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Case Reports

CARDIO-PULMONARY COMPLICATION CAUSED BY AUTONOMIC NERVOUS SYSTEM IMPAIRMENT WITH THE MILLER FISHER SYNDROME IN A PATIENT WITH THROMBOCYTOPENIA AND RIGHT ATRIUM MASS. A CASE REPORTS

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A R T I C L E I N F O

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A B S T R A C T

Aim: literature review of MFS and case report.
Miller Fisher syndrome (MFS) is an acute inflammatory polyradiculoneuropathy that is generally considered a variant of Guillain-Barré syndrome (GBS) and is characterized by the clinical triad of ataxia, areflexia, and ophthalmoplegia. Causes Both GBS and MFS are triggered by a viral infection, most commonly the flu or a stomach bug. Symptoms generally start appearing from one to four weeks after infection with the virus. No one is entirely sure why GBS and MFS develop in response to these common illnesses. Several reports of familial Guillain-Barré syndrome have been reported, indicating a possible underlying genetic and/or environmental predisposition to the development of Guillain-Barré syndrome. Recently a few reports of MFS have been reported, with possible genetic predisposition. We report a case of 26 years old man with Cardio-pulmonary complication caused by autonomic nervous system impairment with the Miller Fisher syndrome with thrombocytopenia, renal failure and right atrium mass.
Conclusion: This report has shown that rare life threatening cardiovascular complications may occur not only in GBS, but also in MFS.

INTRODUCTION

MFS was first recognized by *James Collier* in 1932 as a clinical triad of ataxia, areflexia, and ophthalmoplegia. Later, it was described in 1956 by *Charles Miller Fisher* as a possible variant of Guillain-Barré syndrome (GBS). MFS is a subgroup of a more common, yet still rare nerve disorder known as Guillain-Barré syndrome (GBS). While GBS affects just 1 person in 100,000, MFS is even more uncommon. It makes up just 1 to 5 percent of Guillain-Barré cases in the Western world. Miller Fisher syndrome (MFS) is a variant of GBS that presents as rapidly evolving ataxia and areflexia of limbs without weakness and ophthalmoplegia, often with pupillary abnormalities.1

Miller Fisher Syndrome is observed in approximately 1-5% of all Guillain-Barre cases in Western countries. Patients with Miller Fisher Syndrome usually have good recovery without residual deficits.2

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Venous thromboembolism is a common complication of Guillain-Barre Syndrome and has also been reported in Miller Fisher Syndrome, but it has generally been reported in the presence of at least one prothrombotic risk factor such as immobility. 2

The MFS variant accounts for approximately 5% of all GBS cases. Anti-GQ1b immunoglobulin (Ig) G antibodies are found in >90% of patients with MFS, and titters of IgG are highest early in the course.

MFS involves both adults and children. MFS is a geographically variable variant of GBS observed in about 1% to 5% of all GBS cases in Western countries, yet up to 19% and 25% in Taiwan and Japan respectively.3

It is largely a clinical diagnosis and this distinctive syndrome comprises ophthalmoplegia, ataxia and areflexia which all develop over a period of few days without significant limb weakness.**1-4** There is an established male predominance at a ratio of 2:1 and a mean age of 43.6 years, although cases of MFS have been reported in all age groups.**5-6** The most common presenting symptom of MFS is diplopia caused by external ophthalmoplegia.**3,6** The spinal fluid protein is

elevated after the first week and patient recovers over a matter of weeks.3

The serum of over 90% of patients contain antibodies against the GQ1b and GT1a gangliosides of both peripheral and central nervous system. Brighton criteria is used for the diagnosis of MFS. MFS is usually having a self-limiting course.1,4

The treatment options are same as that of GBS, i.e. intravenous immunoglobulin and plasmapheresis but the overall impact of these treatment on eventual recovery is questioned.1,4 Upper respiratory tract infections is the most commonly described prodromal entity, followed by gastrointestinal illness 7,8,9.

The recovery period is marked by gradual improvement and resolution of symptoms, although rarely serious complications such as respiratory failure or cardiac arrhythmia have been reported.5 Ataxia and ophthalmoplegia resolve in 1 to 3 months after onset and near complete recovery is expected over several months to a year.3

There is very little published information about thromboembolic complications of MFS; however, in GBS, deep vein thrombosis (DVT) is common 10.

The treatment options are same as that of GBS. The recovery period is marked by gradual improvement and resolution of symptoms, although rarely serious complications such as respiratory failure or cardiac arrhythmia have been reported.11 Immobility, mechanical ventilation, and intravenous immunoglobulin (IVIg) therapy are common risk factors in GBS patients predisposing them to thromboembolic diseases 10,12.

However, the concomitant presence of both thromboembolic disease and GBS or MFS in a patient, without having an acquired or hereditary risk factor for thrombophilia, has not been previously documented in the literature.

Manifestations of autonomic dysfunction in patients with GBS include cardiac arrhythmias, BP fluctuations, sweating disorder, gastrointestinal dysfunction, and sphincter disturbance.13

Recent studies on GBS indicate that these complications are not only limited to severely disabled patients who require mechanical ventilation, but also affect those with milder forms of the condition, who do not require artificial ventilation and are able to walk more than 5 meters. 14,15Acute renal failure can occur in cases with GBS in those with dysautonomia.16 Some researchers speculate that the viruses may somehow change the structure of cells in the nervous system, causing the body's immune system to recognize them as foreign and fight them off. When this happens, nerves can't transmit signals well. The muscle weakness that's a characteristic of both diseases can result. Previous studies found that clinically overt cardiovascular autonomic disturbances such as tachycardia, bradycardia, hypotension, hypertension, and fluctuating BP were present in 27–79% of patients with GBS.17Similarly, to GBS, cardiovascular complications are rarely reported in MFS and are a complication of dysautonomia. 18,19

Case report

26 years old man with MFS, thrombocytopenia and renal failure with severe dyspnoea and pressure over the chest. In recent weeks, he became more tired and difficult with breathlessness. He was under treatment for MFS, but he had are flexia without ophthalmoplegia and ataxia. He was admitted to emergency department with suspicion of pulmonary embolism or MI.

Chest X-ray showed enlargement of right atrium with atelectasis of basal lob on left side. ECG on admission demonstrated incomplete (RBBB) and unspecific ST-T changes. *Fig-1*

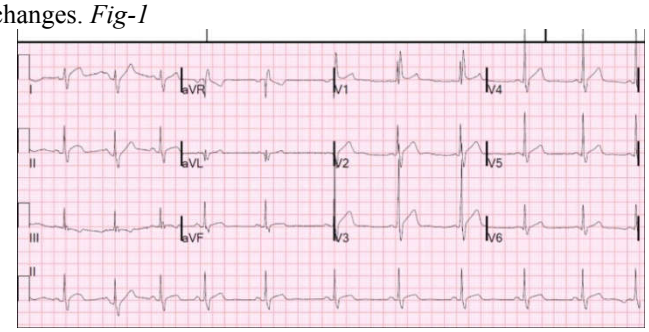

Fig 1 EKG with RBBB

A number of tests, including electrocardiograms (ECGs), blood tests, and coronary angiography were down, without evidence of MI.

Echocardiography showed a huge mass which was mobile in right atrium with changes in preload and afterload with Ejection Fraction = 45 and extension of right ventricle. ***Fig-2*** After consultation with hematologist and Cardiologist, He became subject to cardiac surgery.

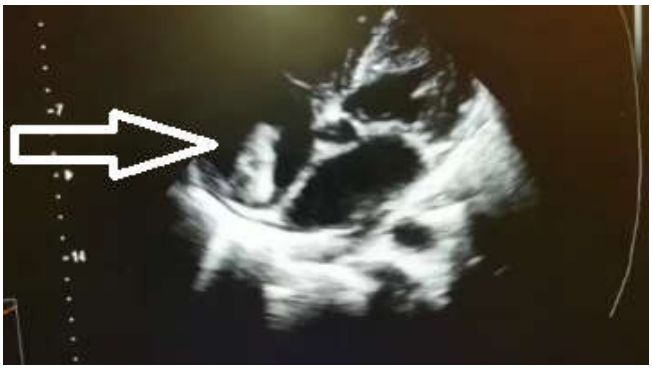

Fig 2 Echocardiography of right atrium mass

On operation with ECC without cardioplegia, the cardiac surgeon removed the mass which was 3*4 cm from right atrium and did a valvuloplasty of tricuspid valve. 24 hours post operatively, the patient discharge to cardiac rehabilitation ward and 5 days later discharge home. *Fig-3 and 4*

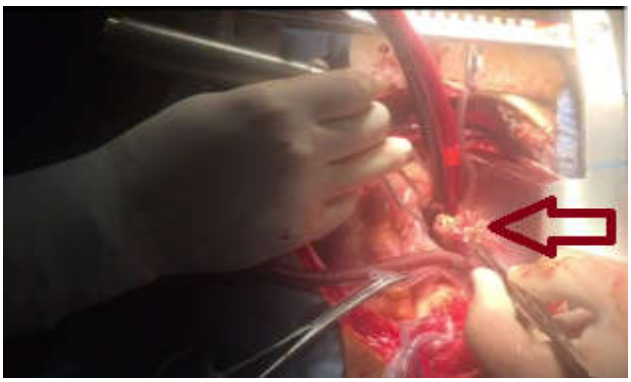

Fig 3 On ECC, right atriotomy and excision of the mass

Fig 4 Total excision of the cardiac Mass

CONCLUSION

Miller Fisher syndrome is a variant of GBS that presents as rapidly evolving ataxia and areflexia of limbs without weakness and ophthalmoplegia, often with pupillary abnormalities. Anti-GQ1b immunoglobulin (Ig) G antibodies are found in >90% of patients with MFS, and titters of IgG are highest early in the course.

The recognition of this syndrome is important as, despite its alarming nature, the course and outcome (after removing the offending cause and with early initiation of treatment with intravenous immunoglobulin or plasmapheresis) are optimistic. The familiarity with this rare syndrome will give a clue to the clinician to consider MFS as at least a differential diagnosis in a patient presenting with ataxia, areflexia and ophthalmoplegia.

This report has shown that rare life threatening cardiovascular complications may occur not only in GBS, but also in MFS.

***Disclosure**:* The authors declare no conflicts of interest.

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