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Mitral Facies: A Case Report

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Abstract

A 42-year-old woman presented with sudden weakness of left side. Her medical history was rheumatic mitral stenosis 8 years ago. On physical examination was remarkable her rosy cheeks, whilst the rest of the face has a bluish tinge called Mitral Face. It is the classic facial appearance in mitral stenosis, a plum-colored malar flush, occurs only when cardiac output is low and pulmonary hypertension is severe; cause is cutaneous vasodilation and chronic hypoxemia. In addition to cardiac auscultation was an irregularly irregular rhythm, a loud S1 and accentuated P2, an opening snap, and a III/IV decrescendo diastolic rumble at the apex. Transthoracic echocardiography revealed

findings typical of rheumatic mitral stenosis. The mitral leaflets were pliable, with restricted mobility of the leaflet tips, resulting in doming and a “hockey-stick” appearance of the anterior leaflet in diastole. There was notable subvalvular thickening. Cross-sectional imaging of the stenotic orifice demonstrated thickening restricted to the leaflet tips. Continuous-wave Doppler ultrasonography demonstrated a transvalvular gradient persistent throughout diastole, with a mean transvalvular gradient of 14.6 mm Hg. The estimated valve area was 0.9 cm² on direct planimetry of the restrictive orifice. Systolic pulmonary artery pressure was 45 mmHg.

Keywords: Mitral Facies, Mitral Stenosis, Rheumatic Heart Disease

Introduction

Mitral stenosis is a condition characterized by the narrowing of the mitral valve orifice due to structural abnormalities in the mitral valve apparatus^[1]. Rheumatic disease is the most common cause of MS worldwide. The prevalence of MS varies geographically, with fewer cases reported in industrialized countries (around 0.2–0.3 per 1000) compared to developing nations (approximately 5.5–5.7 per 1000)^[2]. Patients with MS may remain asymptomatic for years, but eventually, they experience a gradual decline in functional capacity. As the severity of MS worsens and left atrial pressure increases, symptoms such as exertional dyspnea (shortness of breath during physical activity), orthopnea (difficulty breathing while lying flat), and paroxysmal nocturnal dyspnea (sudden nighttime breathlessness) become more pronounced^[3]. In advanced stages, patients may also complain of symptoms related to right heart failure, including abdominal discomfort, ascites (fluid accumulation in the abdomen), and leg swelling^[4]. Recognizing clinical signs during physical examination is crucial. Some characteristic findings include a low-pitched rumbling diastolic murmur and an opening snap. Additionally, the presence of “mitral facies” serves as a clue to more severe disease. Mitral facies refers to the appearance of the patient’s face, which can be indicative of impaired cardiac output and pulmonary hypertension^[5]. Identifying this sign not only guides appropriate treatment but also helps prevent complications that can increase mortality and morbidity. Remember, echocardiography is the primary diagnostic modality for confirming MS and assessing its severity and anatomical consequences.

Case Report

A 42-year-old woman presented with sudden weakness of left side. Her medical history was rheumatic mitral stenosis 8 years ago. On physical examination was remarkable her rosy cheeks, whilst the rest of the face has a bluish tinge called Mitral Facies (Fig 1). It is the classic facial appearance in mitral stenosis, a plum-colored malar flush, occurs only when cardiac output is low and pulmonary hypertension is severe; cause is cutaneous vasodilation and chronic hypoxemia. In addition to cardiac auscultation was an irregularly irregular rhythm, a loud S1 and accentuated P2, an opening snap, and a III/IV decrescendo diastolic rumble at the apex. Electrocardiogram showed atrial fibrillation (Fig 2). Chest Xrays showed cardiomegaly with prominent pulmonary artery segment and large left atrium (Fig 3). Transthoracic echocardiography revealed findings

typical of rheumatic mitral stenosis. The mitral leaflets were pliable, with restricted mobility of the leaflet tips, resulting in doming and a “hockey-stick” appearance of the anterior leaflet in diastole. There was notable subvalvular thickening. Cross-sectional imaging of the stenotic orifice demonstrated thickening restricted to the leaflet tips. Continuous-wave

Doppler ultrasonography demonstrated a transvalvular gradient persistent throughout diastole, with a mean transvalvular gradient of 14.6 mm Hg. The estimated valve area was 0.9 cm² on direct planimetry of the restrictive orifice. Systolic pulmonary artery pressure was 45 mmHg (Fig 4).



Fig 1: Mitral facies

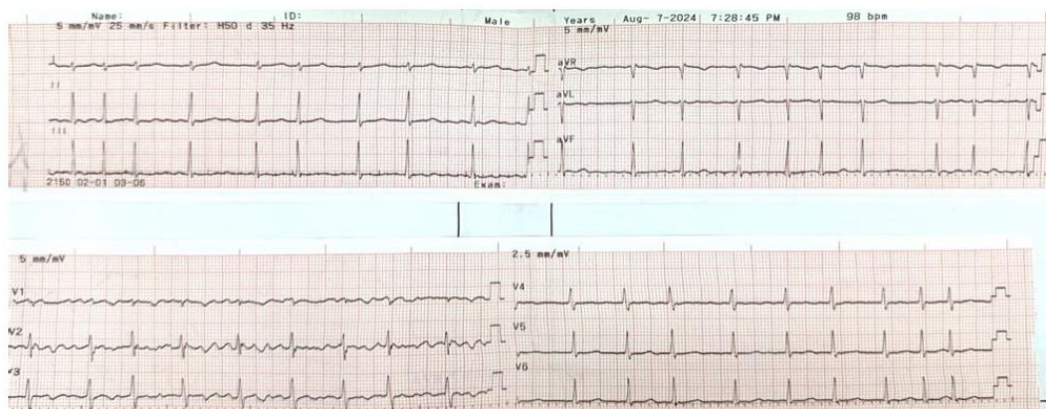


Fig 2: Electrocardiogram showed atrial fibrillation



Fig 3: Chest Xrays showed cardiomegaly with prominent pulmonary artery segment and large left atrium



Fig 4: Echocardiography showed Parasternal long axis view showed typical rheumatic mitral disease such as chordal and leaflet thickening of MV, hockey stick deformity of anterior mitral leaflet, and an enlarged LA

Discussion

The “mitral facies” is not an uncommon finding in MS, yet this sign is often under recognized. There is very limited literature on this classical sign. It is typically described as plethoric cheeks punctuated by bluish patches^[3]. Some also described it simply as pinkish-purple patches on the cheeks^[6, 7]. “Mitral facies” has become rare in the developed world because it is usually encountered in patients with long-standing untreated MS^[8]. This distinctive malar rash occurs in chronic severe MS with pulmonary hypertension^[6]. In this case, the patient’s symptomatic condition has occurred for the last 8 years and left untreated due to her socioeconomic status.

The “mitral facies” is quite similar to malar rash that can be caused by local and systemic disease such as systemic lupus erythematosus, erysipelas, rosacea, and pellagra^[9]. Systemic lupus erythematosus manifests as erythema, edematous, and sometimes purpuric rash with patchy macules, that usually precedes arthritis, fever, fatigue, and other debilitating autoimmune symptoms. In erysipelas, injury to blood vessels by streptococcal toxins produces a painful, well-circumscribed, erythematous, and shiny facial rash that is often accompanied by fever and malaise^[10]. Rosacea produces erythema, telangiectasia, and pustules or papules with episodes of flushing that can be triggered by hot drinks, stress, or alcohol. Dermatitis related to pellagra is a bilateral symmetric eruption on sun-exposed areas of the skin, that causes a painful, striking facial rash that usually involves the upper and lower extremities. In this patient, the facial rash is painless, more purplish in color, and less well demarcated with no triggering or precipitating factors. There are no accompanying systemic symptoms such as fever, malaise, or joint pain. The pathophysiology of mitral facies alone is not commonly described. Wood stated in his article that “mitral facies” was attributed to peripheral vasoconstriction secondary to low cardiac output. This reflex vasoconstriction causes peripheral cyanosis of the face or hands^[11]. He described that elevated pulmonary vascular resistance also contributed to make the cardiac output low and fixed. Atrial fibrillation (AF) and arterial oxygen saturation of less than 90% were also associated with this

facial rash. As MS severity worsens, flow restriction causing reduced diastolic filling, limits left ventricular output^[3]. AF further impairs this limited diastolic filling. LV filling may also further be impaired by RV pressure or volume overload causing abnormal septal motion. In severe MS, pulmonary vasoconstriction in addition to LA hypertension produces severe pulmonary hypertension^[3].

Conclusion

We present a case of a 42-year-old woman with severe mitral stenosis (MS) and regurgitation due to rheumatic heart disease. She exhibited clinical signs of right heart failure, including a distinctive malar rash—a potential marker of advanced MS. Echocardiography confirmed severe pulmonary hypertension and low cardiac output. The presence of mitral facies, characterized by flushed cheeks and bluish tinge, highlighted the chronic impaired cardiac output and pulmonary hypertension.

Conflict of interest

None.

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