Nerve conduction assessment and magnetic resonance imaging for the diagnosis of localized hypertrophic neuropathy of the sciatic nerve and the lumbo-sacral plexus

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ABSTRACT

Localized hypertrophic neuropathy (LHN) are slowly growing nerve lesions causing progressive nerve deficit and weakness. We present the case of a 32-year old woman with long history of motor and sensory deficit complains along the sciatic nerve territory. The muscles involved were featured by delay in F waves at nerve conduction assessment. Magnetic resonance imaging (MRI) showed specific patterns, low intense on T1 and abnormally hyper intense on short tau inversion recovery (STIR) and T2, with no obvious enhancement, features compatible with either LHN or intraneural perineurioma (IP) of the sciatic nerve and/or the lumbosacral plexus. Focal thickening and hypertrophy of the sciatic nerve with preserved fascicular configuration and progressive enlargement of the right lumbosacral plexus could be noted. A nerve conduction assessment followed by an MRI eventually allowed to diagnose LHN, without performing a nerve biopsy. Although similar, LHN and IP are two distinct lesions which should be diagnosed and differentiated as soon as possible, to avoid potential complications due to delayed diagnosis and/or misdiagnosis.

1. Background

In 1964, Imaginariojda et al. reported the first case of localized hypertrophic neuropathy (LHN) [1], an idiopathic fusiform bulging of a peripheral nerve causing progressive weakness, painless and sensory neurological deficit along the nerve territory involved [2,3]. LHN is a relatively rare and slowly growing "onion-bulb" lesion leading to fusiform local enlargement of the peripheral nerves involved [4,5].

2. Case presentation

We present the case of a 32-year-old woman with long history of numbness and paresthesia in the sciatic nerve distribution, with gradual onset of atrophy in her right lower leg muscles. Although being under treatment for the latter condition, the patient's symptoms gradually worsened over time.

The patient medical history was negligible. At neurologic examination, a clear strength diminishment of the lower limb muscles – especially the gluteus – could be noted. Furthermore, the Achilles tendon reflex was nearly absent.

Conduction velocity showed delay in F waves of the peripheral nerves involved, especially the right posterior tibial nerve. MRI images revealed typical imaging pattern of LHN in the sciatic nerve and the lumbosacral plexus: focal fusiform nerve thickening and hypertrophy; progressive enlargement of the right lumbosacral plexus and sciatic nerve diameter with preserved normal fascicular and linear configuration, without any pathologic enhancement. However, whilst a low signal on T1-weighted images and an abnormally hyperintense signal on STIR and T2-weighted images could be noted, no intense enhancement at axial view following i.v. administration of gadolinium was detected.

Abbreviations: LHN, Localized hypertrophic neuropathy; IP, intraneural perineurinoma; MRI, magnetic resonance imaging; STIR, short tau inversion recovery; EMA, epithelial membrane antigen (); PNST, peripheral nerve sheath tumor ().

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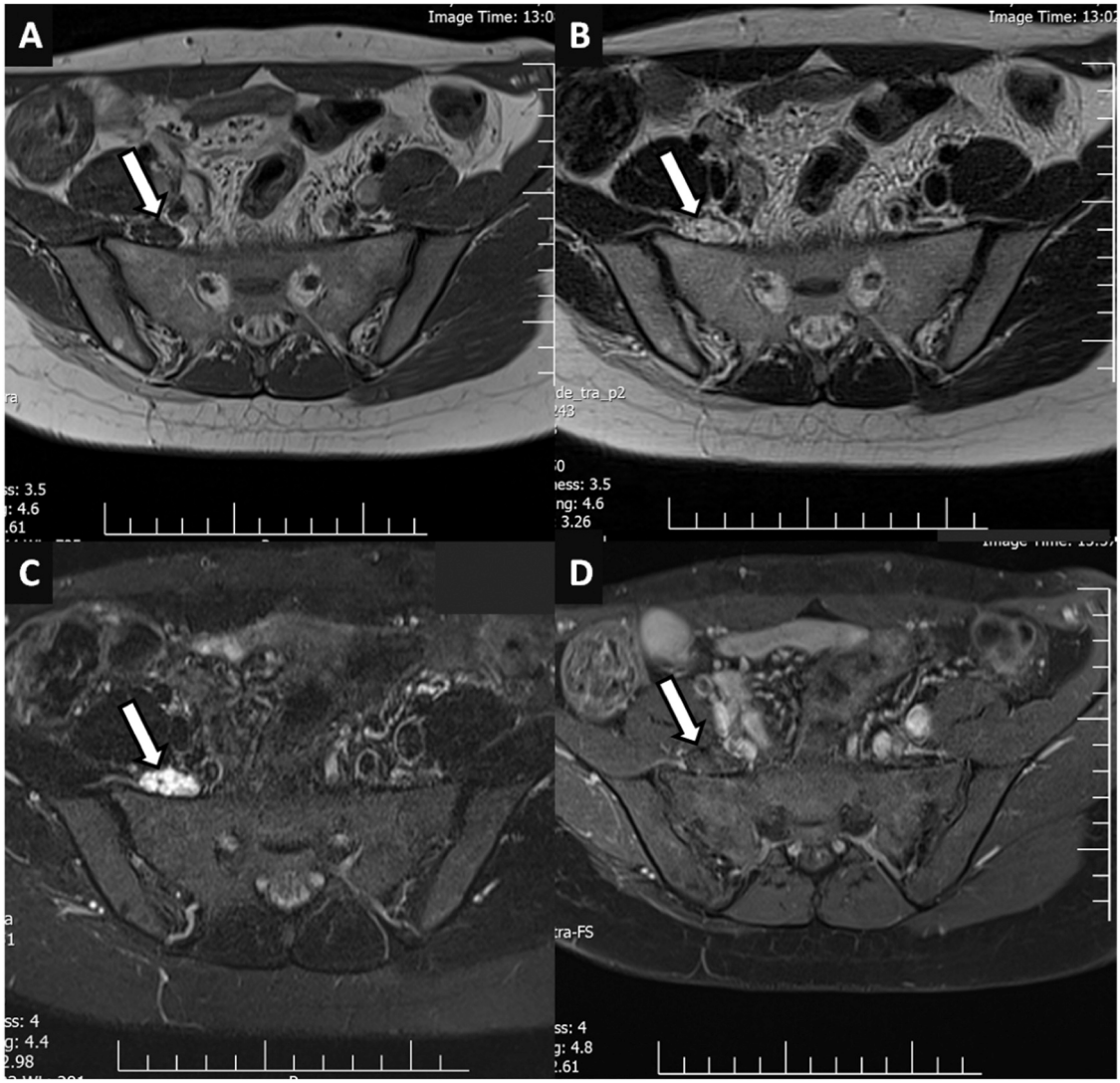

Fig. 1. Axial Magnetic Resonance Imaging (MRI) show focal thickening and hypertrophy with progressive enlargement of the right lumbosacral plexus or sciatic nerve diameter (white arrow) compatible with either localized hypertrophic neuropathy (LHN) of the sciatic nerve and lumbosacral plexus: (A) low signal on T1-weighted images; (B and C) abnormal hyperintensity of the nerves on STIR and T2-weighted images; (D) no obvious enhancement after intravenous gadolinium administration.

Furthermore, severe atrophy and fatty infiltration in the right gluteus maximus muscle were visible (Figs. 1 and 2). Coronal images confirmed specific mentioned neural findings (Fig. 3), prompting the diagnosis of LHN of the sciatic nerve and the lumbosacral plexus.

3. Discussion

A confined whorl proliferation of Schwann cells and fibrous tissues wrapping a variably myelinated axons of peripheral nerves [4,5]. LHN most frequently involves the brachial plexus and the radial, median, sciatic, peroneal, tibial and trigeminal nerves [4,6]. LHN of the sciatic nerve was reported in a 73-year-old man affected by Klippel-Trenaunay Syndrome, a rare congenital vascular disorder in which a limb may be affected by port wine stains [6]f. LHN of spinal nerve roots – from the conus into the upper leg – have also be reported [4].

LHN is most likely a hyperplastic reaction to nerve damage [5,7] and should be distinguished from other benign peripheral nerve sheath tumors (PNST) as neurofibroma, Schwannoma and especially intraneural perineurioma (IP) [8].

IP is a proliferation of perineural cells forming a “pseudo-onion” in the peripheral nerves affected [4,9]. The nerves most frequently affected by IP are the sciatic nerve, followed by the radial nerve [10]. Since pure perineuriomas are rare and most lesions classified as IP are instead LHN, the term “perineurioma” was proposed as a comprehensive category for a range of nerve conditions including neurofibroma, LHN and IP [11]. Nevertheless, the term IP should be properly restricted to PNST of perineural cells [8]. According to their location, perineuriomas are classified into extra-neural (soft-tissue) or intraneural [12].

Whilst Schwann cells forming LHN are typically S100-positive and epithelial membrane antigen (EMA)-negative, perineurial cells surrounding neural fascicles inside a nerve affected by IP are EMA positive and S-100 negative [5,8,13]. Moreover, IP predominantly presents affects children and young adults of 14–23 years of age [14].

MRI allows to diagnose LHN, as in our 32-year-old female patient, excluding PNST or nerve compressions. Whilst both IP and LHN manifest in fact as a fusiform enlargement of the affected nerve with preserved fascicular architecture at high field MRI – isointense on T1 and hyperintense on T2 weighted images – IP is featured by atrophy and fatty infiltration of the muscle involved a well as variable enhancement following intravenous gadolinium administration [3,9,15,16].

MRI successfully enabled to diagnose LHN of the sciatic nerve in four children aged 2-12 years [3] and a LHN of the brachial plexus in a

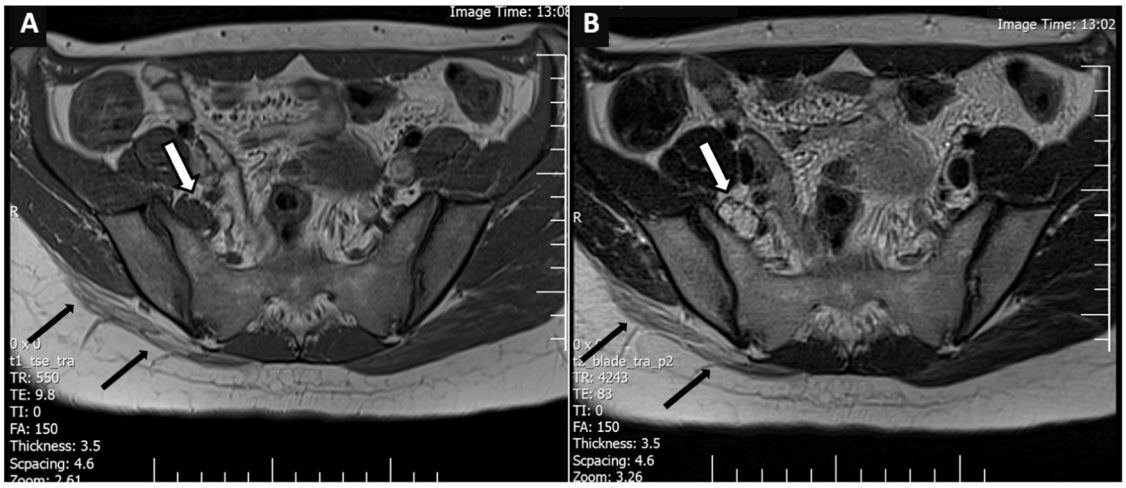

Fig. 2. Axial Magnetic Resonance Imaging (MRI) show focal thickening, hypertrophy and enlargement of right sciatic nerve diameter (white arrow), with severe atrophy and fatty infiltration of the right gluteus maximus muscle (black arrows): (A) low signal on T1-weighted images; (B) abnormal hyperintensity of the nerves on T2-weighted images.

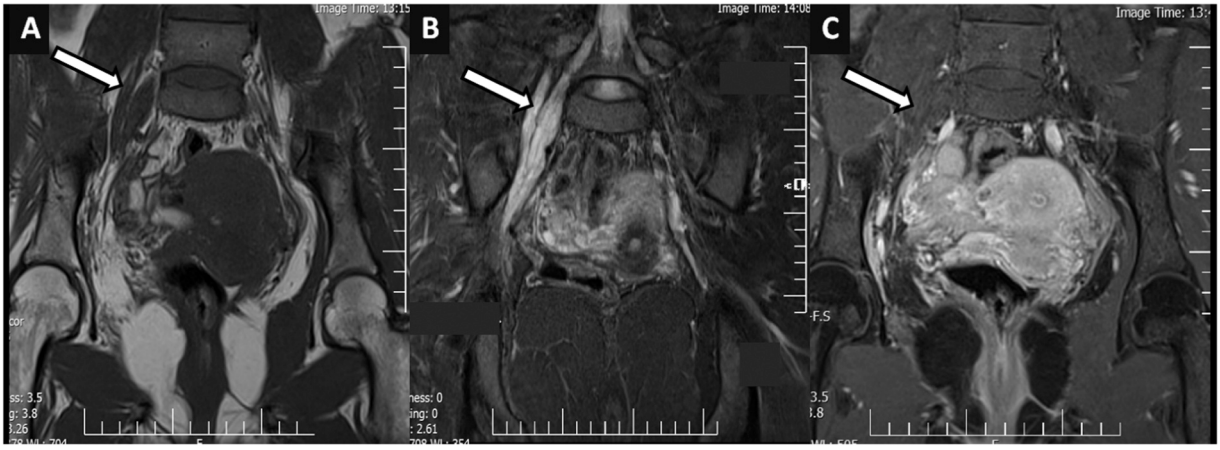

Fig. 3. Coronal Magnetic Resonance Imaging (MRI) show fusiform focal thickening and hypertrophy with progressive enlargement of the right lumbosacral plexus and sciatic nerve diameter (white arrow), compatible with either localized hypertrophic neuropathy (LHN) of the sciatic nerve and the lumbosacral plexus: (A) low signal on T1-weighted images; (B) abnormal hyperintensity of the nerves on STIR images; (C) preserved normal fascicular and linear configuration; (C) no enhancement following intravenous gadolinium administration.

10-year-old girl [17]. Several other reports emphasized the benefit of MRI in the characterization of LHN [3,18–22].

Radiologists should therefore always consider the differential diagnosis of LHN with other PNST (as IP, Schwannoma, neurofibroma) or neuropathies as Charcot-Marie-Tooth disease [2,14,23,24]. Although similar, LHN and IP are two distinct lesions which should be diagnosed and differentiated as soon as possible, to avoid potential complications due to delayed diagnosis and/or misdiagnosis. MRI can support clinical decision-making, since whilst the main treatment approach for some PNST is surgery, there are no established clinical guidelines for LHN, whose treatment is decided based upon patient's symptoms and medical conditions.

Therefore, a nerve conduction assessment followed by a typical MRI pattern allowed a reliable diagnosis of LHN in the present case, with no need of performing invasive nerve biopsy.

Ethics approval and consent to participate

The institutional review board of the University of Baqiyatallah University of Medical Sciences ethics approval for this study.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report.

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Authors' contributions

Ramezan Jafari, Luca Cegolon, Nima Mohseni Kabir, Fatemeh Dehghanpoor, and Mohammad Javanbakht performed data collection, case description, and acquisition of data, interpretation, and critical revision of the manuscript. The author(s) read and approved the final manuscript.

Competing interests

The authors declare that there are no competing interests.

Data availability

All data collected, generated and analyzed in this study are included in the published article.

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