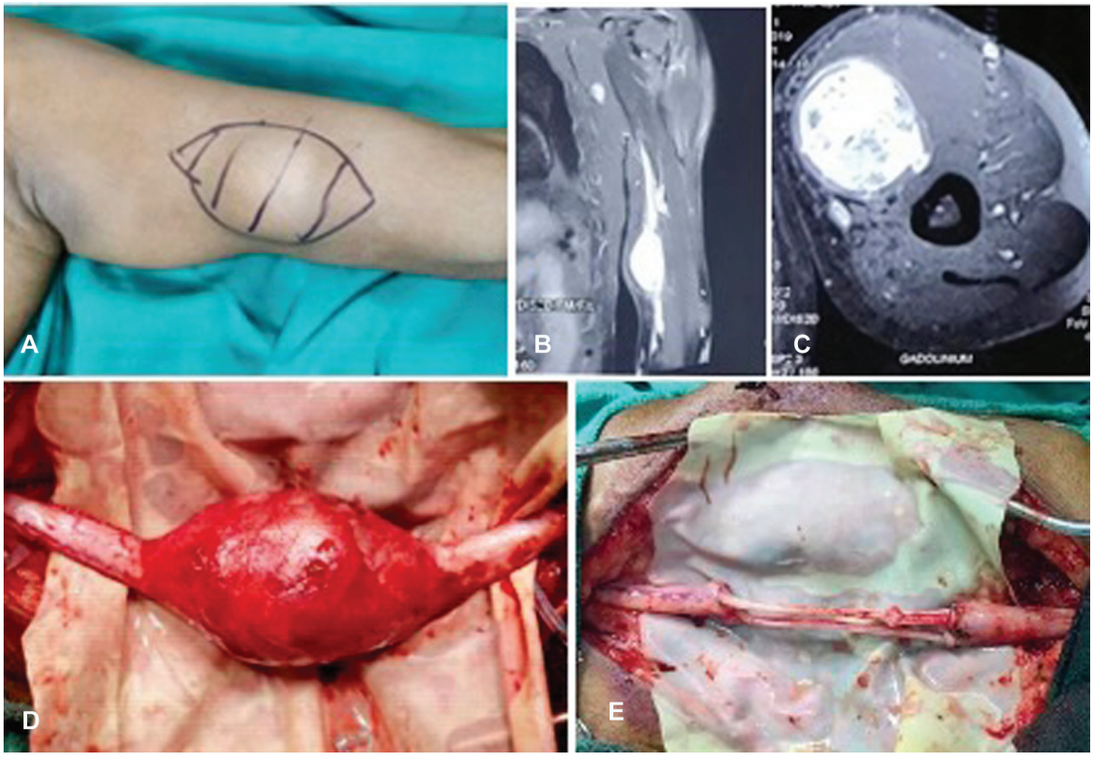

Fig. 6 Left median nerve schwannoma. (**A**) Preoperative clinical image. Magnetic resonance imaging of the left arm showing an enhancing tumor along the left median nerve in (**B**) coronal and (**C**) axial views. (**D**) Intraoperative image showing a tumor arising from the left median nerve. (**E**) Postoperative image showing reconstruction of the nerve defect with the sural nerve.

moderately vascular, firm, with good plane of demarcation from other neurovascular structures. The involved segment of the median nerve also excised. The resulting nerve defect was approximately 5 cm long. Therefore, nerve grafting was done using the left sural nerve (double cable) with Prolene 8–0. There was weakness of the left thumb and the index finger as present before the surgery. A biopsy of the lesion confirmed schwannoma.

Tumors in the Forearm

Case 7 (Ulnar Nerve Schwannoma)
A 9-year-old girl presented with a small swelling measuring 3 × 3 cm in the right medial forearm (►**Fig. 7**) and associated tingling sensation along the medial aspect of the right little finger. A T1-isointense and T2-hyperintense, well-defined lesion was seen over the extensor digitorum muscle. Exploration and total excision of the tumor were done. A biopsy of the excised mass confirmed schwannoma. The postoperative period was uneventful.

Case 8 (Unnamed Nerve Neurofibroma)
A 35-year-old man presented with a small swelling of approximately 3 × 3 cm in the radial aspect of the left elbow not easily mobile in both axes and firm in consistency (►**Fig. 8**). A T1-isointense and T2-hyperintense well-defined lesion was observed, suggestive of nerve sheath tumor. Exploration and total excision of the tumor were done. A subcutaneous dissection was done, and the extensor digitorum muscle sheath was identified and split open and the muscle fibers were dissected to find a well-defined, spindle-shaped tumor adhered to the nerve fiber. The tumor was excised. A biopsy confirmed neurofibroma. The postoperative period was uneventful.

Case 9 (Median nerve Parachordoma)
A 62-year-old woman presented in January 2017 with a painful swelling at the flexor aspect of the right forearm (►**Fig. 9**). There was a painful solid mass, 2 cm in size and restricted mobility in both transverse and longitudinal directions. Tinel's sign was present. There was no motor weakness or muscle atrophy. MRI showed an enhancing lesion below the flexor tendon of the forearm. Provisional diagnosis of a nerve sheath tumor arising from the median nerve was made. The tumor had an eccentric position and was firmly attached to the median nerve. The epineurium was longitudinally incised and the soft tumor mass was dissected from surrounding nerve fascicles and enucleated. The postoperative course was uneventful with no new neurological deficit. The histopathology confirmed parachordoma. The patient remained in yearly follow-up. About 6 years later, the patient again presented with the same site swelling in the right forearm and pain and tingling over the right hand for 6 months. A contrast-enhancing lesion in the lateral aspect in the intermuscular plane along the median nerve was seen on MRI. Tinel's sign was present along the

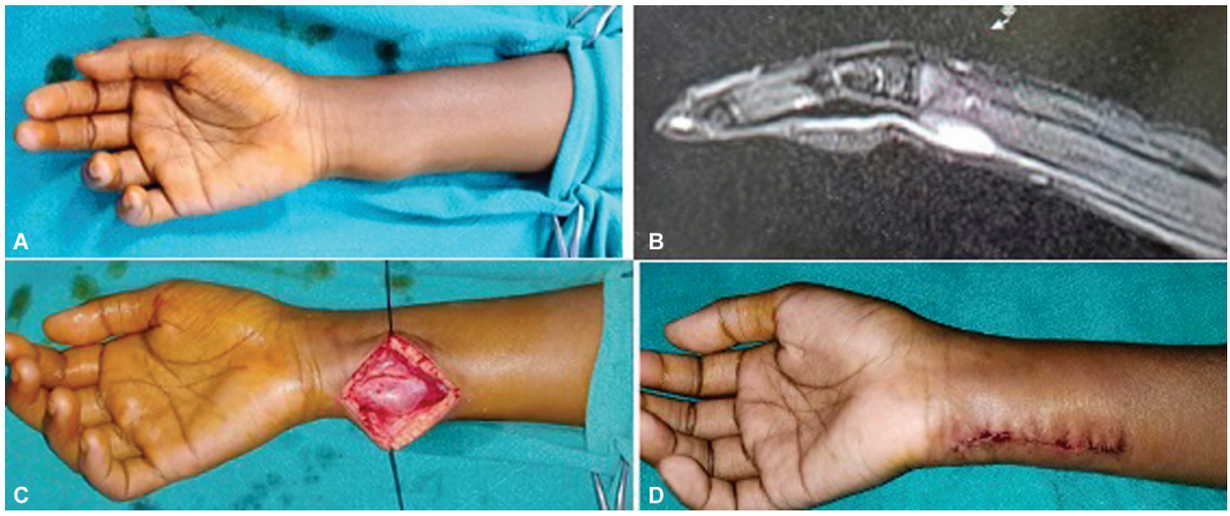

Fig. 7 Right ulnar nerve schwannoma. (**A**) Preoperative clinical image. (**B**) Magnetic resonance imaging of the right forearm showing an enhancing tumor along the right ulnar nerve. (**C**) Intraoperative image showing a tumor arising from the right ulnar nerve. (**D**) Postoperative image.

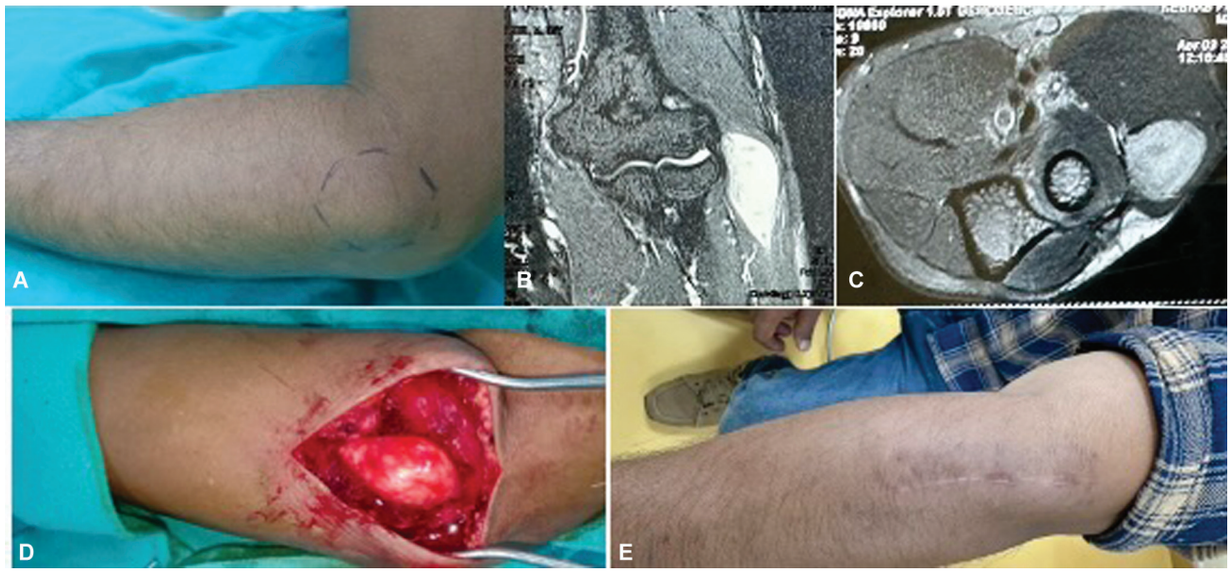

Fig. 8 Neurofibroma in the left forearm. (**A**) Preoperative clinical image. Magnetic resonance imaging of the left forearm showing an enhancing tumor in (**B**) coronal and (**C**) axial views. (**D**) Intraoperative image showing the tumor. (**E**) Postoperative image.

right median nerve. Exploration and total excision of the right median nerve tumor were done. A biopsy again confirmed parachordoma. The postoperative period was uneventful. The patient is kept in regular follow-up.

Tumors in the Wrist

Case 10 (Ulnar nerve schwannoma)

A 15-year-old adolescent boy presented with a swelling at the dorsum of the left wrist for the past 6 years (►**Fig. 10**). A 6 × 4 cm, firm, nontender, mobile swelling in the extensor aspect of the hand, 1 cm distal to the left wrist joint was observed. No other swelling was noted over the other parts of the body. FNAC of the swelling confirmed schwannoma. MRI showed a well-defined heterogenous contrast-enhancing lesion with thin septation in the deep subcutaneous plane of the dorsum of the left wrist along the tendons of the extensor digitorum longus. Excision of the tumor was done. A biopsy confirmed schwannoma. The postoperative period was uneventful.

Tumors in the Leg

Case 11 (Superficial Peroneal Nerve Schwannoma)

A 54-year-old man presented with a gradually progressive radiating pain and tingling sensation over the right leg and foot for 1 year and painful swelling in the outer aspect of the right leg for 10 months (►**Fig. 11**). A 3 × 3 cm swelling was

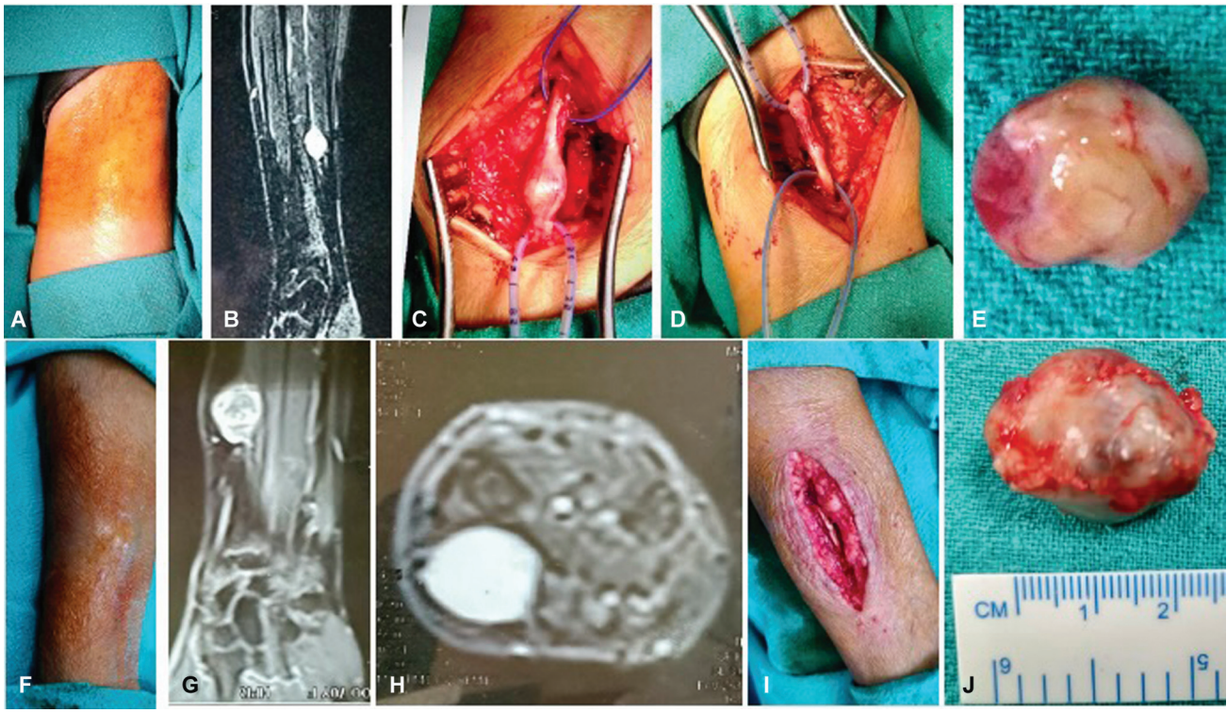

Fig. 9 Right median nerve parachordoma. (**A**) Preoperative clinical image. (**B**) Magnetic resonance imaging (MRI) of the right forearm showing an enhancing tumor in coronal view. (**C**) Intraoperative image showing a tumor along the right median nerve. (**D**) Intraoperative image after excision of the tumor. (**E**) Excised tumor specimen. (**F**) Clinical image after recurrent swelling in the right forearm. (**G**) MRI of the right forearm showing an enhancing tumor in (**G**) coronal and (**H**) axial views. (**I**) Intraoperative image after excision of the tumor. (**J**) Excised tumor specimen.

present in the lateral aspect of the middle one-third of the right leg. Overlying skin was healthy. The swelling was firm, tender, and nonpulsatile. MRI showed a well-defined, T1-isointense, T2-hyperintense lesion in the right leg antero-lateral aspect in the intermuscular plane. Exploration and enucleation of the tumor were done. The tumor was yellowish, solid, poorly vascular, and arising from the covering sheath of the superficial peroneal nerve. The upper and lower poles were dissected and the tumor was excised. The traversing nerve with splayed fibers were all preserved. A biopsy confirmed schwannoma. The postoperative period was uneventful.

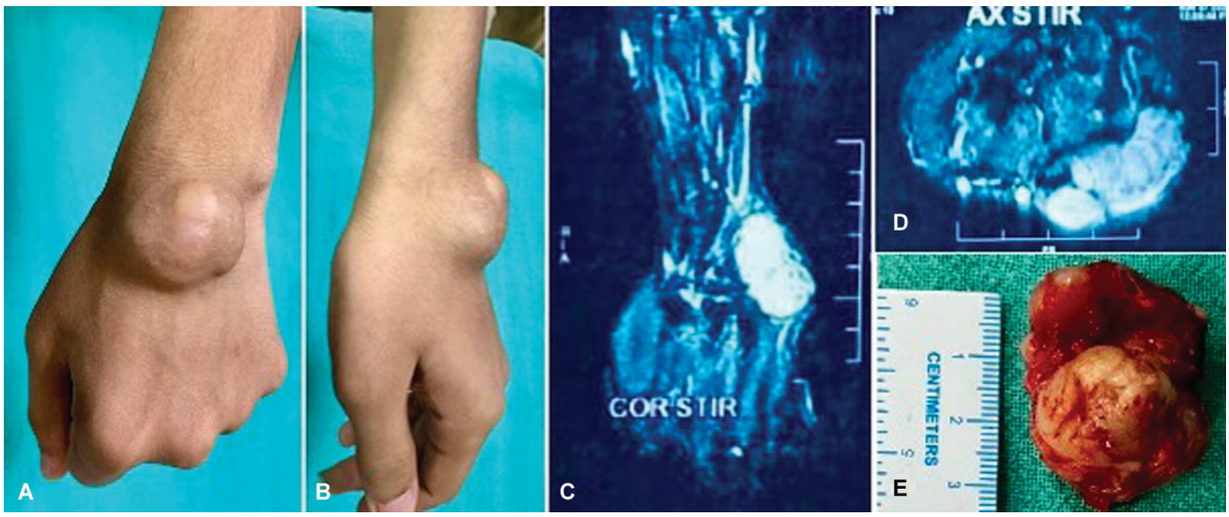

Fig. 10 Left ulnar nerve schwannoma. (**A, B**) Preoperative clinical images. Magnetic resonance imaging of the left wrist showing enhancing tumor in (**C**) coronal and (**D**) axial views. (**E**) Excised tumor specimen.

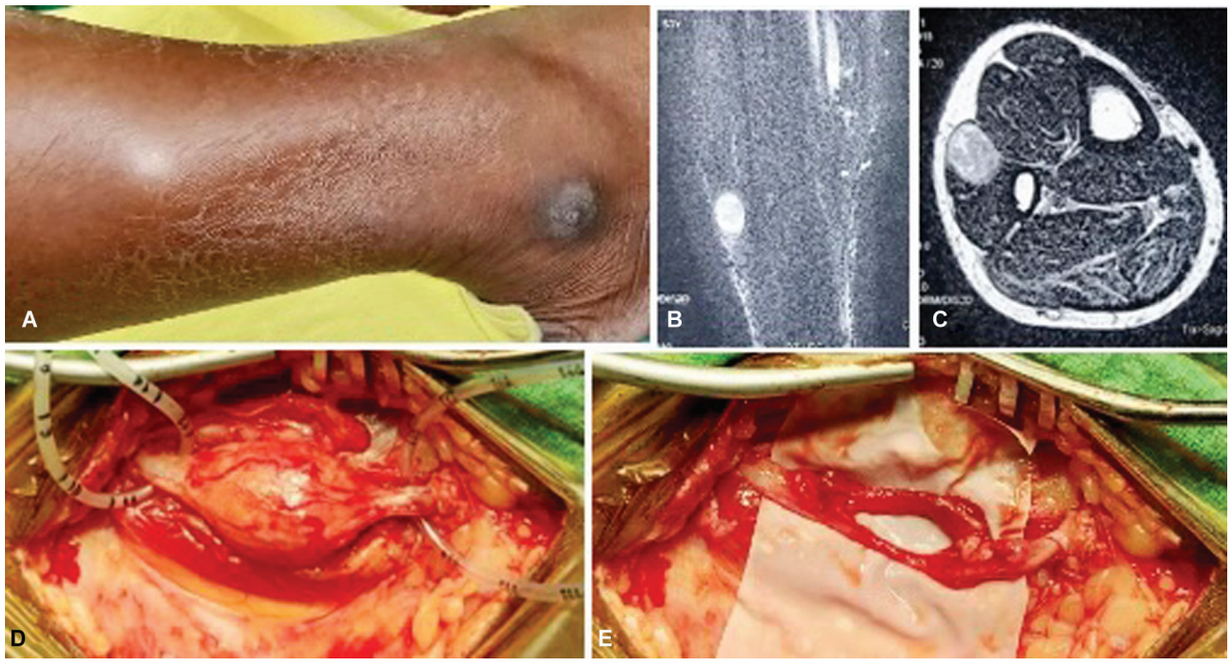

Fig. 11 Right superficial peroneal nerve schwannoma. (**A**) Preoperative clinical image. Magnetic resonance imaging of the right leg showing an enhancing tumor in (**B**) coronal and (**C**) axial views. (**D**) Intraoperative image showing the tumor. (**E**) Intraoperative image after excision of the tumor.

Tumors in the Ankle

Case 12 (Tibial nerve MPNST)

A 36-year-old woman presented with a painful lump behind the left ankle for the past 12 months (►**Fig. 12**). This was insidious and gradually progressive in size. A lump measuring 20 × 5 cm behind the left ankle, tender with overlying skin thinned out and shiny with no finger insinuation and nonpulsatile and nonreducible, was present. A slightly horizontal movement was possible, but there was no perpendicular movement along the course of the tibial nerve. MRI showed a well-defined, contrast-enhancing focal lesion measuring 20 × 4.5 × 2.4 cm at the distal course of the tibial nerve and provisional diagnosis was made as tibial MPNST. Nerve conduction velocity showed left axonal tibial neuropathy. Left foot drop was present. Exploration and gross total excision of the left posterior tibial peripheral nerve sheath tumor were done. Linear skin incision on the medial border of left tibia was made extending from the middle part of the left leg to the left medial malleolus along the course of the left posterior tibial nerve. Layered soft tissue dissection was done to expose the tumor arising from the left posterior tibial nerve. A large multilobulated, pinkish-gray, moderately vascular tumor, extending from the middle of the leg to the medial malleolus arising along the course of the left posterior tibial nerve was seen. The left posterior tibial nerve was not separately visible from the tumor mass. Gross total excision of the tumor was done. Nerve reconstruction was not done due to the large nerve defect and the possibility of a malignant disease. A biopsy of the lesion confirmed MPNST. The postoperative period was uneventful with no new deficit. The patient was attached to radiation oncology for adjuvant radiotherapy.

Discussion

Schwannomas are benign peripheral nerve tumors derived from Schwann's cells.[2] They are predominantly solitary tumors ranging from 1.5 to 3 cm in diameter. Reported incidence of schwannomas is 12 to 19% in the upper extremity and 13.5 to 17.5% in the lower extremity.[3] Schwannomas are well encapsulated and display a slow and noninfiltrating growth pattern.[4] Clinical symptoms are mainly caused by compression of nerve fascicles.[5]

Neurofibromas occur sporadically or with neurofibromatosis.[6] Malignant transformation is rare in isolated neurofibromas, but it has been reported in up to 13% cases of von Recklinghausen's disease.[7]

The definitive diagnosis is done by histopathological examination and immunohistochemistry.[8] Schwannomas are more amenable to intraneural dissection due to their encapsulated structure, whereas surgery for neurofibromas can cause structural nerve damage and require nerve reconstruction.[9] Extracapsular excision, an operative technique described by Hussain et al,[10] is commonly used for removal of schwannomas. The intracapsular technique was described by Date et al.[11] Intracapsular enucleation has been found to be superior due to lower risk of complications.[11]

Parachordomas (or myoepitheliomas) are rare soft tissue tumors with only 20 cases reported in the upper limbs to date.[12] They typically occur in the fourth decade of life, with a male predilection, and usually in extremities. There are

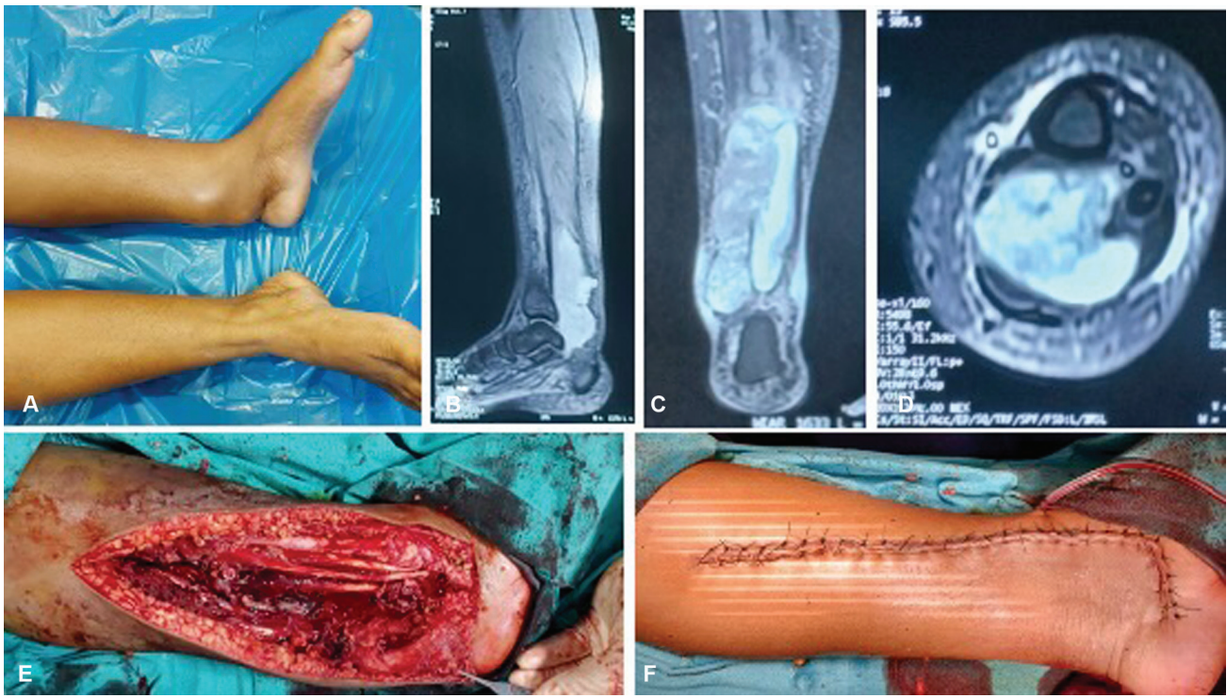

Fig. 12 Left tibial nerve malignant peripheral nerve sheath tumor. (**A**) Preoperative clinical image. Magnetic resonance imaging of the left leg showing an enhancing tumor in (**B**) sagittal, (**C**) coronal, and (**D**) axial views. (**E**) Intraoperative image after excision of the tumor. (**F**) Postoperative image.

known for late recurrence and metastasis. Originally a parachordoma was believed to be a chordoma occurring in nonaxial sites, but now a parachordoma is considered a unique entity.[13] The management for these lesions is wide surgical excision with clinical and imaging follow-up to exclude recurrence. Diagnosis is made only on histopathology.

MPNSTs constitute approximately 2% of sarcoma tumors. Half of the MPNSTs are associated with neurofibromatosis type 1 (NF1). MRI is useful to determine adjacent tissue involvement.[14] Early diagnosis and treatment offers the highest chance of survival; delay makes the outcome poor.[15] Radiotherapy may provide local control, but chemotherapy is ineffective.[16]

Conclusions

Benign tumors occur more frequently than malignant tumors in the peripheral nerves. Imaging modalities like MRI, CT angiography, and intraoperative USG are useful tools for managing peripheral nerve tumors. Electrophysiological tests and intraoperative nerve monitoring are important for diagnosis and prognosis. Histopathology confirms the diagnosis. The most suitable surgical intervention for schwannoma is complete removal with preservation of neurological function, whereas MPNSTs require a wide margin resection to obtain the most favorable prognosis.

Funding
None.

Conflict of Interest
None declared.

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