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Lutembacher Syndrome: A Case Report

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Abstract **Case Report**

Lutembacher syndrome, first described in 1916 by French physician René Lutembacher, has undergone several definitions over the last few decades. However, the current consensus defines Lutembacher syndrome as any combination of atrial septal defect (congenital or iatrogenic) and mitral stenosis (congenital or acquired). We present the case of a 58-year-old woman in whom Lutembacher syndrome was discovered during the management of her right heart failure.

Keywords: Lutembacher syndrome, atrial septal defect, heart failure.

INTRODUCTION

Lutembacher syndrome is a rare clinical syndrome referring to the presence of mitral stenosis with coexisting atrial septal defect. It owes its name to French physician René LUTEMBACHER, who described his first case in 1916. This condition is usually treated surgically, with valve replacement and closure of the atrial septal defect. We present a case of Lutembacher syndrome in a 56-year-old female patient who was admitted to the hospital with right heart failure.

CASE REPORT

Mrs. F.O, 56 years old, was admitted to the cardiology department with right heart failure. She had a history of recurrent angina and joint pain. She was married with three children. She reported a dyspnea class II of NYHA classification, for 2 months, progressively worsening to stage IV with orthopnea associated with edema of the lower limbs.

Clinical examination revealed blood pressure 112/72 mmHg, heart rate 85 bpm, oxygen saturation 92%, cyanosis of the mouth. Signs of right heart failure were present, with jugular vein distention, hepatojugular reflux, and edema of the lower limbs. Auscultation revealed a diastolic rumble in the mitral area and a systolic murmur is heard over the precordium, mainly accentuated at the tricuspid area.

Her laboratory test results were normal. The electrocardiogram showed atrial fibrillation with a ventricular rate of 83 bpm, complete right bundle-branch block and secondary repolarization disorders (Figure 1).

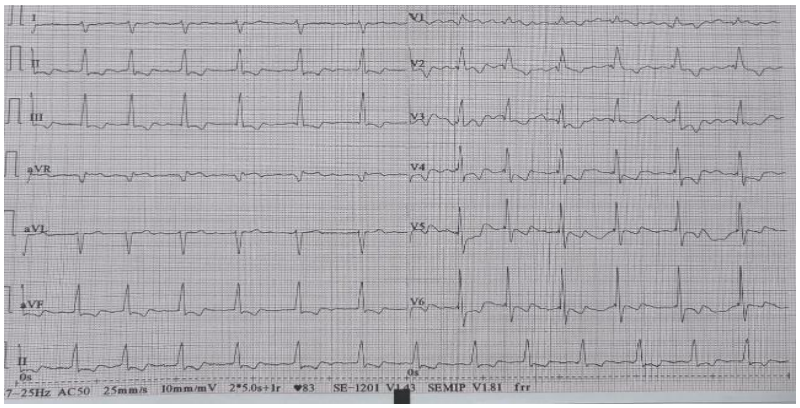

Figure 1: Complete atrial fibrillation arrhythmia with mean ventricular rate at 83 bpm, BBD with repolarization disorders

The frontal chest X-ray showed cardiomegaly with a cardiothoracic index of 0.62, and dilatation of the pulmonary artery. Two- dimensional echocardiography in apical view shows a 41mm wide OS-type atrial septal defect, tight mitral stenosis with a transmitral gradient of 5mmHg, a mitral valve area of 0.8 cm2, severe bi-atrial dilation, severe right ventricular dilation with hypokinesis, along with severe tricuspid regurgitation and severe pulmonary arterial hypertension at 73 mmHg. The left ventricle was of normal size with good systolic function EF: 75% (Figure 2, 3, 4, 5).

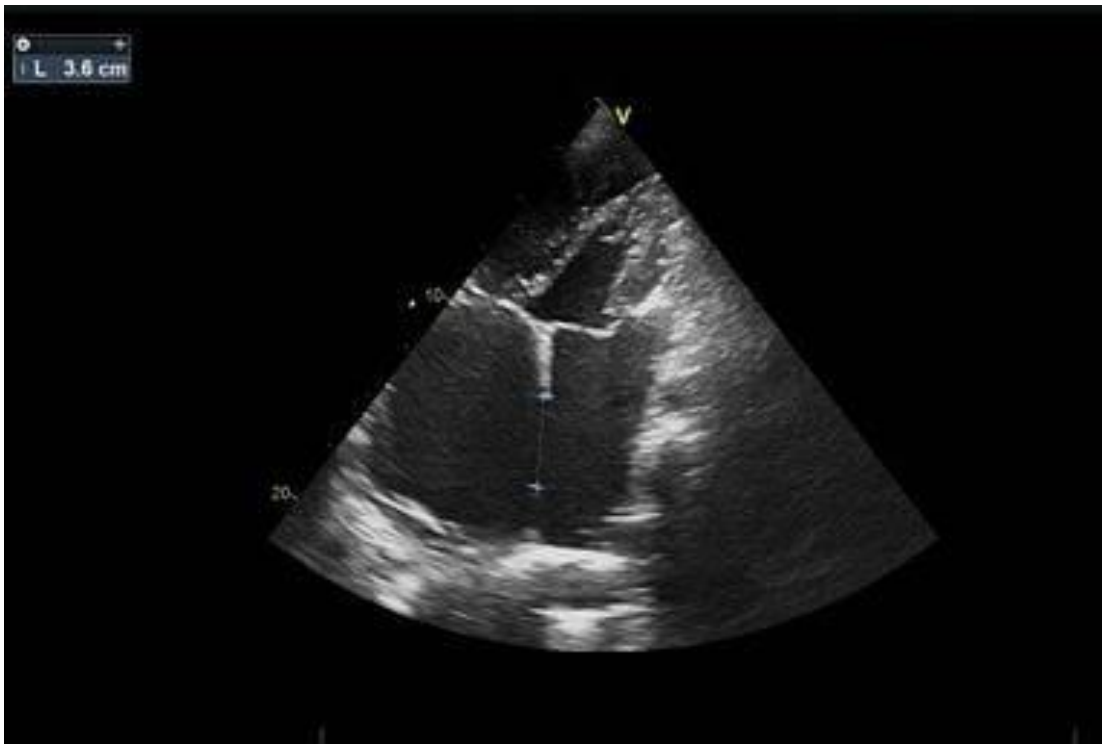

Figure 2: Two-dimensional four-chamber echocardiography image showing wide ASD and bi-atrial dilatation

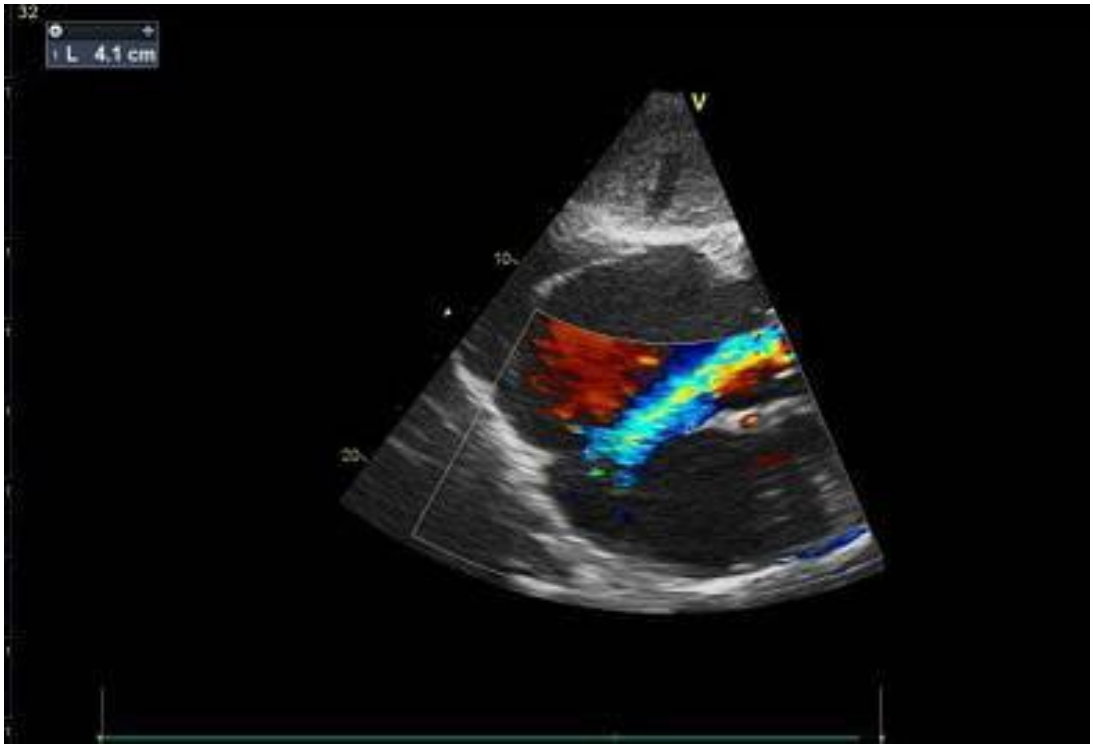

Figure 3: Two-dimensional color Doppler echocardiogram of the subcutaneous rib section showing the 41 cm wide ASD

Figure 4: Two-dimensional 4-cavity echocardiography image with mitral surface measurement

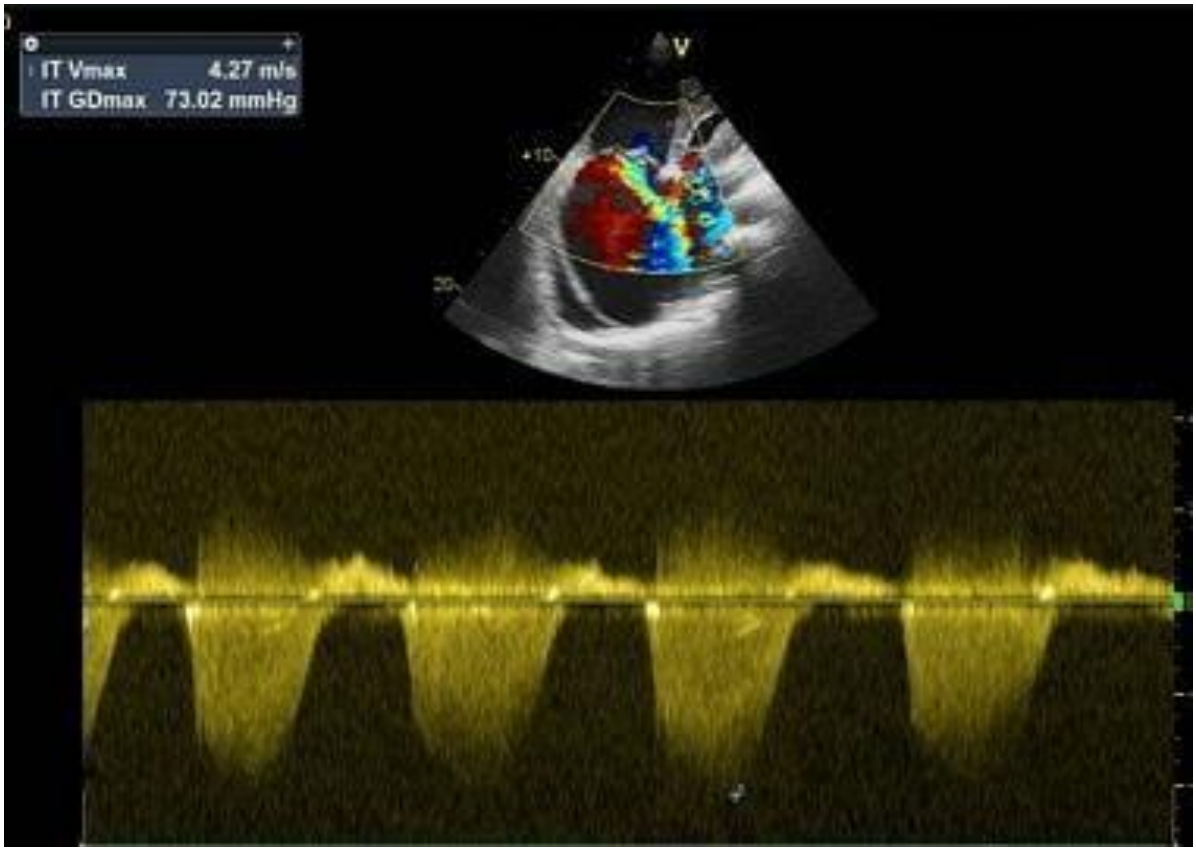

Figure 5: Two-dimensional echocardiography image showing the flow of tricuspid insufficiency

The patient had been receiving treatment for heart failure combined with anticoagulant therapy. At 10 days, peripheral signs of right heart failure had disappeared.

DISCUSSION

Both Lutembacher syndrome anomalies can be congenital or acquired. The incidence rate of congenital interatrial communication in patients with mitral stenosis is 0.6% to 0.7% [1]. Assuming that the incidence of interatrial communication is globally uniform; coexisting rheumatic mitral stenosis depends on the geographical prevalence of rheumatic disease. Lutembacher syndrome is well tolerated by patients over a long period; the hemodynamic effects of this syndrome result from the interaction between the relative effects of interatrial communication and mitral stenosis.

When mitral stenosis is severe and the interatrial septum is not restrictive, decompression of the left atrium occurs through the interatrial septum. As a result, the pressure in the left atrium does not increase proportionally to the severity of the mitral stenosis. Therefore, pulmonary venous hypertension takes a longer time to develop.

However, this results in an increase in the left-to-right shunt through the atrial communication and a progressive dilation of the right atrium and right ventricle [1]. This is the case with our patient, who remained asymptomatic until the age of 56. In her circumstance, atrial fibrillation decompensated this syndrome.

Two-dimensional echocardiography with color Doppler allows for the diagnosis of Lutembacher's syndrome; it assesses the severity of mitral stenosis as well as the size and type of atrial communication. The gradient across the mitral valve is lower despite severe mitral stenosis. The Doppler half-pressure time generally overestimates the surface area of the mitral valve orifice because the atrial communication relieves pressure in the left atrium and consequently reduces the transmitral gradient [2].

The natural progression of the syndrome is variable and depends on the size of the atrial communication and the severity of the mitral stenosis. The management of this syndrome is based on symptomatic treatment with beta-blockers and surgical intervention, traditionally through open-heart surgery, which used to be the gold standard. This procedure involves closing the septal defect and valve replacement, but nowadays, this technique is limited to large atrial communications [3]. Currently, given the therapeutic advancements and percutaneous interventional techniques, percutaneous transcatheter therapy in the form of balloon mitral valvuloplasty for mitral stenosis, followed by closure of the atrial communication device, has become the preferred treatment choice [4, 5].

In our case, due to the insufficient technical capabilities and lack of resources, our patient continued to be monitored in the department with a stable clinical condition.

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