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MRI neurography and diffusion tensor imaging of a sciatic perineuroma in a child

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Abstract Perineuroma, rare in children, presents as a painless mononeuropathy of a major nerve trunk. Resection of the lesion with end-to-end sural nerve grafting appears to be the treatment of choice. This technique is not recommended if the unhealthy segment of nerve is too long or if spinal roots are involved. However, in children, reports of direct MR evaluation of nerve trunks and of the exiting nerve roots are limited. We report a 7-year-old girl with an intramural sciatic nerve perineuroma in whom the diagnosis was made by MRI and confirmed by biopsy. The MR protocol combining 3-D T2-W STIR SPACE, fat-saturated gadolinium-enhanced T1-W images, and diffusion tensor imaging with tractography was a valuable tool for depicting peripheral nerve and roots in order to plan surgical treatment.

Keywords Perineuroma · MR neurography · Diffusion tensor imaging · Child

Introduction

Perineuroma, previously known as localized hypertrophic neuropathy, is rare in children and presents as a painless mononeuropathy of a major nerve trunk. This definition should be reserved for cases with immunohistochemical confirmation of perineural cell lineage by demonstrating positive staining with epithelial membrane antigen (EMA) and negative staining with S100 and/or a deletion in chromosome 22 [1, 2].

Advances in MR imaging have improved the visualization of both normal and pathological peripheral nerves. However, MR neurography remains complementary to clinical examination and electrodiagnostic studies when evaluating peripheral nerve disorders [3]. In children the latter is not always performed due to poor patient cooperation. Furthermore, direct MRI evaluation of the exiting nerve roots in children is difficult [4]. Information regarding spinal nerve integrity is, however, of great value to the surgeon in selecting the most appropriate treatment. Resection of the lesion with end-to-end and sural nerve grafting is the treatment of choice, but is not recommended if spinal roots are involved or if the unhealthy nerve segment is too long [5].

We report a 7-year-old child in whom the diagnosis of intramural sciatic nerve perineuroma was made by MRI and confirmed by biopsy. We describe an MRI protocol combining three-dimensional (3-D) T2-weighted (T2-W) STIR SPACE, fat-saturated gadolinium-enhanced T1-W images, and diffusion tensor imaging (DTI) with tractography, and emphasize the diagnostic value of this technique

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not only in depicting fusiform nerve hypertrophy and the extent of the lesion in relation to the nerve trunk, but also in evaluating the involvement of spinal roots so that the most suitable surgical treatment (sural nerve graft vs. neurolysis) can be planned.

Case report

A 7-year-old girl presented with a 2-year history of progressive weakness of the left leg with painless atrophy. There was weakness of the tibialis anterior, peroneus tertius, extensor hallucis longus and extensor digitorum longus, but no sensory deficit. The patient did not meet the criteria for neurofibromatosis and there was no family history of other heritable neuropathies. Electrodiagnostic studies were not performed due to poor patient cooperation.

On a 1.5-T Avanto MR scanner (Siemens, Erlangen, Germany), we examined the sciatic nerves from the sacral plexus (L4–L5) to the knees. The following sequences were obtained:

- Axial turbo spin-echo (TSE) T2-W (TR/TE 3,100/88 ms, flip angle 150°, TI 0, voxel size 1×1×7 mm, FOV 250×176 mm).
- Coronal 3-D T2-W SPACE (sampling perfection with application optimized contrast using different flip angle evolutions) with short tau inversion recovery (STIR) (TR/TE/TI 2,000/174/160 ms, turbo factor 85, TA 6–7 min, FOV 448×448 mm, matrix size 448×482, number of slices 104, iPAT factor 3) with maximum intensity projection (MIP) and multiplanar reconstruction.
- Axial diffusion-weighted images (DWI) (TR/TE 7,100/92 ms, iPAT factor 2, NEX 1, FOV 256×256 mm, matrix 128×128, voxel size 3×3×5 mm, b values 0 and 900 s/mm²) in 30 diffusion-encoding directions). Fractional anisotropy (FA), apparent diffusion coefficient (ADC) maps and tensor calculation were performed. The 3-D dataset was analyzed using Neuro 3D Syngo software (Siemens, Erlangen, Germany). Colour-coded FA maps were calculated from the DTI dataset and fibre tracks were generated and fused with the 3-D STIR image dataset. Sciatic nerve fibre tracks were calculated using a four-order Runge-Kutta algorithm from several seed points placed in the nerve roots.
- Fat-saturated T1-W (TR/TE 507/11 ms, flip angle 126°, voxel size 1.2×1.2×3 mm) gadolinium-enhanced sequences in the axial, coronal and sagittal planes.

The 3-D T2-W STIR SPACE sequence depicted hyperintensity within an enlarged left sciatic nerve of linear shape. The gadolinium-enhanced T1-W sequence with fat saturation demonstrated enhancement within the same long portion of the hypertrophied nerve. DTI allowed superb

depiction of the sciatic nerve, well differentiated from the adjacent vessels, and sacral root involvement. It showed a decreased FA value (0.32 left, 0.38 right) and increased ADC (1.8×10^{-3} mm²/s) as compared with the contralateral side (1.6×10^{-3} mm²/s) (Fig. 1), as measured in the proximal, middle and distal parts of the lesion.

The patient underwent biopsy of the posterior tibial branch of the left sciatic nerve. Histology of the enlarged extremity showed expanded nerve fascicles containing multiple whorls of cells around nerve fibres (“pseudo-onion bulbs”) with nearly complete loss of myelin. Immunostaining of these circumferentially arranged cells showed EMA positivity, confirming their perineural nature. The cells were negative for S-protein, arguing against a Schwann cell origin (Fig. 2). Thus the lesion was considered an intra-neural perineuroma.

Discussion

We identified 53 definite cases of intraneural perineuroma from 29 articles in the literature [4]. Up to 44% of patients were children and the sciatic nerve was involved in 13% of the cases. Few authors have described the use of MRI in “localized hypertrophic neuropathy” due to perineural cell proliferation (EMA positive, S-100 negative) [6]. The differential diagnosis is chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), which is often symmetrical and multifocal; Guillain-Barré syndrome, where imaging is usually normal; and HMSN (hereditary motor and sensory neuropathy) III (Dejerine-Sottas disease), in which contrast enhancement is almost always absent. The linear, nonfusiform morphology of a solitary lesion distinguishes perineuroma from other peripheral nerve tumours such as posttraumatic neuroma, neurofibroma and schwannoma.

Fat-saturated T1-W contrast-enhanced images are most helpful in demonstrating the extension of the lesion, but the vessels are very difficult to separate from enhancing nerve fibres. Saguintaah et al. [7] described a lesion that was misinterpreted as a vascular lesion on T1-W contrast-enhanced images. Visualization of nerve roots is difficult with thick slices and consequently low-quality multiplanar reconstructed and MIP images.

Affected nerves are of higher signal than normal on T2-W images [6]. The 3-D T2-W STIR SPACE sequence combines the properties of fat-saturated T2-W images with better visualization of the spatial extension of the lesion within and along the nerve tracts due to large anatomical coverage and the ability to slice through the volume of interest. It also provides an appropriate 3-D background for anatomical fusion with fibre tracts of the complete nerves. Heavily saturated T2-W sequences distinguish them from the surrounding soft tissue but, again, not from vessels.

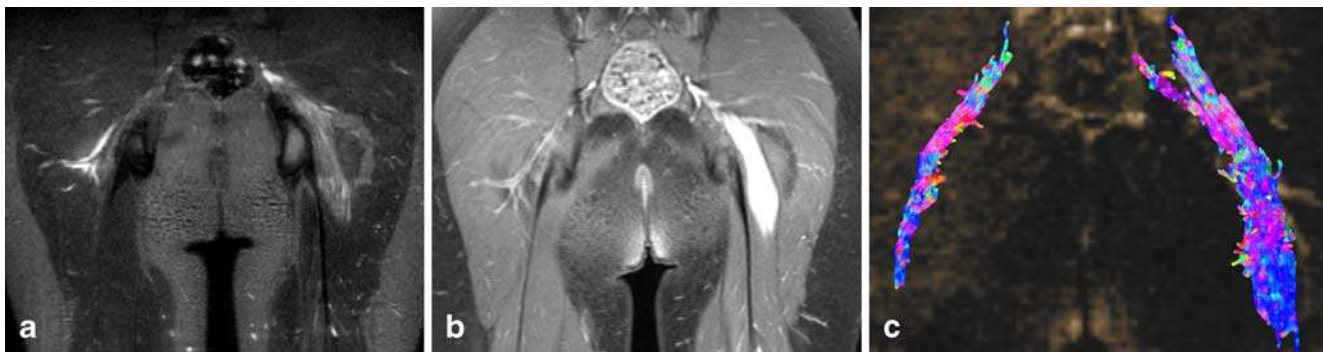


Fig. 1 MRI. **a** 3-D T2-W STIR SPACE image demonstrates hyperintensity within an enlarged left sciatic nerve. **b** Coronal fat-saturated gadolinium-enhanced T1-W image demonstrates enhancement within the same long portion of the hypertrophied nerve. However,

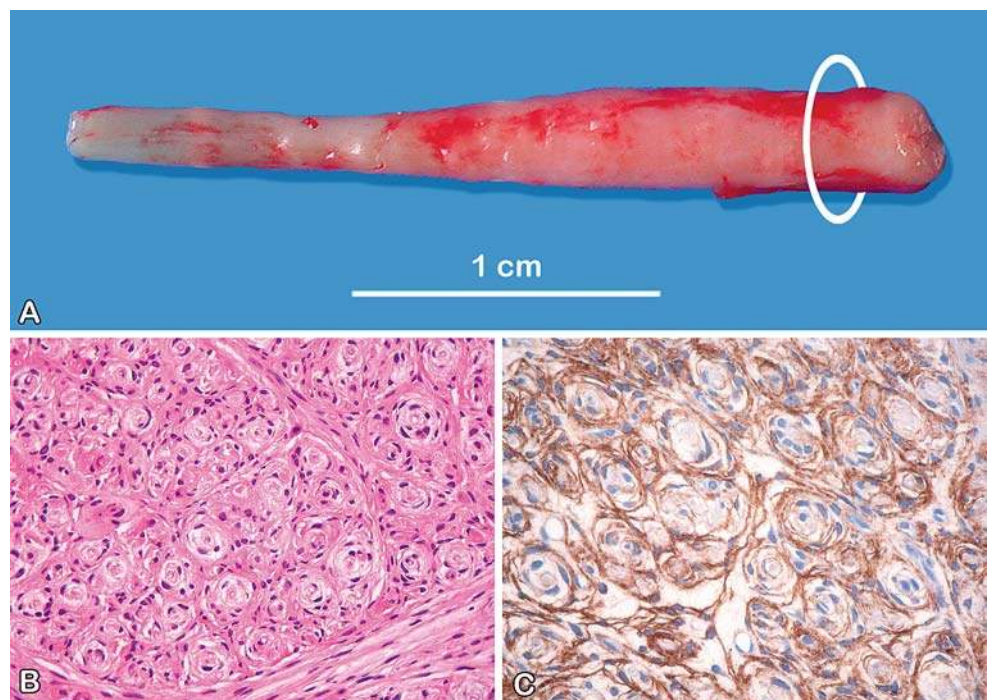
involvement of nerve roots is unclear and differentiation from femoral vessels is difficult. **c** Tractography is useful in distinguishing nerve from vessel and is better at depicting sacral root involvement

The fibre-tract method is based on the fact that peripheral nerves have anisotropic diffusion properties and can be visualized by DTI, distinguishing nerves from other tissues, such as vessels, that do not possess anisotropic diffusion [8, 9]. Marked prolongation of the T2-relaxation time is nonspecific and does not differentiate between denervation oedema, demyelination and tumour. In our study, DTI showed a decrease in FA value and an increase in ADC in the sciatic nerve, i.e. a ‘tumour-like’ appearance. We have been unable to identify in the literature other examples of pathological peripheral nerves imaged by DTI. It would be interesting to determine if the FA value increases and ADC decreases in inflammatory and degenerative causes of hypertrophic neuropathy. Further studies are needed to find out if this technique will allow

differentiation of tumour from hypertrophic neuropathy with histological features of demyelination/remyelination, such as Dejerine-Sottas disease, or CIDP.

Resection of the lesion with end-to-end and sural nerve grafting appears to result in clinical stabilization, and in partial improvement in muscle function. However, this technique is not recommended if spinal roots are involved or if the unhealthy segment of nerve is too long [5]. Fat-saturated T1-W images show lesion extension but do not indicate whether the sacral roots are involved. Involvement of the sacral roots was well depicted by DTI with tractography in this patient. This, together with the long length of sciatic trunk hypertrophy, indicated that neurolysis was the best possible surgical option after biopsy and excluded sural nerve grafting.

Fig. 2 Pathology. **a** Macroscopic view of the nerve trunk, with progressive diameter enlargement. Histological sections are at the level of the white circle. **b** H&E stain demonstrates multiple ‘pseudo-onion bulbs’ that are composed of circumferentially arranged cells (original magnification $\times 400$). **c** EMA positivity of tumour cells (original magnification $\times 600$)



In conclusion, MRI can exclude nonsurgical causes of painless neuropathy involving major nerve trunks. This technique distinguishes hypertrophic nerves from vessels and clearly depicts nerve root involvement. A protocol combining 3-D T2-W STIR SPACE, DTI with tractography and fat-saturated gadolinium-enhanced T1-W images is a promising new method that may offer a novel approach to imaging peripheral nerves and roots in children, and provide a better understanding of the underlying pathophysiology. However, further studies are required to prove the usefulness of this technique in this and other paediatric peripheral nerve disorders such as brachial plexus birth injury, demyelination/remyelination conditions and other tumours.

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