Chronic inflammatory demyelinating polyneuropathy in a child: clinical-spinal MR imaging correlation

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ABSTRACT

Spinal magnetic resonance (MR) imaging of a 3-year-old girl with chronic inflammatory demyelinating polyneuropathy (CIDP) showed thickened and marked enhancement of the lumbosacral nerve roots. These abnormalities resolved after steroid treatment. MR imaging of the cauda equina may be helpful in the diagnosis of CIDP.

Keywords: Chronic inflammatory demyelinating polyneuropathy, magnetic resonance imaging, nerve root enhancement, spinal root hypertrophy

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INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired, immune-mediated peripheral neuropathy[1]. It is characterised by chronic progressive or relapsing and remitting weakness, and sensory loss in multiple limbs[1]. The spinal magnetic resonance (MR) imaging findings of CIDP have been documented but mostly for adult patients[2-5]. We report a case of CIDP in a child with thickening and abnormal enhancement of the sacral nerve roots that resolved after steroid treatment.

CASE REPORT

A previously-healthy 3-year-old girl presented with a two-week history of progressive difficulty in walking. There was no history of similar illness in her family. On examination, there were mild proximal and more pronounced distal weakness, and absent tendon reflexes of all limbs. Cerebrospinal fluid (CSF) analysis showed no cells but slightly increased protein (80mg/dL). An electrophysiological study showed prolonged distal latency, slow nerve conduction velocity, and absence of F-waves for the upper and lower limbs. Her weakness was stable at three weeks after onset without specific treatment. A diagnosis of acute inflammatory demyelinating polyneuropathy was made. Eight weeks later, her weakness was worse and there was an onset of neck extensor weakness. CSF analysis revealed an elevated protein of 102mg/dL but no cells. Unenhanced MR imaging of the spinal cord was normal. T1-weighted imaging after gadolinium injection showed thickening and marked enhancement of the nerve roots in the conus medullaris and cauda equina (Fig. 1). CIDP was diagnosed and the patient was treated with 2mg/kg/day of prednisolone. One month later, she was able to walk with a walker and at two months, she was able to walk independently. Follow-up spinal MR imaging at seven months after treatment was normal (Fig. 2).

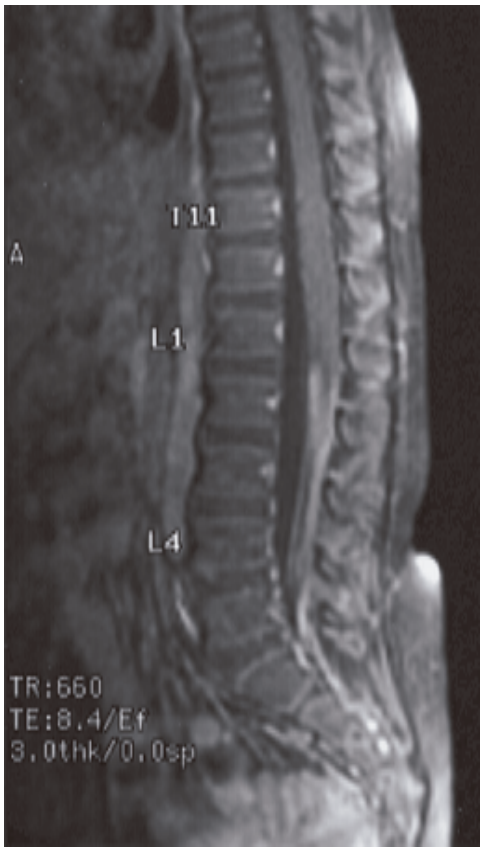

Fig. 1 Enhanced sagittal T1-W image (TR660/TE8.4) of the thoracolumbar spine shows abnormal enhancement of lumbosacral nerve roots.

DISCUSSION

CIDP is relatively rare in children[6-8]. One epidemiological study reported an overall prevalence of CIDP of 1.9 per 100,000 and a childhood prevalence

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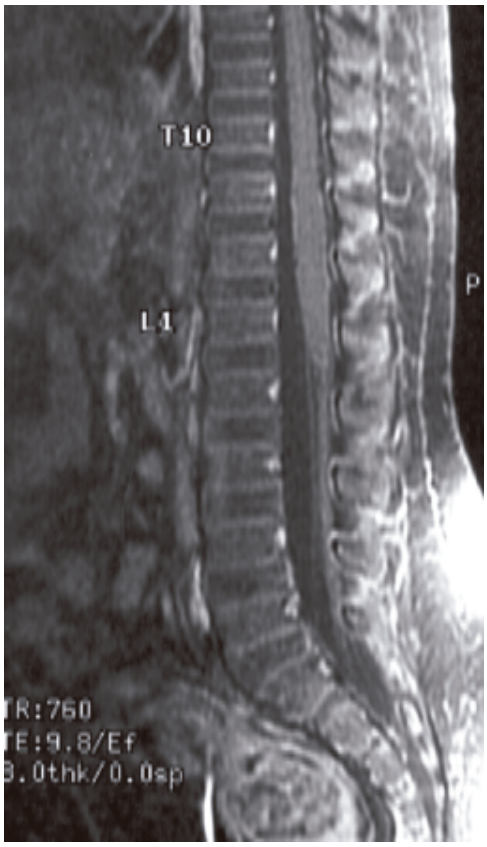

Fig. 2 Follow-up enhanced sagittal T1-W MR image (TR760/TE9.8) taken 7 months after prednisolone treatment shows no abnormal enhancement of lumbosacral nerve roots.

(age less than 19 years) of 0.48 per 100,000[6]. The prognosis of CIDP in children and younger patients (age less than 30 years) is better than the prognosis for patients who are older at onset[6-8].

Although a nerve biopsy was not done, our patient's clinical history, CSF findings, and electrophysiological findings supported a diagnosis of relapsing-remitting CIDP. MR imaging of the spine showed thickening and enhancement of the sacral nerve roots. These findings have been reported in patients with CIDP, most of whom were adults. In one study, gadolinium enhancement might have been present in active disease[4] but another study did not support this finding[5]. Thickening of the nerve roots with abnormal enhancement of the cauda equina and lumbosacral spinal roots has been reported in patients with various disorders, including arachnoiditis, meningitis, multiple schwannomas, acquired immunodeficiency syndrome-related polyradiculopathy, sarcoidosis, lymphomatous/leukemic infiltration, metastatic disease, and Guillian-Barre syndrome[9,10].

No family history, a rather rapid rate of progression, a relapsing-remitting course of illness and multifocal abnormlities found on neurophysiological study ruled out a diagnosis of hereditary motor and sensory neuropathy (HMSN) and Dejerine-Sottas disease. MR imaging of patients with HMSN may show hypertrophy of nerve roots but if it does, it is without contrast enhancement[5]. Abnormally enhanced lesions of the conus medullaris and cauda equina are seen in Guillian-Barre syndrome[10], but enlargement of nerve roots and a relapsing-remitting course suggests a diagnosis of CIPD[5].

It is now believed that CIDP is an autoimmune disease that targets the myelin sheaths of peripheral nerves[1]. Abnormal enhancement of the spinal nerve roots probably results from an inflammatory or infiltrative process breaking the blood-nerve barrier[2], while hypertrophy of the spinal nerve roots and cauda equina has been attributed to recurrent segmental demyelination and remyelination[3]. The normal T1- and T2-weighted images in our case may have been due to the very subtle lesions. However, on a contrast study, MR imaging of the spine showed thickening and enhancement of the sacral nerve roots, which disappeared after steroid treatment. These findings support the hypothesis that CIDP is an immune-mediated disease.

As with adult patients with CIDP, MR imaging of the spine may be a very helpful diagnostic tool in childhood-onset CIDP. In addition, abnormal enhancement of nerve roots may indicate the presence of active disease.

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