

A RARE CASE OF ISOLATED SEPTUM PRIMUM ATRIAL SEPTAL DEFECT: PRESENTING AS NEW-ONSET ATRIAL FIBRILLATION IN A MIDDLE-AGED MALE

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Background: Atrial septal defects (ASD) of the septum primum type are typically diagnosed in infancy or childhood and are most often associated with atrioventricular valve malformations or syndromic conditions. We present the first reported case of an isolated septum primum ASD presenting as new-onset atrial fibrillation in a middle-aged male.

Case: A 41-year-old man with a history of asthma and tobacco use presented with sudden retrosternal chest pain and palpitations after alcohol intake. Electrocardiogram demonstrated atrial fibrillation with rapid ventricular response, which converted to sinus rhythm following intravenous diltiazem. Computed tomographic angiography ruled out pulmonary embolism but showed a 4 cm dilated main pulmonary artery and multichambered cardiac enlargement. Transthoracic echocardiography revealed LVEF 45-50%, severely enlarged right atrium and ventricle, moderate to severe tricuspid regurgitation, and reduced right ventricular systolic function. Right heart catheterization revealed a significant left-to-right shunt at the atrial level with a pulmonary-to-systemic flow ratio (QP: QS) of 3:1. Transesophageal echocardiography with bubble study confirmed a 1.9 cm superior-inferior isolated septum primum ASD without a mitral valve cleft or ventricular septum defect. Patient was discharged on metoprolol and apixaban and referred for surgical ASD closure.

Decision making: Septum primum ASDs are typically part of atrioventricular septal defects and frequently coexist with mitral valve clefts or ventricular septal defects. The absence of these associated abnormalities in this patient suggests a rare isolated septum primum defect. Chronic left to right shunting likely resulted in progressive right heart dilation, pulmonary artery enlargement, and atrial remodeling, predisposing the patient to new onset atrial fibrillation. Multimodality imaging with transthoracic echocardiography, transesophageal echocardiography, and right heart catheterization were essential for accurate diagnosis and hemodynamic assessment.

Conclusion: Septum primum ASD can present as an isolated defect, manifesting as atrial fibrillation in a middle-aged male. Early identification with echocardiography and right heart catheterization is vital to prevent progressive pulmonary hypertension and right heart failure.