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CIDP PRESENTING AS MYELORADICULOPATHY

Neurology

Sundarachary.N.V
Mythri.A.* *Corresponding Author

ABSTRACT

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a relapsing or chronic progressive disorder which is one of the causes of hypertrophic neuropathy. Enlarged nerve roots were identified in few patients. We now report a case of CIDP with highly thickened or enlarged nerve roots severe enough to cause cord compression and myelopathy. A 38 year old woman presented with weakness and sensory disturbances in lower limbs with sensory level at L1 and bladder disturbances in form of urgency and urge incontinence . MRI showed a non enhancing isointense mass lesion from L1 to S1 causing compression over conus . During surgery the cauda equina nerve roots were found to be thickened , entangled with ' bag of worms' appearance. Nerve conduction studies showed all her peripheral nerves to be inexcitable. Her CSF protein was mildly elevated. Diagnosis of CIDP with thickened nerve roots was considered and she was started on steroids and subsequently showed improvement. Thus, CIDP may present with symptoms of myeloradiculopathy due to thickened nerve roots causing cord compression.

KEYWORDS

CIDP, root hypertrophy , bag of worms appearance, cord compression.

INTRODUCTION

CIDP is a inflammatory demyelinating polyradiculoneuropathy with relapsing or chronic progressive weakness of limbs associated with distal sensory disturbances and hyporeflexia.[1] CSF protein is almost always raised.[2] NCS shows slowing of motor conduction with conduction blocks and sural nerve biopsy shows segmental demyelination and remyelination. [1,2]. CIDP is one of causes of hypertrophic neuropathy . Thickened peripheral nerves were detected in 11 percent of one large series and in up to one third of relapsing cases.[3] Greatly enlarged nerve roots are identified in a few typically affected patients at necropsy [4,5], by myelography [6], at surgery [7] and by MRI[8,9,10].

We now report a case who presented with symptoms suggestive of myeloradiculpathy in whom surgery and MRI showed massive nerve root hypertrophy . Subsequent investigations showed it to be CIDP.

CASE REPORT

A woman aged 38 years , presented with complaints of low backache radiating down to both thighs , weakness and tingling and numbness of both lower limbs and girdle like sensation around lower trunk of six months duration. She complained of urinary urgency and urge incontinence.

Examination showed weakness of (4/5) lowerlimbs both proximal and distal , and decreased pain and temperature sensations below L1. Joint position sensation was lost in lower limbs. All deep tendon reflexes were absent and plantar response was mute. Romberg's sign was strongly positive. Contrast enhanced MRI of spinal cord showed non enhancing intradural mass lesion in spinal canal from L1 to S1 causing compression over conus. Thoracic spinal cord showed a prominent central canal.

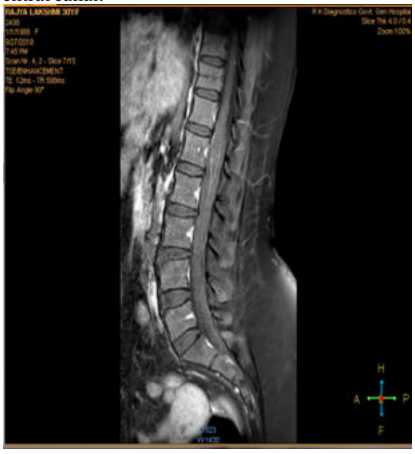

Figure 1. *MRI showing intradural mass lesion in spinal canal from L1 -L5 causing compression over conus.*

She was operated with aim of decompression and during surgery the cauda equina nerve roots were found to be thickened and entangled.

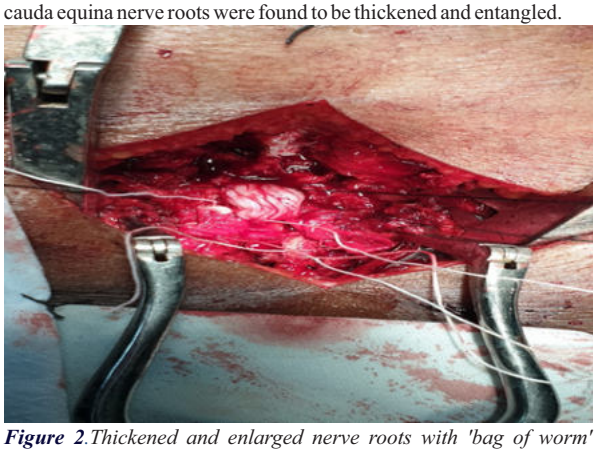

Figure 2*.Thickened and enlarged nerve roots with 'bag of worm' appearance.*

She was then referred to us and further questioning revealed tingling and numbness in hands and loss of joint position and graded sensory loss in upperlimbs with mild hand weakness. Nerve conduction studies showed all the peripheral nerves to be inexcitable. CSF protein was 46 mg/dl and cells were 9 lymphocytes/cu.mm . Diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy with thickened nerve roots was made. Nerve biopsy was not done because patient did not give consent. She was started on steroids and on follow up her sensory symptoms and power improved.

DISCUSSION

Our patient presented with symptoms of myeloradiculopathy. Myelopathic features include urinary urgency and urge incontinence and sensory level at L1 and girdle like sensation around lower trunk.

Polyradiculopathy features include pain radiating down both thighs and equal proximal and distal weakness of lower limbs. Myelopathy could be due to cord compression by enlarged nerve roots, or due to syrinx in lower cord (due to obstruction to CSF flow due to enlarged roots). Peripheral nerve thickening was not a feature in our patient.

Radiculopathy is an unusual presentation of CIDP.[1,2] But, it has been stressed that a pattern of weakness which include both proximal and distal is distinctive feature of CIDP[11]. This probably indicates painless root involvement. Commonly involved roots are lumbosacral and cervical. Why dorsal roots should be spared is unclear ,it raises the possibility that nerve thickening is in someway dependent on influence which arises in limbs but not trunk[12]. Most of the hypertrophic nerve roots are in cauda equina. [2]

Spinal root and plexus hypertrophy is seen on MRI , particularly in

cases of CIDP of long duration , gadolinium enhancement may be present in active disease.[13] Thus spinal MRI is a valuable addition to diagnostic armamentarium in CIDP.Even in absence of enlargement , abnormal enhancement of cauda equina is seen after intravenous Gadolinium.[12]

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