Coexistent Mitral Stenosis and Multiple Sclerosis: An Unprecedented Case
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ABSTRACT
Mitral stenosis is a valvular heart disease characterised by narrowing of mitral orifice. It can lead to a dilated left atrium with atrial fibrillation culminating into thrombus formation. Patients with mitral stenosis, presenting with stroke-like episodes, most likely experience cardio-embolic phenomenon; but unusual and unprecedented associations do occur. Reported association of cardiac MS with demyelinating CNS MS has never been proven; but authors have speculated theories based on case reports and series. We report a rare case of mitral stenosis coexistent with multiple sclerosis, which is a chronic inflammatory demyelinating disease of central nervous system that affects both white and gray matters.

CASE REPORT
A 28-year gentleman known to have severe mitral stenosis and non-compliant to treatment for the last 4 years (Figure 1), presented to us with history of multiple stroke-like episodes for 18 months without visual impairment. His symptoms included variable combinations of weaknesses including monoparesis, paraparesis, hemiparesis, triparesis, ataxia, and weakness of one arm with opposite leg at different times, making him essentially bed bound. During each episode, he had a CT brain done and was labelled as having recurrent embolic strokes. We investigated in detail, and the patient was found to have classical MRI features and CSF findings of multiple sclerosis with multiple lacunar infarcts, warranting different line of treatment in addition to cardio-embolic strokes.


INTRODUCTION
Mitral stenosis is a valvular heart disease characterised by narrowing of the orifice of the mitral valve, mostly as a sequel of rheumatic heart disease. It can lead to a dilated left atrium with atrial fibrillation culminating into thrombus formation. It can be a source of cardio-embolic stroke per se, or via formation of mural thrombus, by showering emboli into CNS vasculature. Reported association of cardiac MS with demyelinating CNS MS has never been proven, but authors have speculated theories based on case reports and series.

We report a rare case of mitral stenosis coexistent with multiple sclerosis, which is a chronic inflammatory demyelinating disease of central nervous system that affects both white and gray matters.

Figure 1: Echocardiography showing severe mitral stenosis in the patient: (A) 4-chamber view showing mitral stenosis (arrow heads), (B) Mitral valve stenosis with calcified valves (arrow heads).

and was, consequently, treated with anti-platelets and anti-coagulants.

He presented to our facility with a similar event mimicking stroke by left sided hemiparesis with additional bilateral cerebellar signs. His deep tendon reflexes were brisk globally with bilateral extensor plantar response. Cranial nerves were intact and symmetric. Fundoscopic examination revealed normal discs and retina. He was investigated by extensive neuroimaging of both the brain and the spinal cord, using MRI with contrast as well as DWI and ADC sequences for brain. Spinal tap was also performed and CSF analysed for biochemistry, cell counts, and oligoclonal bands. The extended workup revealed classical MRI features and CSF findings of multiple sclerosis. These included periventricular, juxtacortical, infratentorial and spinal cord lesions and oligoclonal bands respectively. No evidence of new or old infarcts was found (Figure 2). As he fulfilled the clinical and radiological criteria, he was diagnosed with relapsing remitting type of multiple sclerosis (RRMS).

He was administered methylprednisolone pulse therapy for 5 days, followed by maintenance therapy. Supportive therapy and rehabilitation was offered to alleviate the
symptoms. He eventually underwent mitral valve replacement (MVR) with good clinical outcome. On follow-up at 6 months, he was asymptomatic regarding the cardiac symptoms and ambulatory with support. He still had marked ataxia, so was eventually put on maintenance therapy for multiple sclerosis with good remission on follow-up at six months till date.

**DISCUSSION**

Evidence regarding the prevalence of vascular co-morbidities in multiple sclerosis is conflicting. While some studies have reported that individuals with multiple sclerosis have an increased risk of ischemic heart disease, others have suggested that patients with multiple sclerosis are less likely to be hospitalised for ischemic heart disease. Findings regarding stroke also conflict for such patients.\(^5\) In 2015, Marrie et al. published a systemic review describing two studies conducted from 1977 to 2009 in Europe. They reported that prevalence of cardiac valvular disease at the time of first diagnosis of multiple sclerosis ranges from 0 to 2.3%. Neither study reported a statistical comparison of the prevalence of valvular disease between the multiple sclerosis population and controls, but valvular disease consistently occurred less often in the multiple sclerosis population.\(^5\) In 1981, Castaigne et al. presented the case of a 60-year woman with mitral and aortic stenosis with cortical deafness and transient right hemiparesis. She died 5 years later. Brain examination showed infarcts involving both middle cerebral artery territories. There were also numerous old plaques in the periventricular areas, thalamus, internal capsule, centrum semi-ovale, brain stem, and right nucleus dentatus. Authors suggested that the asymptomatic character of that multiple sclerosis case could be explained by the location of the plaques and the lack of spinal cord and optic tract involvement. They further suggested that it could also be due to the small size of the plaques.\(^6\) Uesugi and colleagues in 1999 described a case of mitral valve replacement under cardiopulmonary bypass, which got complicated by multiple sclerosis.\(^7\)

Papageorgiou and associates presented a case of multiple sclerosis in association with Williams-Beuren syndrome characterised by aortic valve disease.\(^8\) Some authors have suggested genetic association of multiple sclerosis rendering individuals susceptible to CNS MS; and no wonder, valvular heart disease has its own genetic affiliations. However, no correlation between the two has so far been proven.\(^9,10\) In all of these cases, authors have narrated the rarity of association of cardiac and CNS MS. As described, these cases pose a diagnostic challenge for physicians; and for some the diagnosis had not been clear until autopsy further elaborates the rarity of this relationship. Similarly, our case posed great diagnostic difficulty misleading to different diagnosis and, therefore, different treatment protocols. We, too, suspected cardio-embolic phenomenon with little chance of demyelination. Detailed review of the case and thorough work-up, however, did reveal the underlying association which seemed to be unprecedented.

We, therefore, conclude that while patients with mitral stenosis presenting with stroke-like features are most likely to have cardio-embolic phenomenon, unprecedented associations do occur. Reported association of cardiac MS with demyelinating CNS MS has never been proven, but authors have speculated theories based on case reports and series. This can pretty much be a diagnostic challenge and one, therefore, needs to keep demyelinating disease in differential diagnosis while dealing with challenging cases. This is important because failure to make a correct diagnosis can lead to wrong line of treatment, depriving patient of potentially manageable symptoms.

**REFERENCES**


