Concurrent Miller Fisher Syndrome Variant in Ossification of Posterior Longitudinal Ligament

1Alexander Cahyadi, 2Arwinder Singh, 3PS Ramani, 4Sudhendoo Babhulkar, 5Sumeet Pawar, 6Amrita Shenoy

ABSTRACT

Introduction: Miller fisher syndrome (MFS) could be found in coincidence with ossification of posterior longitudinal ligament. High index of suspicion is required that lead to further investigation.

Case report: A 56-year-old male presented with four days history of loss of sensation on both lower and upper extremity. The complaint was felt more on the lower than upper extremity. The patient felt imbalance during walking. Muscle strength of all extremity was normal, but sensory lost was found in all extremity. Deep tendon reflexes were absent in all extremity.

Investigations: Computed tomography (CT) scan and MRI showed canal stenosis due to ossification of the posterior longitudinal ligament. Nerve conduction velocity suggested peripheral neuropathy on both upper and lower extremity. Antibody anti GQ1b was positive. Cerebrospinal fluid examination showed cytoalbuminemic dissociation.

Treatment: Patient was treated conservatively.

Results: Improvement was achieved in 5 days, and progressively return to normal condition.

Conclusion: Peripheral polineuropathy could be found in coincidence with ossification of the posterior longitudinal ligament and required specific management.

Keywords: Ossification, Posterior longitudinal ligament, Miller Fisher syndrome.


Source of support: Nil

Conflict of interest: None

INTRODUCTION

The incidence of the ossification of posterior longitudinal ligament (OPLL) has been known to be 1.9 to 4.3% in Japan. Compared with the past, frequency of the disease has been observed to be gradually increasing possibly due to interest in OPLL.1 Up to 25% of patients presenting with cervical myelopathy have OPLL rather than spondylotic and stenotic myelopathy or disk disease alone.2 Patients with OPLL, increasing myelopathy and magnetic resonance imaging (MRI) evidence of progressing cord edema should be considered candidates for surgery if under 65 years of age. The conservative management of patients with OPLL may apply where patients have minimal neurological symptoms or signs.2 Cervical spondylotic myelopathy can present with subtle and varied presentations which require a high index of suspicion in the examiner.3 Miller Fisher syndrome (MFS) is a rare neurological disorder. It accounts for about 5% of patients with Gullain-Barrè Syndrome (GBS). In MFS patient, paresis of extraocular muscles progresses to complete bilateral ophthalmoplegia within 1 to 2 weeks, accompanied by areflexia, sensory disturbance and ataxia.4 Both OPLL and MFS could be found as a coincidence in the same patient as happened in our patient.

CASE REPORT

A 56-year-old male presented with 4 days history of numbness and tingling on both lower and upper extremity. The complaint was felt more on the lower than upper extremity. The patient felt imbalance during walking. It was so severe that he could not walk for more than few steps. The complaint increased since 2 days later, then he could not stand properly on his feet. There was no complain of vision. History of trauma or fever was negative, no diabetes and high blood pressure.

PHYSICAL EXAMINATION

He was in normal weight. Both arms and legs’ range of motion (ROM) was normal. There was no wasted muscle. Both bisp and tricep jerks were absent, and both knee and ankle jerks were absent. The muscle strength of all extremity was normal. Plantars were flexors. Straight leg raise (SLR) was normal (70º) on both sides. There were sensory loss to pain and propriocep-
tive on both legs. Peripheral pulses in both limbs were normal. Sphincters and blood pressure were also normal.

INVESTIGATIONS

Routine hematology was normal computed tomography (CT) scan of the cervical spine (Figs 1A and B) showed segmental ossification of posterior longitudinal ligament at C3-C4. The MRI of cervical spine (Figs 2A and B) showed significant compression of cervical spinal cord due to thickening of posterior longitudinal ligament at C3-C4-C5-C6-C7.

Electromyography (EMG) study were then performed and showed doubtful response in upper and lower limbs, suggestive of early GBS. Antibody examination was performed, and found anti-GQ1b antibody (+). Cerebrospinal fluid (CFS) examination showed no cell and protein was 235 mg/dl.

MANAGEMENT

The patient was administered corticosteroid (Solu Medrol) injection 3 × 1 gm for 3 days and followed by IV immunoglobulin 25 mg/day for 5 days.

FOLLOW-UP

The patient had significant improvement in his symptoms after having corticosteroid injection on day 3. Further improvement was noted after administration of immunoglobulin. He was mobilized at day 5, and sent home at day 13 while he was able to walk without assistance but still with imbalance. Two weeks after sending home, he came for follow-up, and able to walk freely without any symptom.

DISCUSSION

Surgical treatment of OPLL is clearly indicated when the patient shows severe and/or progressive cervical myelopathy, and surgery should be performed as soon as possible. However, for patients without myelopathic symptoms or with only mild myelopathy instead of evident OPLL on plain radiography of the cervical spine, the indications and timing of surgical treatment remain controversial. The critical ratio for the development of myelopathy has been reported within a range of 30 to 60%. Several authors have focused on the size of the space available for the cord (SAC). This indicator of myelopathy varied from 6 to 9 mm. No evidence has been accumulated for surgical treatment of asymptomatic or mildly myelopathic, considering the risks of surgery and the variable natural course of OPLL. Classically, myelopathic patients complain of gait instability and have problems with balance. In OPLL patient, the symptom was developed slowly progressive. Family members may comment that the patient walks as if he/she is intoxicated. Upper extremity complaints may include impairment of fine-motor function, grasp and sensation, leading to feelings of ‘clumsiness’ or ‘dropping things’ and diffuse (typically nondermatomal) numbness. Subjective weakness is a relatively late complaint. It was also happen in our patient, while the main sign and symptom were ataxia.

Hyperreflexia may be present in the upper and/or lower extremities and is suggestive of spinal cord compression. However, because peripheral nerves and roots must be functioning properly to transmit the hyperreflexia of myelopathy, patients with concomitant myelopathy and peripheral nerve disease from conditions, such as diabetes and peripheral neuropathy or those with root level compression from foraminal stenosis, may have diminished or absent reflexes. In our patient, the deep tendon reflex was negative. This finding makes suspicion of coincidence of peripheral neuropathy or neuronopaties. It was supported by the acute and rapid progression of the symptom.

Miller Fisher Syndrome, a variant of the GBS, is characterized by ophthalmoplegia, ataxia and areflexia. In our patient, ataxia and areflexia was not accompanied by ophthalmoplegia, and further examination was done accordingly. The high protein content and low cell count in the CSF, named albuminocytological dissociation, is a typical feature in MFS. The CSF protein concentration is normal in some patients during the first week of MFS but
may rise due to serial lumbar punctures made in subsequent weeks. Anti-GQ1b IgG antibody was found in MFS patients during each week after the onset of illness. Anti-GQ1b antibody testing in particular was much more useful than a CSF examination for supporting a diagnosis of MFS during the first week. In our patient, we found both of them was elevated in the first week. Peripheral nerve dysfunction is demonstrated in the patient, that been confirm by nerve conduction velocity. The sensitivity of electrophysiological investigation in supporting a polyneuropathy is 85% at admission and 93% at the nadir with a specificity of 100% in patients who are not able to walk without support. Miller Fisher syndrome is generally regarded as a self-limiting, benign condition MFS patients in the largest published series returned to normal activities 6 months after neurological onset. The respective median (range) periods between neurological onset and the disappearance of ataxia and ophthalmoplegia were 32 (8-271) and 88 (29-165) days. Our patient was mobilized at day 5, and sent home at day 13 while he was able to walk without assistance but still with imbalance. Two weeks after sending home, he came for follow-up and able to walk freely without any symptom.

**CONCLUSION**

High index of suspicion of coincident of peripheral polyneuropathies in OPLL patient is very important. Careful anamnesis, physical examination and further examination were necessary to exclude them.

**REFERENCES**