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BARLOW'S SYNDROME: A CASE STUDY

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ABSTRACT

Barlow's Syndrome a.k.a. Mitral valve prolapse (MVP) is a very common heart disorder which approximately affects 2-3% of the world population. It is described by typical fibrous and myxomatous changes in the mitral leaflet tissue with superior displacement of one or both leaflets into the left atrium. As 2-3% of the world is affected by it, MVP now would be affecting approximately over 176 million people worldwide. MVP could be related with significant mitral regurgitation (MR), bacterial endocarditis, congestive heart failure, and even sudden death.

MVP is a clinical term that is not fully discovered and perceived, despite human beings were aware about it for more than a century. In 1887 'mid - systolic click' was first depicted by Cuffer and Barbillon. In the year 1963 using angiography Barlow demonstrated the presence of MR in patients with the 'click-murmur' syndrome. Later in that way Criley coined the term mitral valve prolapse. MVP can be familial or sporadic type. Despite being the most widely recognised reason of isolated MR requiring surgical repair, little is known about the genetic mechanisms underlying the pathogenesis and progression of MVP. Studies on the heritable features of MVP have been limited by the analysis of relatively small families and by self-referral and selection biases, including a huge chunk of data from hospital - based partners. Nonetheless, a majority of data favours an autosomal dominant pattern of inheritance in a large proportion of individuals with MVP. Despite the variability in the clinical features, familial MVP might be considered very common Mendelian cardiac abnormality in humans. While filamin A has been identified as causing an X-linked form of MVP, the causative genes for the more common form of autosomal dominant MVP have yet to be defined.

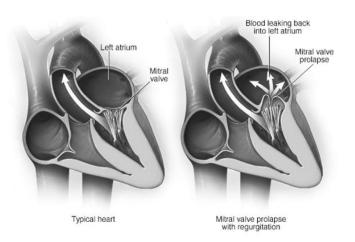
Keywords: Mitral valve prolapse, fibromyxomatous, atrium, regurgitation, angiography, autosomal

1. INTRODUCTION

The reason behind naming it mitral valve is because of its resemblance to a bishop's mitre, is the heart valve that prevents the backflow of blood from the left ventricle into the left atrium of the heart which is made up of two leaflets, one anterior and one posterior, that close when the left ventricle contracts. Each leaflet is composed of three layers of tissue : the *atrialis*, *fibrosa*, and *spongiosa*. Patients with classic mitral valve prolapse condition have excess connective tissue that thickens the spongiosa and separates collagen bundles in the fibrosa. This is due to an excess of dermatan sulfate, a glycosaminoglycan. This weakens the leaflets and adjacent tissue, resulting in increased leaflet area and elongation of the chordae tendineae. Elongation of the chordae tendineae often causes rupture, commonly to the chordae attached to the posterior leaflet. Advanced lesions — also commonly involving the posterior leaflet — lead to leaflet folding, inversion, and displacement toward the left atrium.

The real cause behind Mitral Valve Prolapse condition is still unknown to mankind, but is thought to be linked to heredity. Primary and secondary forms of Mitral Valve Prolapse are described as follows:

- **Primary Mitral Valve Prolapse**: Primary Mitral Valve Prolapse is characterised by thickening of one or both valve flaps. Other than that fibrosis (scarring) of the flap surface, thinning or lengthening of the chordae tendineae, and fibrin deposits on the flaps. The primary form of Mitral Valve Prolapse is seen more commonly in people suffering with Marfan's Syndrome or other inheri ted connective tissue diseases or disorders, but the people having no other forms of heart disease are found to be the most common victim of it.
- Secondary Mitral Valve Prolapse: In case of secondary Mitral Valve Prolapse, the flaps are not thickened. The prolapse may be due to ischemic damage (caused by decreased blood flow as a result of coronary artery disease) to the papillary muscles attached to the chordae tendineae or to functional changes in the myocardium. Secondary Mitral Valve Prolapse may result from damage to valvular structures during acute myocardial infarction, rheumatic heart disease, or hypertrophic cardiomyopathy (occurs when the muscle mass of the left ventricle of the heart is larger than normal).



CASE PRESENTATION

In April 2022, A 66 years old female house wife visited I.G.M.C., Shimla cardiology department with the complaints of chest pain and breathlessness from moderate to severe intensity, she was experiencing these symptoms from last 1 week. On physically examining the patient it was found that she already had Type 2 Diabetes Mellitus, some edema around various body parts and Hypertension. With unequal pulses of both the arms. Blood pressure measured from right arm (180/90 mmhg) and left arm (110/80 mmhg). Blood sugar was 250 mg/dl.

Past Medical History

Patient has a past medical history a caesarean section done in 1992. Patient also suffered a stroke of paralysis in 2018.

Past Medication History

Patient has a past medication history and was taking medications for blood pressure and diabetes from past 4 years. Tabs like Vildafree M550, Azulix MV2/0.2, Azulix MV1/0.2 and Tabs for blood pressure.

General Examination

Weight: 44 kg.

Height: 4 foot 10 inches

B.M.I.: 20.4 kg/m²

Physical activity: close to none after the stroke felling breathless and chest pain.

SPECIAL INVESTIGATIONS

C.B.C. - Complete Blood Count, P.P.B.S. - Postprandial Blood Sugar Test, ECHO Echocardiograppy and X - Ray Chest PA View

TREATMENT

Tab. Vildafree M550, Tab. Azulix MV2/0.2, Tab. Azulix MV1/0.2, Cap. Rosulip - ASP (10 & 75), Tab. Dytor (5mg), Tab. Etorica P (60 & 325), Gel Divon Gel (10 gm), Cap. Mylin Gard PG and Tab. Montair AB.

INTERVENTIONS

Patient was advised to control her blood sugar level and blood pressure by taking care of herself by walking daily as suitable. New medications were given to her along with diet plan.

CARE PLAN

Eat a healthy diet - Patient was advised to use til oil for cooking food, use saindhava salt, curry patta, gheeya, moong dal, kulcha daal, chana, only seasonal fruits, seasonal vegetables, use lehsun, methi, adaraka and coriander.

Patient was advised daily walking as much as possible. Patient was advised to avoid sweets and outside food.

OUTCOME

After getting results for all the tests the patient and her attendant was given a brief knowledge about her current health status. Patient was advised to take the prescribed medications. Patient was advised to visit hospital after 1 month for follow up.

2. DISCUSSION

MVP may be seen frequently in individuals suffering with Ehlers-Danlos syndrome, Marfan syndrome or polycystic kidney disease. Other risk factors include Graves disease and chest wall deformities such as pectus excavatum. For unknown reasons, MVP patients generally have a low body mass index (BMI) and are typically leaner than individuals without MVP. Also women tend to have joint hyper mobility.

Rheumatic fever is very common worldwide and responsible for many cases of damaged heart valves. Chronic rheumatic heart disease is described by repeated inflammation with fibrinous resolution. The cardinal anatomic changes of the valve occurs that includes leaflet thickening, commissural fusion, and shortening and thickening of the tendinous cords. The recurrence of rheumatic fever is relatively common in the absence of maintenance of low dose antibiotics, especially during the first three to five years after the first episode. Heart complications may be long-term and severe, particularly if valves are involved.

Rheumatic fever, since the advent of routine penicillin administration for Strep throat, has become less common in developed countries. In the older generation and in much of the less-developed world, valvular disease (including mitral valve prolapse, reinfection in the form of valvular endocarditis, and valve rupture) from undertreated rheumatic fever continues to be a problem. In an Indian hospital between 2004 and 2005, 4 of 24 endocarditis patients failed to demonstrate classic vegetations. All had rheumatic heart disease (RHD) and presented with prolonged fever. All had severe eccentric mitral regurgitation (MR). (One had severe aortic regurgitation (AR) also.) One had flail posterior mitral leaflet (PML)

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