Advanced MR Imaging of Peripheral Nerve Sheath Tumors Including Diffusion Imaging

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Abstract

Keywords

► peripheral nerve sheath tumor
► magnetic resonance imaging
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► diffusion tensor imaging

Peripheral nerve sheath tumors (PNSTs) are neoplasms derived from neoplastic Schwann cells or their precursors. Whereas benign PNSTs are relatively common and considered curable lesions, their malignant counterparts are rare but highly aggressive and require early diagnosis and treatment. MR imaging has been the modality of choice for noninvasive evaluation of PNSTs. This article discusses the features of PNSTs in conventional and advanced MR imaging, and it emphasizes the features that help differentiate benign and malignant variants.

Peripheral nerve sheath tumors (PNSTs) are defined as neoplasms that clearly arise from an identifiable peripheral nerve or whose component cells show evidence of nerve sheath cell differentiation.[1] PNSTs can form in the peripheral nerve network anywhere in the body. They may arise either sporadically (with an incidence of ~ 1 per 100,000 individuals per year; they are most common in individuals 20 to 50 years of age, but without sex or racial predilection) or in relation to neurocutaneous syndromes including neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and schwannomatosis.[2,3] The major types of these neoplasms include neurofibromas, schwannomas, and perineuriomas, which are benign lesions that usually require only symptomatic treatment. Malignant PNSTs (MPNSTs) are aggressive lesions that require rapid diagnosis and treatment because of their high rate of local recurrence and distant metastasis.

MR imaging has been the modality of choice for noninvasive evaluation of PNSTs and has been used as a tool for presurgical planning and image-guided biopsies of these lesions. Moreover, MR imaging has been suggested as a screening tool in individuals with NF1 for the early diagnosis of malignant degeneration in preexisting PNSTs to avoid unnecessary biopsy.[4] This article discusses the features of PNSTs in conventional and advanced MR imaging, and it emphasizes the features that assist in differentiating benign from malignant variants.

Neurofibromas

Neurofibromas arise from Schwann cells but exhibit multiple additional cell types including neuronal axons, fibroblasts, mast cells, macrophages, perineural cells, and extracellular

matrix materials such as collagen.[5] Neurofibromas account for 5% of all benign soft tissue tumors, and they can occur both sporadically (95%) and in NF1.[6] These lesions are usually inseparable from normal nerves, and therefore complete surgical excision must include at least some part of the involved nerves.[6] Three types of neurofibromas are described.

Solitary neurofibromas (also known as localized, sporadic, or nodular) are most common (90%) and most often encountered in patients who do not have NF1. These tumors occur in young to middle-aged individuals and manifest as slow-growing masses that are discovered incidentally or present with mild to moderate sensorimotor symptoms. Solitary neurofibromas are usually cutaneous or subcutaneous, although deep-seated lesions with involvement of larger nerves also occur. They also tend to be larger, multiple, and deeper in location in the setting of NF1.[7,8]

Diffuse neurofibroma is an uncommon but distinct variety that primarily affects children and young adults. It most frequently involves the subcutaneous tissues of the head and neck.

Plexiform neurofibromas are composed of the same cell types as localized ones but have an expanded extracellular matrix. Plexiform neurofibromas may involve multiple fascicles, nerves, or even plexuses, and they grow along the nerve sheath, spreading the axons as the abnormal cells proliferate and increased extracellular matrix is deposited. Plexiform neurofibromas can arise in various regions of the body. In NF1, the most common location is in the trunk including the paraspinal region (41%), followed by the neck/upper trunk (24%), and the extremities (17%). They may be present at birth or become apparent later in life, usually preceding cutaneous neurofibromas.[9] Overall, 15 to 30% of plexiform neurofibromas are isolated to the head and neck.[6] At early stages, nerve expansion might only be caused by an increased amount of endoneurial material. Later, the lesions typically extend beyond the epineurium and spill into the surrounding soft tissues. When involving an entire limb, plexiform neurofibroma might induce *elephantiasis neuromatosa*, a condition associated with enlargement of the affected extremity, hypertrophy of bone, and redundant skin.[10] Plexiform neurofibromas are difficult to manage clinically because they can cause pain, disfigurement, and neurologic dysfunction, and can progress to malignant sarcomas.[6] Diffuse and plexiform neurofibromas are more likely to be found in NF1.[6]

Managing solitary neurofibromas depends on the symptoms. They do not typically need surgical removal, unless they are associated with pain, progressive neurologic complications, cosmetic problems, compression of adjacent tissues, and/or suspicion of malignant transformation. Plexiform neurofibromas can be difficult to resect and often recur after surgery because of residual tumor cells deep in the soft tissues. Complete removal is generally not possible without loss of neurologic function, and in some cases even subtotal removal of the tumor leads to some loss of function. For large tumors and in cases with severe pain, decompression with removal of a portion of the tumor bulk may provide benefit.[11]

On MR imaging, solitary neurofibromas appear as unencapsulated, fusiform, or round soft tissue masses that are typically < 5 cm in size and demonstrate intermediate (similar to muscle) signal intensity on T1-weighted images and heterogeneously high signal intensity on T2-weighted images. After contrast administration, small lesions feature intense and homogeneous or targetoid enhancement, whereas larger ones can show predominantly peripheral, central, or heterogeneous nodular enhancement.[6] Additional MR imaging features, which indicate the neurogenic origin of a soft tissue mass, and are common, although not pathognomonic, of solitary neurofibroma include the following.

Location in the Region of a Major Nerve/Tail Sign

1. On MR images, oriented along the long axis of a lesion, the involved nerve can be seen entering and/or exiting the neoplasm, resembling a tail coming off the tumor. Virtually pathognomonic for PNSTs, this feature is usually easy to detect in lesions affecting large deep nerves, but it is often difficult or impossible to assess in superficial or in small lesions.[4,12] This feature is seen in both benign PNSTs (BPNSTs) and MPNSTs. In most cases, PNSTs also acquire a fusiform shape that is related to the growth along the course of the affected nerve. In addition, visualization of a nerve eccentrically entering a mass must be considered a strong indicator of a schwannoma[13] (►**Fig. 1**).

Split-Fat Sign

1. Because neurovascular bundles are normally surrounded by fat, benign masses arising in relation to these structures usually maintain a complete thin rim of fat about them as they slowly enlarge and remodel the surrounding fat plane. In intramuscular PNSTs, this rim is best depicted on T1-weighted images oriented along the long axis of the lesions. The rim separates the tumor from the surrounding muscle tissue and appears more prominent at the tapering margins of the neoplasm. Known as the split-fat sign, this configuration suggests a tumor origin in the intermuscular space about the neurovascular bundle, and neurogenic neoplasms are the most frequent cause.[12] The sign is frequent in neurofibroma, but less common in schwannoma or other slow-growing lesions, and MPNSTs, although in the latter neoplasms, the rim of fat is typically incomplete reflecting their infiltrative growth pattern.[8]

Target Sign

1. The target sign appears on T2-weighted images in PNSTs and refers to central low or intermediate signal intensity, surrounded by a rim of higher signal intensity. This pattern reflects the histologic features of the tumor, with T2 hyperintense myxomatous tissue surrounding the low to intermediate signal fibrocollagenous core. Enhancement may follow a reverse or similar pattern. Rarely, a reverse pattern of target appearance on T1-weighted images is seen, with central hyperintensity and peripheral hypointensity (►**Fig. 1**). Cutaneous PNSTs are less likely than deeper lesions to demonstrate the target sign. Initially
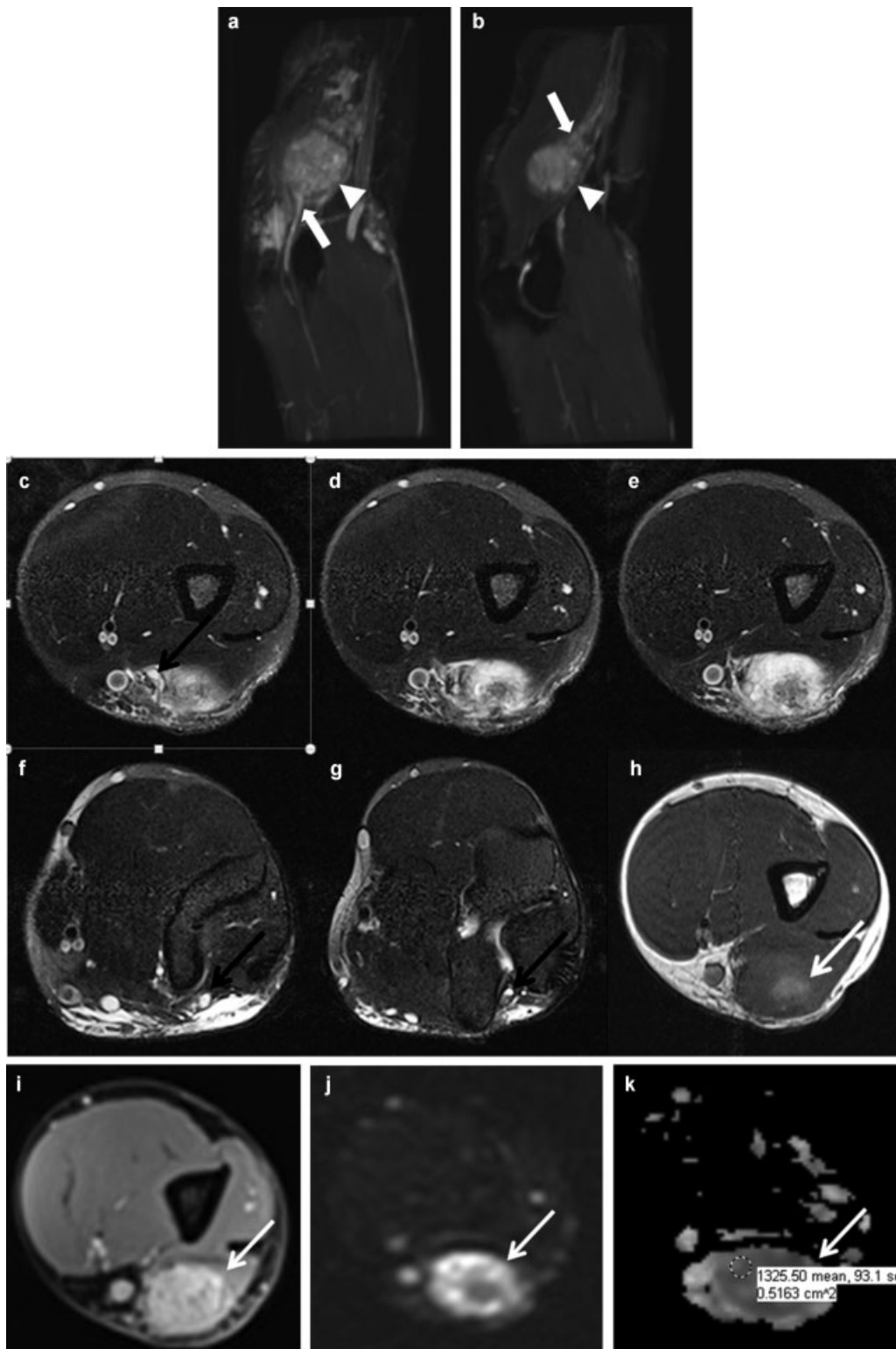
Fig. 1 Schwannoma of the ulnar nerve in a 24-year-old man. (**a, b**) Sagittal maximum intensity projection images from three-dimensional reverse fast imaging with steady-state free precession sequence at the level of the elbow demonstrate a heterogeneous hyperintense round lesion (arrowheads) at the anatomical location of the ulnar nerve. Note the involved nerve as it enters (arrow in a) and exits (arrow in b) the neoplasm, resembling a tail coming out of the tumor and creating the "tail sign." (**c–g**) Sequential axial T2 spectral attenuated inversion recovery images depict the involved fascicle (arrows) coursing within the neoplasm. (**h**) Axial T1-weighted image reveals target appearance (arrow) within the lesion. (**i**) On the respective postcontrast fat-suppressed T1-weighted image, the tumor (arrow) shows intense and slightly heterogeneous enhancement. (**j**) The neoplasm exhibits restricted diffusion on axial diffusion-weighted image with a targetoid appearance and (**k**) apparent diffusion coefficient map shows value of 1.3×10^{-3} mm^2/second.

considered pathognomonic for neurofibromas, the target sign has also been observed in schwannomas, and, rarely, in MPNSTs.[14,15] The target sign may not be appreciated because of improper window and level settings that can obscure the hypointensity in the central area of the neoplasm. The ability to detect the target sign can be improved by using wide window settings that allow better characterization of the internal architecture of the mass.[16]

Fascicular Sign

1. In the fully developed nerve, a layer of connective tissue, or an epineurium, surrounds the entire nerve trunk. Bundles of nerve fibers are surrounded by a perineurium. This gross appearance can be recognized on MR imaging and appears

as multiple ringlike prominent T2 hypointense structures within the lesion that possibly reflect the enlarged fascicular bundles seen histologically. A thin hypointense capsule is occasionally identified on T2-weighted images, particularly if the tumor is surrounded by fat. This sign is highly suggestive of PNST and slightly more common in schwannoma than in neurofibroma, but it does not appear in MPNSTs.[6,16] Similar to the detection of the target sign, detecting the fascicular sign may require wider window settings.[16]

Muscle Denervation Changes

1. PNSTs can be associated with denervation of the muscle(s) innervated by the involved nerve. Imaging features are

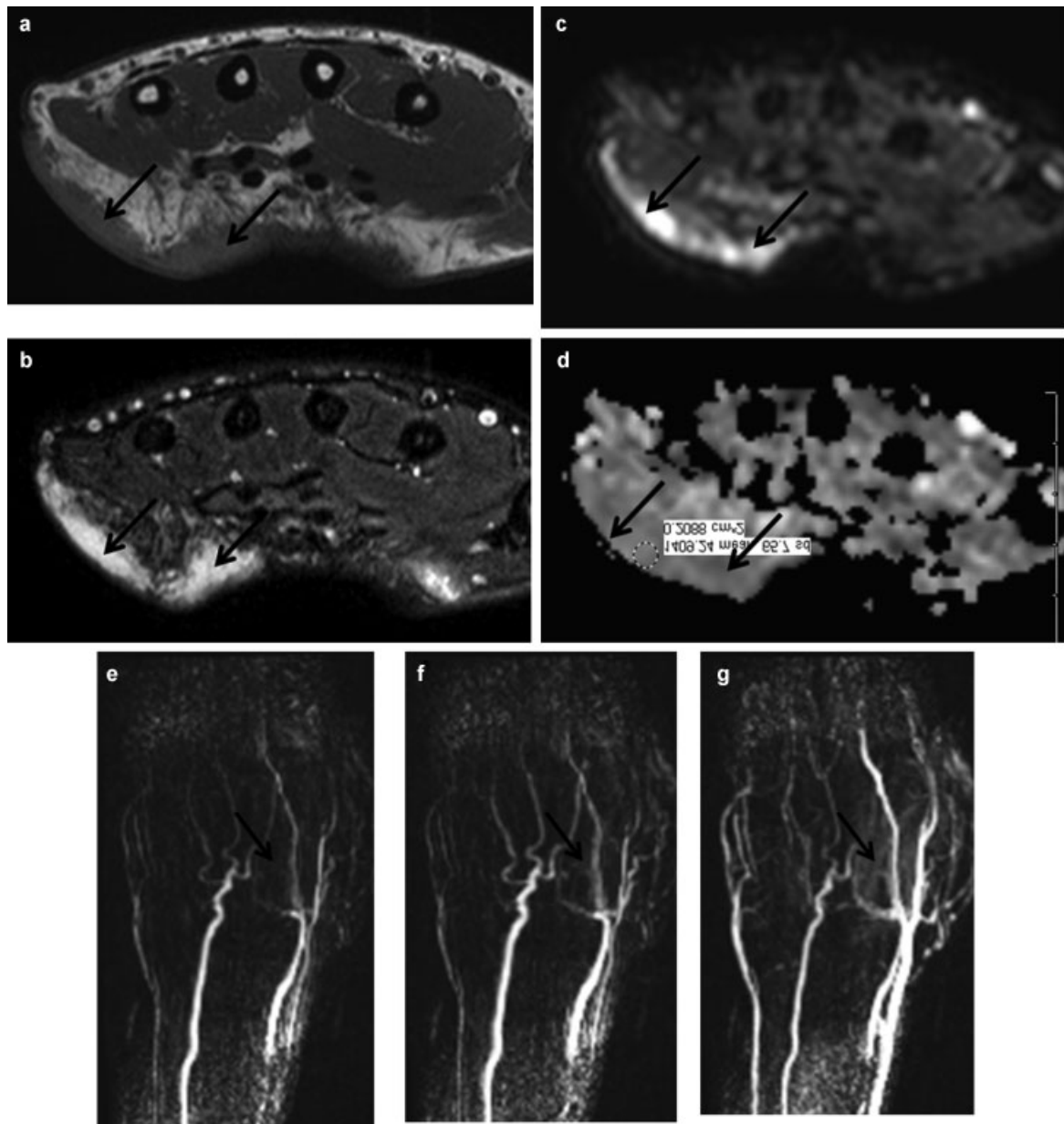**Fig. 2** Diffuse neurofibroma in a 38-year-old man. (**a**) Axial T1-weighted and (**b**) fat-suppressed T2-weighted images demonstrate an ill-defined lesion (arrows) of intermediate T1 and high T2 signal involving the subcutaneous tissues of the palmar aspect of the hand. The lesion (arrows in c–g) exhibits restricted diffusion (apparent diffusion coefficient [ADC]: 1.4×10^{-3} mm^2/second) on the respective (**c**) diffusion-weighted image and (**d**) ADC map, as well as delayed enhancement on the dynamic contrast-enhanced coronal images (**e–g**).

diffusely distributed within the affected muscle(s) and can include edema-like signal (high signal on fluid-sensitive sequences), fatty infiltration (replacement of muscle tissue by T1/T2 hyperintense fat), and/or atrophy (loss of muscle volume). Findings are occasionally subtle and can require comparison with the normal contralateral side.[12]

Diffuse neurofibromas demonstrate similar signal and enhancement characteristics as solitary ones, but they are often ill defined and spread extensively along connective tissue septa and between adipose tissue. Diffuse neurofibromas typically involve the subcutaneous tissue down to the level of the fascia[17] (►Fig. 2). Plexiform neurofibromas almost invariably show a pathognomonic MR imaging appearance that reflects their gross pathologic aspects of diffuse and multifocal nerve thickening. They appear as multinodular lesions that often feature the target sign and involve multiple nerve branches, creating a serpentine "bag-of-worms" configuration (►Fig. 3).[12] Plexiform neurofibromas can present with three types of growth patterns: *superficial, displacing,* and *invasive.* Superficial lesions are cutaneous or subcutaneous, and they show asymmetric diffuse extension with no clear demarcating borders or internal space occupation. They feature intermediate signal, are isointense to the skin on T1-weighted images,
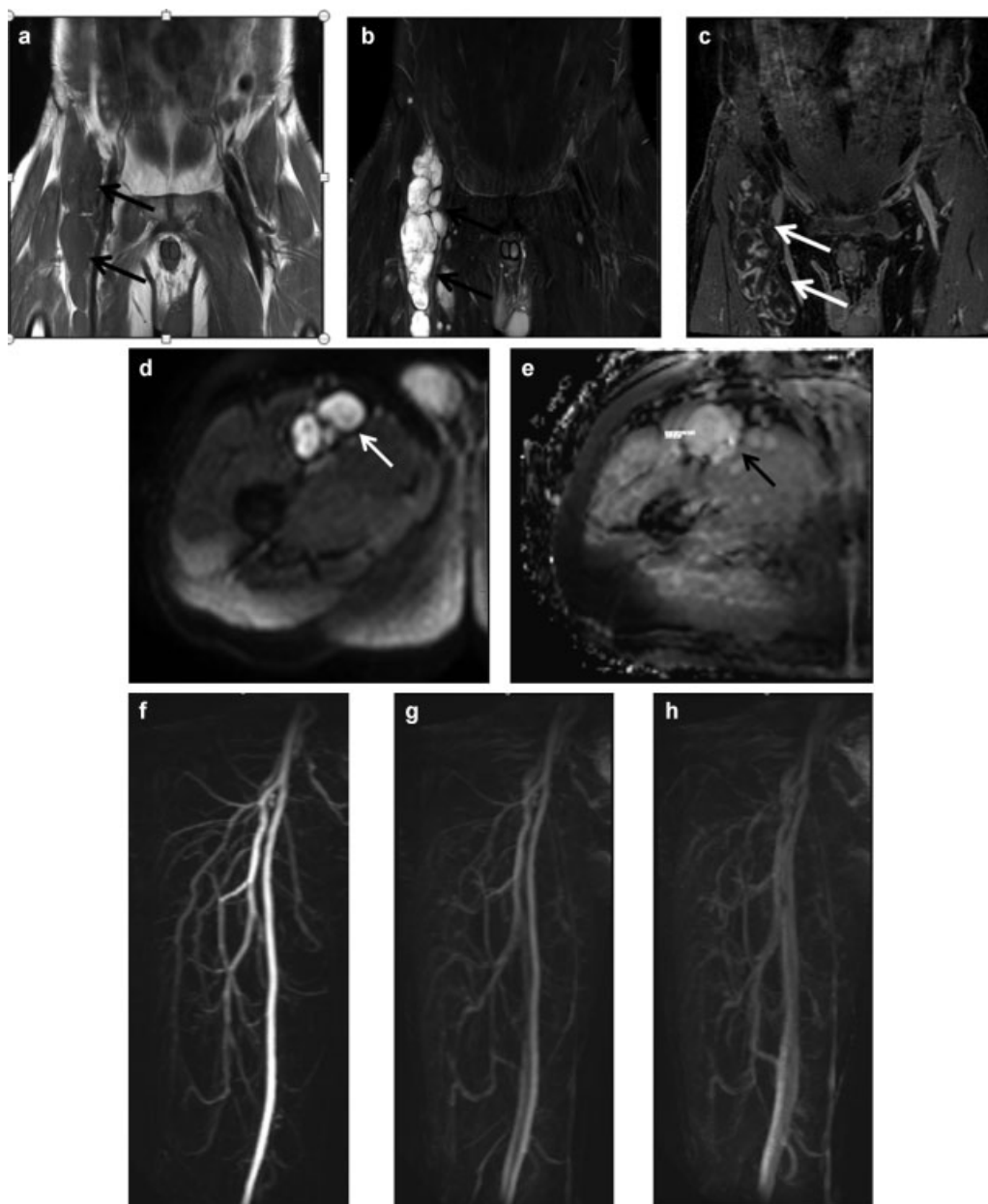

Fig. 3 Plexiform neurofibroma of the femoral nerve. (**a**) Coronal T1-weighted, (**b**) fat-suppressed T2-weighted, and (**c**) fat-suppressed postcontrast T1-weighted images demonstrate a well-defined elongated multilobulated mass (arrows) located along the anatomical course of the right femoral nerve and shows thin peripheral enhancement. The lesion exhibits restricted diffusion on the (**d**) axial diffusion-weighted image and (**e**) apparent diffusion coefficient (ADC) map with ADC 2.1×10^{-3} mm^2/second, as well as delayed enhancement on the dynamic contrast-enhanced coronal images (**f–h**).

and appear homogeneously hyperintense with good demarcation against the subcutaneous fat on fluid-sensitive images. They also show intense and homogeneous contrast enhancement. Superficial neurofibromas can mimic venous malformations on MR imaging, and MR angiography or Doppler ultrasound can be necessary for differentiation. Displacing lesions develop along main nerves and feature multinodular, smoothly defined borders, and they compress adjacent structures. They appear bright on fluid-sensitive images and feature homogeneous and moderate contrast enhancement. Invasive lesions feature ill-defined borders and appear as multiple conglomerating lesions that cannot be divided. They can penetrate muscles, fasciae, joints, and surrounding tissues. Similar to the displacing variety, invasive lesions appear bright (although slightly more inhomogeneous) on fluid-sensitive images, and they show moderate contrast enhancement. MPNSTs are associated with the displacing and invasive growth types of plexiform neurofibromas.[6,9] After surgical excision, the involved nerve(s) usually exhibit(s) T2 hyperintensity and contrast enhancement.

On advanced diffusion tensor imaging (DTI), benign neurofibromas show high minimum apparent diffusion coefficient (ADC) values ($> 1.1 \times 10^{-3}$ mm^2/second) or a lack of substantial restricted diffusion. Target appearance is also well seen in BPNSTs on DWI, whether as a solitary neurofibroma or as part of neurofibromatosis (►Fig. 4). On tractography, there are nearly normal or partially disrupted nerve tracts. The affected nerve shows increased ADC and low functional anisotropy (FA) values. Neurofibromas also tend to display delayed enhancement on dynamic contrast-enhanced MR imaging.[6,18,19]

Schwannomas

Schwannomas (also known as known as neurilemomas, neurinomas, and perineural fibroblastomas) are benign tumors composed solely of Schwann cells and ensheathed by a continuous basal lamina in association with a variable proliferation of collagen fibers. They can develop at any age, with a peak incidence in the fourth to sixth decade, but they are rare in the pediatric age group.[12] They usually present as firm, solitary, slow-growing masses that are either incidentally discovered or manifest with mild to moderate sensorimotor symptoms. They develop as multiple lesions in schwannomatosis and occasionally in NF2. They can be found in the cranial, spinal, and sympathetic nerve roots, as well as in the peripheral nerves of the flexor surfaces of the extremities.[6] Schwannomas can also be plexiform. The latter may be associated with NF2 and are usually dermal or subcutaneous, and they most often develop in the trunk or upper extremities. They may be locally aggressive and tend to recur after resection but are not considered overtly malignant.[1,6]

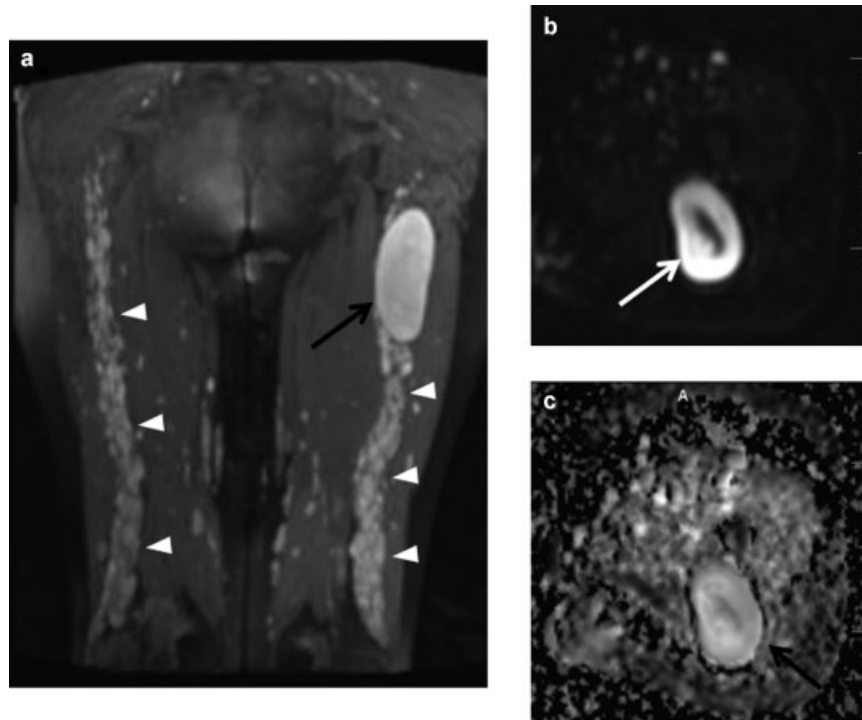Schwannomas associated with symptoms are surgically removed. Some surgeons advocate removing asymptomatic

Fig. 4 Large neurofibroma in a 38-year-old woman with neurofibromatosis type 1. (**a**) Coronal fat-suppressed T2-weighted maximum intensity projection (MIP) image demonstrates multiple scattered T2 hyperintense lesions in the thighs that correspond to neurofibromas, the vast majority of which are distributed along the anatomical course of the sciatic nerves (arrowheads). One of them (arrow), located in the proximal left thigh, is significantly larger than the others and exhibits target sign and restricted diffusion on the axial diffusion-weighted image (**b**) and apparent diffusion coefficient (ADC) map (**c**), the ADC = 2.0×10^{-3} mm^2/second. Notice on the MIP image (**a**) or narrow window settings that the target sign may not be inconspicuous.

lesions because they often will grow. Because schwannomas are eccentrically encapsulated within the perineurium, they can be surgically excised, with sparing of the affected peripheral nerve.[8] Schwannomas can recur but rarely undergo malignant degeneration.[6,12]

Schwannomas share MR imaging features with solitary neurofibromas. They are usually fusiform in shape, < 5 cm in size, and feature intermediate (similar to muscle) signal intensity on T1-weighted images and high signal intensity on T2-weighted images. On the latter images, a thin hypointense capsule (epineurium) can also be identified, particularly if the tumor is surrounded by fat.[6] Similar to neurofibromas, schwannomas commonly demonstrate the split-fat sign, fascicular sign, and target sign. The target sign is usually absent in large lesions or in schwannomas that have undergone cystic, hemorrhagic, or necrotic degeneration. The latter are known as ancient schwannomas and can mimic sarcomas on MR imaging.[20] The eccentric and separate relationship of schwannomas relative to the involved peripheral nerves is not shown confidently on conventional MR imaging to distinguish them from neurofibromas. However, dedicated MR neurography (MRN) can show the one or two prominent fascicles that are individually affected by the tumor, similar to microsurgical findings (►**Fig. 1**).[6] After surgical excision, involved nerves usually exhibit T2 hyperintensity and contrast enhancement but to a lesser degree compared with neurofibromas.

Similar to solitary neurofibromas, schwannomas demonstrate high minimum ADC values (> 1.1–1.2×10^{-3} mm^2/second) or a lack of substantial restricted diffusion on advanced DTI. Internal hemorrhagic areas in schwannomas show low ADC values, and these areas should be avoided when calculating the diffusion values by corroborating with T1-weighted images. With the exception of degenerated (ancient) lesions, schwannomas display nearly normal or partially disrupted nerve tracts. The affected nerve shows increased ADC and low FA values. Schwannomas also tend to display delayed enhancement on dynamic contrast-enhanced MR imaging.[6,18,19]

Perineurioma

Perineuriomas are rare underrecognized benign neoplasms defined as peripheral nerve sheath tumors. They are composed exclusively of neoplastic perineurial cells that demonstrate ultrastructural and immunohistochemical features similar to those of their healthy counterparts. Arranged in concentric layers around the central axon and Schwann cells, perineuriomas create an onion-bulb appearance on transverse histologic sections.[10,21] Based on the location, these lesions have been have been traditionally classified into *intraneural* and *extraneural* perineuriomas. Intraneural perineuriomas, also known as localized hypertrophic neuropathy, are composed exclusively of perineurial cells and restricted to the boundaries of a nerve. They cause localized cylindrical or fusiform enlargement of the affected nerve over a length of several centimeters to > 30 cm.[10] Extraneural perineuriomas, which are painless nodules found mainly in the soft tissues and skin, have no association with neurocutaneous syndromes, in contrast to other PNSTs.[22] They typically affect children and young adults, and they most commonly develop in the sciatic, radial, and ulnar nerves, as well as in the sacral plexus. Clinically, these tumors manifest as a slowly progressive or static mononeuropathy that includes weakness of an extremity, denervation signs on electromyography, and consequent atrophy of muscles.[10]
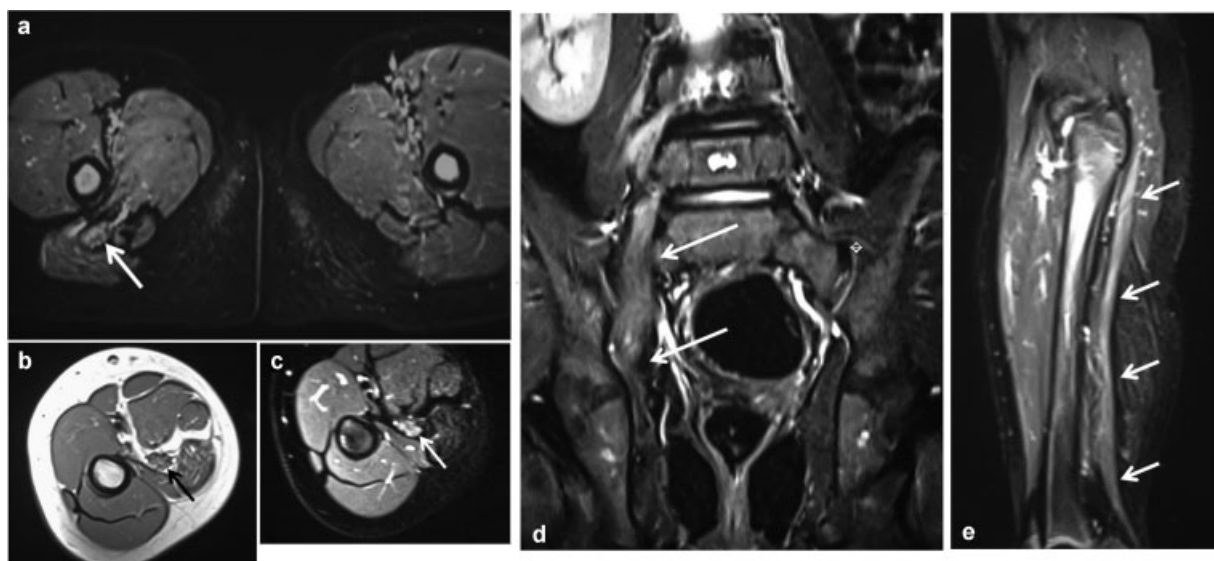

Fig. 5 Perineurioma in a young boy, who presented with painless functional loss of the right lower extremity. (**a**) Axial T2 spectral attenuated inversion recovery, (**b**) T1-weighted, and (**c**) postcontrast fat-suppressed T1-weighted images demonstrate moderate enlargement, moderate T2 hyperintensity, and intense contrast enhancement of a long nerve segment (arrows), extending from the lumbosacral plexus to the sciatic nerve. The extent of the involvement (arrows) is better defined on the respective coronal postcontrast fat-suppressed T1-weightede (**d**) and sagittal short tau inversion recovery (**e**) images (case courtesy of Dr. Jonathan Samet).

Extraneural perineuriomas follow a benign clinical course, and surgical resection with margins free of neoplasm is typically curative. Intraneural perineuriomas, in contrast, have a poorer prognosis. Treatment of the latter lesions is controversial, with some authors advocating diagnostic biopsy followed by neurolysis instead of resection and others preferring resection with neural grafting or end-to-end anastomosis, based on the concept that these neoplasms represent a progressive condition that evolves inexorably to a total loss of nerve function.[6,22] However, most of these lesions do poorly, despite extensive surgery.

On MR imaging, the affected nerve features low to intermediate signal intensity on T1-weighted images, heterogeneous high signal intensity on T2-weighted images, as well as avid contrast enhancement. There is gradual increase of the caliber of the nerve proximally, followed by gradual taper distally.[23] On axial and longitudinal MRN images, the fascicular architecture is typically maintained (►Fig. 5), with the individual fascicles exhibiting uniform enlargement, resulting in a honeycomb appearance.[24] Common associated find-ings include denervation edema, atrophy, and fatty degeneration of dependent muscle groups.[10] The affected nerve shows increased ADC and low FA values. The ADC value of the lesion can approach $1.0–1.1 \times 10^{-3}$ mm^2/second, probably reflecting a more organized lesion in a young compact nerve. Although the imaging appearance can mimic other neurogenic benign neoplasms, a diagnosis is suggested by the combination of slowly progressive mononeuropathy, the patient's young age, and a lack of known tumor syndrome. Because lesions are static or slowly progressive, they can be followed clinically and with imaging, as indicated. Treatment of choice is resection of the neoplasm with end-to-end nerve grafting. However, few patients do well, despite extensive surgery.[6]

Malignant Peripheral Nerve Sheath Tumors

The generic term MPNST is used to describe a confusing multitude of names including malignant schwannomas, malignant neurilemomas, nerve sheath fibrosarcomas, and
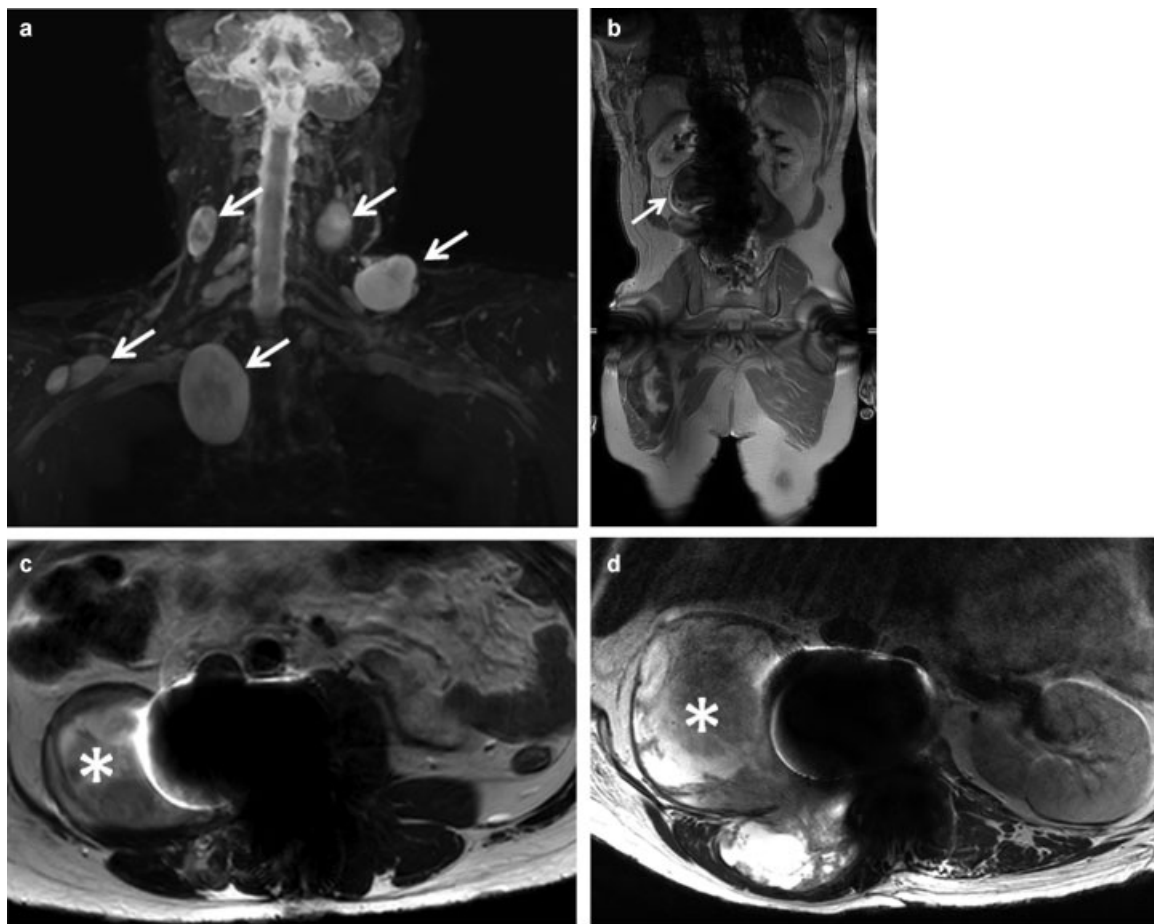

Fig. 6 Malignant peripheral nerve sheath tumor in a 34-year-old man with neurofibromatosis type 1. (a) Coronal three-dimensional short tau inversion recovery image from a whole-body MRI scan reveals multiple ovoid homogeneously T2 hyperintense lesions (arrows) in the neck that are compatible with neurofibromas. (b) Postcontrast T1 mDixon image from the same examination demonstrates a right paraspinal lesion (arrow) that features cystic components, and another lesion in the ipsilateral thigh (arrowhead) that shows targetoid enhancement. On diffusion-weighted imaging, the lesions exhibited apparent diffusion coefficient values of 0.7 and 2.4×10^{-3} mm^2/second, respectively. On biopsy, the paraspinal lesion proved to be a high-grade malignant peripheral nerve sheath tumor. On axial T2 Dixon images at baseline (c) and after 3-month follow-up (d), the malignant lesion (asterisk) exhibits considerable growth in size.

neurogenic sarcomas (neurofibrosarcomas). MPNSTs are spindle cell sarcomas that arise from peripheral nerves or neurofibromas or show nerve tissue differentiation. Histology shows a cellular component resembling fibrosarcoma together with areas of myxoid, hemorrhage, and necrosis.[10] MPNSTs account for 3 to 10% of all soft tissue sarcomas, with 15 to 70% occurring in patients with NF1.[8,25] In addition, ~ 3 to 13% of patients with NF1 develop MPNST, usually after a long latent period of 10 to 20 years.[26,27] MPNSTs usually occur in individuals between 20 and 50 years of age, without gender predilection, and most commonly involve large peripheral nerves including the sciatic nerve, brachial plexus, and lumbosacral plexus. The average age of patients with NF1 who develop an MPNST is 30 years.[10] MPNSTs can also be secondary neoplasms related to previous radiation therapy. Such tumors develop after a latent period (10 to 20 years) and account for 11% of MPNSTs.[6] Clinically, MPNSTs usually present with a new onset of sensory and/or motor deficit, new or intensified pain, and/or as rapid enlargement of a known PNST.[6]

MPNSTs are a leading cause of death in patients with NF1. They respond poorly to chemotherapy and radiation, and surgical ablation with wide resection margins is the only effective therapeutic option.[28] Despite aggressive treatment, however, local recurrence and distant metastases are common. Metastases most frequently affect the lung, bone, pleura, and retroperitoneum, with regional lymph nodes involved in 9% of cases.[12] Mortality rates of 61% are reported,[21] with patients surviving an average of 25 months before succumbing to the disease.[11]

On conventional MR imaging, MPNSTs tend to display irregular shape and indistinct margins. On T2-weighted images, they appear heterogeneous and feature high signal intensity, whereas on T1-weighted images they are usually isointense to muscles, although in some cases they feature areas of T1 hyperintensity that correspond to hemorrhage and are strongly indicative of the diagnosis. Additional features, which are also considered highly indicative of MPNST, include large size (> 5 cm), peripheral contrast enhancement pattern, perilesional edema, and intratumoral cystic changes.[19,29] MPNSTs typically lack the target sign and fascicular sign (►Fig. 6). One should be careful to call a known neurofibroma benign or, in the setting of known NF1, if there is a heterogeneous lesion with perilesional edema. Although the same is not true for schwannomas, ancient schwannomas are typically heterogeneous with cystic-hemorrhagic changes.

On advanced DTI, MPNSTs show low minimum ADC values (< 1.0–1.1×10^{-3} mm^2/second) or substantial restricted diffusion. Limited series have shown that MPNSTs also demonstrate partial or complete nerve tract disruption. On dynamic contrast-enhanced MR imaging, MPNSTs tend to display early arterial enhancement, a feature quite rare in BPNSTs.[6,18,19] In a recent study that used MR spectroscopy to differentiate between BPNSTs and MPNSTs, trimethylamine concentrations and the trimethylamine fraction were found to be relatively lower in the benign neoplasms. A trimethylamine fraction threshold of 50% resulted in 100% sensitivity and 72.2% specificity in distinguishing BPNSTs from MPNSTs.[30]

Normal and Abnormal Postoperative Changes

Postoperatively, the operated nerve can show mild increased signal as a normal variant. Mild prominence of fascicles and small residual lesion in the setting of operated PNST is not uncommon, due to difficulty in resecting the infiltrating lesion and/or additional trauma to the offending nerve. Such changes are more common with larger lesions, particularly neurofibromas as compared with schwannomas (►Figs. 7 and 8). Recurrent mass is indicated by the appearance of a new nodular or enhancing lesion or a lesion with restricted diffusion. Other postoperative complications include perineural hematoma and scarring.

Peripheral Nerve Sheath Tumor Mimics

Entities that can mimic PNSTs on conventional MR imaging and MRN include posttraumatic neuroma, peripheral nerve lipomatosis, Charcot-Marie-Tooth (CMT) disease, amyloidosis, synovial or fibrosarcoma, and intraneural metastatic disease. Posttraumatic neuroma is associated with a history
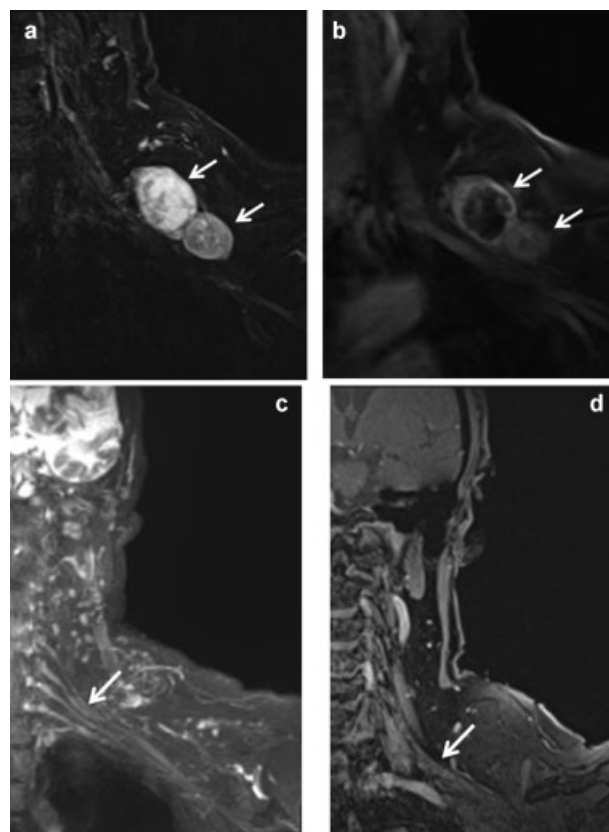

Fig. 7 Normal postoperative findings after surgical excision of a schwannoma in a 65-year-old woman. (**a**) Coronal three-dimensional short tau inversion recovery and (**b**) postcontrast fat-suppressed T1-weighted images from a presurgical scan demonstrate a well-defined bilobed lesion (arrow) involving a left brachial plexus division. The respective images (**c, d**) from a postsurgical scan exhibit a minimal degree of T2 hyperintensity (arrows in c) and contrast enhancement (arrows in d) of the involved nerve branches, the expected postoperative appearance.
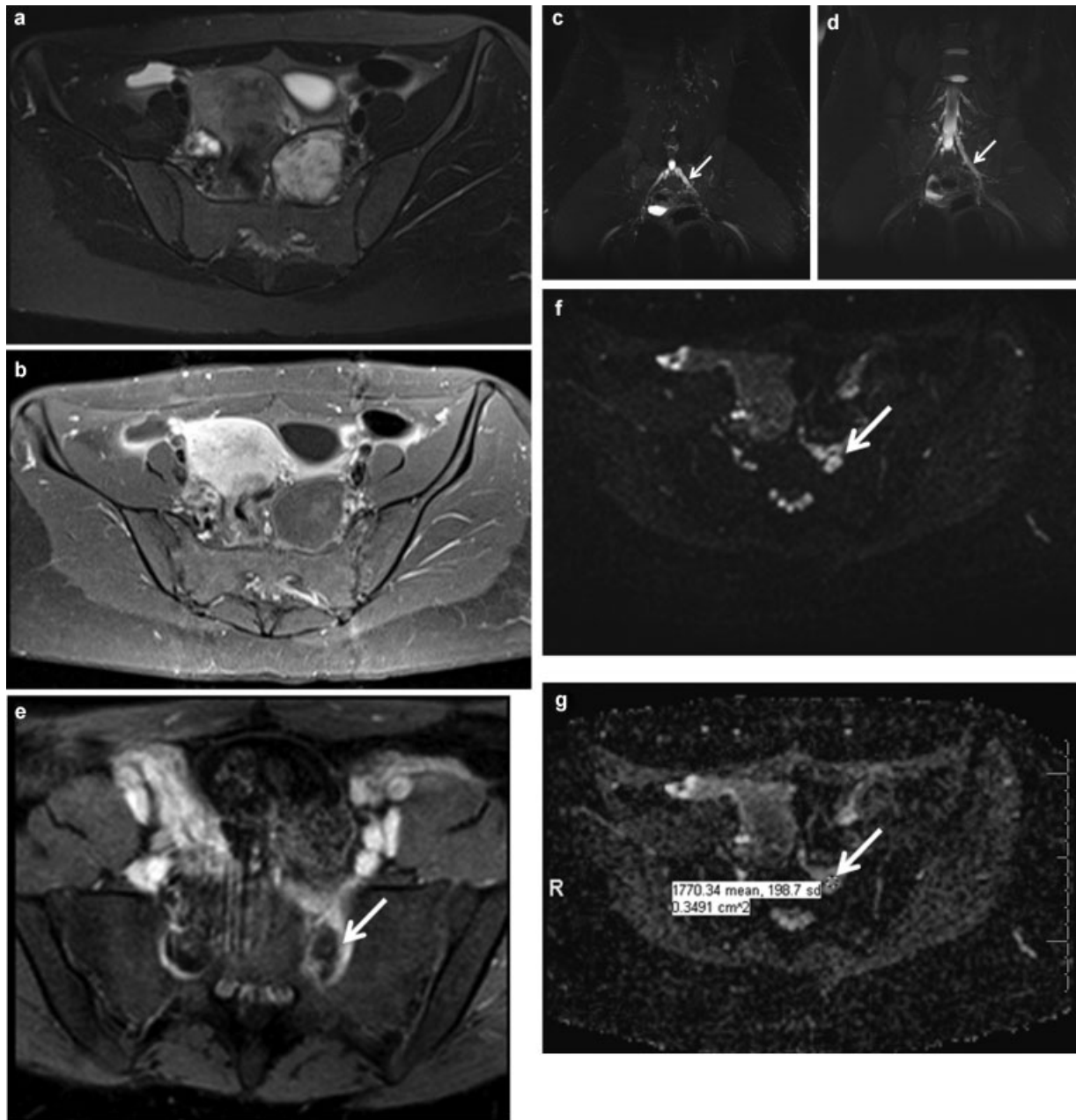
Fig. 8 Normal postoperative findings after surgical excision of a neurofibroma in a 65-year-old woman. (**a**) Axial T2 spectral attenuated inversion recovery and (**b**) postcontrast fat-suppressed (fs) T1-weighted images from a presurgical scan demonstrate a well-defined ovoid lesion (arrow) in the presacral space, compressing and posteriorly displacing the left S1 nerve (arrow). On surgery excision, the lesion proved to be a grade I neurofibroma involving the left S1 and S2 nerve roots. (**c, d**) The respective three-dimensional inversion recovery turbo spin-echo images from a postsurgical scan exhibit a moderate degree of T2 hyperintensity and enlargement of the left S1 (arrow in c) and S2 (arrow in d) nerves. (**e**) Postcontrast fsT1-weighted image shows mildly enhancing perineural fibrosis around the left S1 nerve (arrow). (**f**) Diffusion tensor imaging and (**g**) apparent diffusion coefficient (ADC) images show mild diffusion restriction (ADC: 1.7×10^{-3} mm^2/second), expected postoperatively. No recurrent mass was seen.

of injury or amputation, and these show perineural scarring, lack significant contrast enhancement, and show no split-target sign. Peripheral nerve lipoma (neural fibrolipoma) features enlarged fascicles and extensive areas of intraneural fibrofatty proliferative tissue that create a typical spaghetti-like configuration on long-axis images and coaxial-cable appearance on axial images. CMT, a hereditary neuropathy, can mimic neurocutaneous syndromes on MR imaging. The entity is associated with

positive family history in 80% of cases and appears as mass-like symmetrical enlargement of the peripheral nerves and/or cauda equina. Pseudomasses from excessive demyelination and remyelination can be seen in CMT type 1A in the lumbosacral plexus as diffuse, bland-looking T2 hyperintense lesions. In amyloid neuropathy, MRN depicts unilateral or bilateral, focal (amyloidoma) or diffuse enlargement of lumbosacral plexus segments or sciatic nerves, with the involved nerve branches featuring

Table 1 Features of solitary benign and malignant peripheral nerve sheath tumors on conventional MR imaging

Images	BPNSTs	MPNSTs
T1 signal characteristics	Isointense to muscle, homogeneous	Isointense to muscle (homogeneous or heterogeneous)
T2 signal characteristics	Hyperintense • Target sign (common in schwannomas, uncommon in neurofibromas) • Fascicular sign (common in neurofibromas, uncommon in schwannomas)	Hyperintense (heterogeneous) • No target sign • No fascicular sign
Static enhancement pattern	• Present (homogeneous or targetoid, and occasionally heterogeneous in schwannomas) • Usually delayed	• Marked and heterogeneous, lack of targetoid enhancement • Usually early arterial and peripheral
Shape/size/morphology	Round or oval, usually < 5 cm • Neurofibromas: Continuity with nerve (tail sign) and/or multiple fascicles, encapsulated, can be centrally located • Schwannomas: Continuity with nerve (tail sign) and/or 1–2 fascicles, unencapsulated, can be eccentrically located	Irregular or round, usually > 5 cm
Margin definition	Well defined	Well or ill defined
Invasion	Absent	Can be present
Necrosis/cystic change	Uncommon	Common
Hemorrhage	Uncommon, usually in schwannoma	More common
Calcification	Uncommon, usually in schwannoma	Can be present with necrosis
Perilesional edema	Absent	Common

Abbreviations: BPNST, benign peripheral nerve sheath tumor; MPNST, malignant peripheral nerve sheath tumor.

Table 2 Features of solitary benign and malignant peripheral nerve sheath tumors on advanced MR imaging

Images	BPNSTs	MPNSTs
Dynamic enhancement pattern	Usually delayed	Usually early arterial
Diffusion tensor imaging	• Nerve tracts near-normal or partially disrupted • Minimum ADC values usually $> 1.0–1.1 \times 10^{-3}$	• Nerve tracts partially or completely disrupted • Minimum ADC values usually $< 1.0–1.2 \times 10^{-3}$
Spectroscopy	Trimethylamine fraction usually < 50%	Trimethylamine fraction usually > 50%

Abbreviations: ADC, apparent diffusion coefficient; BPNST, benign peripheral nerve sheath tumor; MPNST, malignant peripheral nerve sheath tumor.

prominent or disrupted fascicles, and occasionally T2 hypointense foci. However, the imaging features are often nonspecific, and clinical findings of a chronic condition such as multiple myeloma is required. Biopsy is required for a definitive diagnosis. Synovial sarcoma and fibrosarcoma are rare malignant masses of the nerve and seen as heterogeneous large hemorrhagic masses. Intraneural metastatic disease and lymphoma are extremely rare. Metastasis most commonly involves the brachial plexus in patients with breast or lung cancer. Neural lymphoma usually occurs in patients with known primary or secondary lymphoma, usually B-cell lymphoma. Multiplicity of lesions and evidence of metastatic deposits or lymphoma elsewhere in the body suggests the diagnosis.[31–33] These lesions show markedly restricted diffusion and early arterial contrast enhancement (►Tables 1 and 2).

Conclusion

MR imaging remains the modality of choice for evaluating suspected neurogenic tumors because it can establish a nerve–tumor relationship and therefore exclude other diagnoses, as well as define the size of the tumor and its association with adjacent structures, which are important for presurgical planning.[11] Despite advances in MR imaging, which have increased the accuracy in characterizing PNSTs, there are still no clear imaging criteria to distinguish between neurofibromas and schwannomas. With respect to differentiating BPNSTs and MPNSTs, it is suggested that, if the characteristic findings described for BPSNT are not present, then a MPNST or other soft tissue neoplasm cannot be excluded, and a carefully planned biopsy should be obtained.[17]

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