Cranial nerve hypertrophy in pediatric chronic inflammatory demyelinating polyradiculoneuropathy

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A 14-year-old boy presented with 12 months of bilateral foot weakness and paraesthesia. His exam revealed areflexia, distal muscle weakness and decreased pinprick and vibration sense to his fingers and below mid-thigh level. Cranial nerves were intact. Nerve conduction studies identified a demyelinating polyneuropathy with dispersion and conduction block. Spinal fluid protein was elevated. MRI revealed cranial nerve, cervical (Fig. 1), thoracic and lumbosacral nerve root hypertrophy (Fig. 2). He recovered well with monthly intravenous immunoglobulin.

Fig. 1 Coronal STIR MRI shows diffuse symmetrical hypertrophy of cervical roots

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is associated with clinical evidence of cranial nerve dysfunction in about 15% of those affected. MRI may reveal cranial nerve hypertrophy and enhancement even without clinical deficit [1]. MR evidence of spinal root hypertrophy occurs in 60% of adult patients, typically those with long disease duration [2]. Root thickening may be seen in paediatric CIDP but, unlike that in our patient, is generally mild.

Fig. 2 Coronal STIR MRI shows lumbosacral roots (enhancement not shown)

References

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