Case Report

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A Rare case of Miller Fisher Syndrome with Ophthalmoplegia with Bulbar Palsy with Bifacial Weakness with Progressive Quadriparesi

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Abstract

A 20 years old female came to ED with c/o giddiness since 4 days, headache since 3 days with diplopia with slurring of speech, lower limb weakness, difficulty in swallowing, and dropping of left eyelid since 1 day. Patient had no any pre-existing comorbidity and not on any regular medication. Last menstrual period was on 5 days back. On examination Patient was vitally and haemodynamically stable. Diplopia on right sided distant vision present. Venous blood gas samples showed Hyponatremia. On CNS examination the Glasgow coma scale was 15/15 with power in upper and lower limb 5/5 bilaterally. There was no neck stiffness. Gag reflex present.

Patient was taken immediately on symptomatic supportive treatment and admitted in Intensive care unit. Patient was planned for contrast enhanced MRI brain, lumbar puncture for CSF study, chest x-ray, ultrasound whole abdomen. Blood samples for complete haemogram and other relevant blood investigations with serum electrolytes sent for further study.

Keywords: Miller Fisher Syndrome; Ophthalmoplegia; Anti GQ1b antibody.

INTRODUCTION

Miller Fisher Syndrome: Is an autoimmune rare acquired nerve disease named after Dr. Charles Miller Fisher, a Canadian neurologist. It causes weakness of eye muscles that leads to difficulty in moving the eyes. The disease is related to Guillain Barre Syndrome and so also leads to unsteadiness and impaired limb coordination and may also leads to absent tendon reflexes. It may also characterised with weakness of facial muscles, tongue, swallowing muscles and limb weakness and also may lead to respiratory failure. Childrens and adults both can be affected. Reasons behind the disease are generally viral and bacterial infections especially Campylobacter Jejuni and Haemophilus Influenzae and the symptoms may occur approximately after 4 weeks. The disease has three defining features as follows:

- Ophthalmoplegia i.e, weakness of eye muscles that leads to impaired eye movements and double vision
- Ataxia i.e, impaired coordination of limbs
- Areflexia i.e, absence of tendon reflexes

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Treatment for the syndrome includes intravenous immunoglobulins, plasmapharesis and steroids. Most of the patients recovers within six months and deaths are noted very rarely.

Ophthalmoplegia: is defined as paralysis of extraocular muscles which helps in eye movements. It is classified as

- Internal ophthalmoplegia in which papillary sphincter and cilliary ciliarymuscles are involved
- External ophthalmoplegia in which extraocular muscles are involved which includes superior rectus, inferior rectus, medial rectus, lateral rectus, the inferior oblique, and superior oblique muscles
- Complete ophthalmoplegia that includes both external and internal

Anti GQ1b Anti body: also known as Antiganglioside antibodies and were first found to react with cerebellar cells. These antibodies are mainly affecting Schwann cells. Initially its difficult to diagnose the Miller Fisher Syndrome because it mimics some other diseases too, for example, myasthenia gravis, botulism, basal meningitis, brain stem stroke, diphtheria, brain stem encephalitis. So the treating doctor can rule out the diagnosis by considering the test for anti GQ1b antibody which help to diagnose Miller Fisher Syndrome.

CASE STUDY

A 20 years old female came to ED with c/o giddiness since 4 days, headache since 3 days with diplopia with slurring of speech, lower limb weakness, difficulty in swallowing, and dropping of left eyelid since 1 day. Patient gave a history of lower respiratory tract infection and gastrointestinal upset one month ago. Patient The patienthad no any pre-existing comorbidity and not on any regular medication. Last menstrual period was on 5 days back. On examination Patient was vitally and haemodynamically stable. ECG showed normal sinus rhythm. Random blood sugar: 73 mg/dl.

On Auscultation: B/L air entry was present. On Per Abdomen examination: soft, non tender and bowel sounds were present. Urogenital The urogenitalexamination was insignificant. Diplopia on right sidedright-sided distant vision present. Venous blood gas samples showed Hyponatremia. On CNS examination the Glasgow coma scale was 15/15 with power in upper and lower limb 5/5 bilaterally. There was no neck stiffness. Gag reflex present.

Patient The patientwas taken immediately on symptomatic supportive treatment and admitted in Intensive care unit. Patient The patient was planned for contrast enhanced contrast-enhanced MRI brain, lumbar puncture for CSF study, chest x-ray, ultrasound whole abdomen. Blood samples for complete haemogram and other relevant blood investigations with serum electrolytes sent for further study.

Patient was admitted and discharged after 19 days in stable condition.

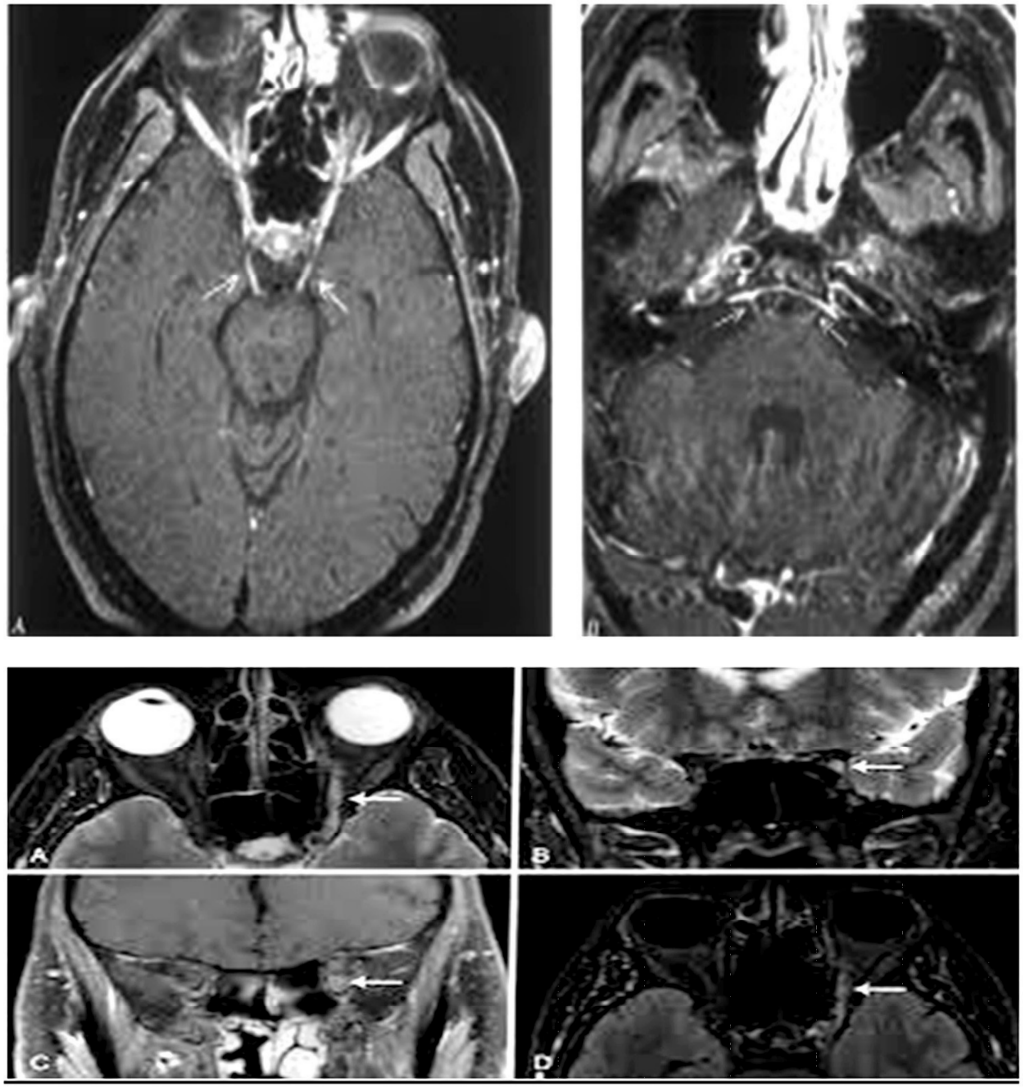
Course in the Hospital and Outcome:

A 20 year old female patient was admitted with above mentioned complaint. Patient was initially evaluated in emergency department and then shifted to intensive care unit for further management. Covid 19 rapid antigen test done in emergency department was negative. Other routine investigations were done. Ryle's tube was inserted and treatment was started with intravenous antibiotic, injection solumedrol, intravenous fluids and other symptomatic supportive treatment.

Patient came with MRI brain report done outside pre-arrival to the hospital which showed right frontal lobe hypodensity. Contrast enhanced MRI brain was done which revealed subcortical white matter edema involving right posterior frontal lobe. Lumbar puncture was done. Clear cerebrospinal fluid was obtained and placed in 4 vials. Needle was removed after adequate fluid collected. Procedure The procedurewas uneventful and sample was sent for evaluation. The differentials include demyelination, focal cortical dysplasia, focal edema.

Close monitoring suggested for further evaluation. MRI whole spine screening was done which revealed a normal study. Complete course of intravenous immunoglobulin (total dose of 90 gm) was given during the hospital stay. Ultrasound whole abdomen was done which revealed mild free fluid in the pelvis. Laboratory report shows hyponatremia. Chest x-ray showed right CP angle blunt.

Speech and language consultation was advised. Repeat laboratory report results appeared normal. The Ppatient was managed conservativelyand responded well to the given treatment. After stabilization, the patient was shifted to the ward. Regular monitoring was done and patient was discharged in stable condition after 19 days.



DISCUSSION AND THERAPEUTIC CONSIDERATIONS

Miller fisher syndrome is a very rare disorder. Some points we have to remember always that if the patient presents with neurological symptoms and ataxia, ophthalmoplegia, arreflexia areflexiaand had a history of respiratory tract infection and/or gastrointestinal upset some weeks/days before, we have to proceed carefully. It may be stroke or transient ischaemic attack or other disease like Miller Fisher Syndrome which mimics stroke. If there is any suspicion of presence of Miller Fisher Syndrome, we can proceed with the approach by following investigating tools:

- CSF study via lumbar puncture: there will be an albuminocytologic dissociation and/or normal cell count present with increased level of protein in CSF.
- CT/MRI scan of spine may show thickening and enhancement of Intrathecal spinal nerve roots and Cauda Equina. It can present with some spinal roots enhancement also.
- Electrodiagnostic studies: There will be diminished or absent sensory responses without slowing of sensory conduction studies.
- There is a validated and quantitative tool known as Brighton criteria that involves clinical history, physical examination, laboratory findings and radiological imaging to diagnose Miller Fisher Syndrome.

CONCLUSION

Why should an emergency physician be aware of Miller Fisher Syndrome: As we discussed above that Miller Fisher Syndrome mimics Acute stroke. Number of acute stroke cases has been increased comparatively as earlier because of more fat rich

unhealthy diet, lack of exercises, smoking, drinking, increased comorbidities and others modern life-induced reason. s, There is always a high chance of misdiagnosis of miller fisher syndrome. An emergency physician must know about miller fisher syndrome for the better approach. The condition can be easily treated if diagnosed earlier with intravenous immunoglobulins, pain control, steroids, respiratory support if needed, plasmapharesis and other symptomatic supportive treatment.

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