Rupture sinus of valsalva and peripheral arterial disease: A rare combination
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Abstract
A 40 year old man admitted in cardiology department, NICVD, Dhaka with complaints of dyspnoea on mild exertion for two months. He was evaluated clinically and investigated with ECG, chest x-ray, Echo and angiography clinically. There was a continuous murmur over second and third intercostal spaces on the left sternal border, low volume pulse on the right lower limb than that of left. Echocardiogram revealed rupture sinus of valsalva into the right atrium. Oxymetry showed 15%, significant step up in the right atrium. Pulmonary and systemic blood flow ratio was 2.17:1. Left to right shunt was 54.1%. Pulmonary vascular resistance was 3.16 wood units. Right atrial systolic, diastolic and mean pressures were 25, 13, and 19 mmHg respectively. Root aortography confirmed rupture sinus of valsalva into right atrium. Coronary angiogram showed normal coronary arteries. Peripheral angiogram showed 90% stenosis in the proximal part of right common iliac arteries. He was diagnosed as a case of rupture noncoronary sinus of valsalva into right atrium with peripheral vascular diseases involving right common iliac artery.

Key words
Rupture sinus of valsalva, Right atrium, Peripheral vascular diseases, Right common iliac artery.

Introduction
Sinus of valsalva fistula is uncommon. It is most common in adults. Congenital failure of fusion of the aortic media with the fibrous skeleton of the heart at the aortic valve ring provides a point of weakness through which a sinus of valsalva aneurysm may develop. Aneurysm of the right coronary sinus is most frequent. Most of the rest protrude from the noncoronary sinus. Congenital aneurysms of the left coronary sinus are rare. In that location, infective endocarditis is a more frequent cause.

Because the root of the aorta is nearly surrounded by cardiac chambers, sinus of valsalva aneurysms may intrude on and may rupture into one of them. An aneurysm of the noncoronary sinus typically protrudes into the right atrium and usually ruptures into that chamber. Rarely recognized prior to rupture, sinus of valsalva aneurysm may be detected on imaging of a patient for some other purpose.

Rupture of a sinus of valsalva aneurysm usually results in a large shunt from the aorta to the right heart chambers. The patient presents with a continuous murmur and bounding arterial pulses. Often severe heart failure is present. With a large shunt, the heart failure is usually rapidly progressive and may result in death very quickly. The diagnosis is readily made from the clinical picture and echocardiography. Cardiac catheterization will confirm the level of the shunt. Because of the natural history, all patients should have this condition corrected surgically. Surgical results are usually quite good. In the large series, the operative survival was 96 percent with no late deaths in an average of 5.9 years of followup.

Peripheral arterial disease affects 10 to 15% of the adult population in developed countries. Arteriosclerosis obliterans is the
Review Article

The most common cause of lower extremity ischemic syndromes in western societies regardless of age. The presentation varies greatly with the time course of progression, the presence and extent of collateral vessels, co morbidities, and activity of the patient. In general, symptoms occur distal to the level of stenosis.

There is a relatively high prevalence of arteriosclerosis obliterans that increases with age. Risk factors for arteriosclerosis obliterans reflect those for coronary disease. The risk of death, usually from a cardiovascular event, increases dramatically as the ankle brachial index (ABI) decreases.

Aggressive risk factor modification should be the cornerstone of therapy in all patients. Smoking cessation is a must. Control of diabetes should be emphasized, since the risk for amputation is increased in this population, although the benefit has not been documented in large arteries. Lipid lowering has a beneficial role in patients with arteriosclerosis obliterans. The goals are similar to those for patients with coronary artery disease. Hypertension control should be optimized.

Case history

A 40 years old normotensive, non diabetic, dyslipidaemic, smoker male patient was admitted into cardiology department, NICVD, Dhaka with the complaints of sudden onset of shortness of breath for two months. There was no history of trauma, prolong fever. His pulse was 88/minute of large volume, regular in rhythm. Pulse volume on the right lower limb was low than that of left. Blood pressure was 120/60 mm Hg. There is a continuous thrill over second and third intercostal spaces on left sternal border. 1st and second heart sound was audible in all areas and was normal. There was a continuous murmur of grade 4/6 over same area.

Electrocardiogram shows normal findings. X-ray chest shows mild cardiomegaly. Echocardiogram revealtd rupture sinus of valsalva into the right atrium (Figure-1). Oxymetry showed oxygen saturation in the mid right atrium was 76% with significant step up (15%) in the right atrium. Pulmonary and systemic blood flow ratio was 2.1 7:1. Left to right shunwas 54.11%. Pulmonary vascular resistance was 3.16 wood units. Right atrial systolic, diastolic and mean pressures were 25,13and 19 mmHg respectively.

Root aortography showed rupture sinus valsalva into right atrium (Figure-2).

Coronaryangiogram showed normal coronary arteries with LVEDP 15 mm Hg. Peripheral angiogram showed 90% stenosis in the proximal part of right common iliac artery (Figure-3). He was diagnosed as a case of rupture noncoronary sinus of valsalva into right atrium with peripheral vascular diseases involving right common iliac artery.

Discussion

Sinus of valsalva fistula is uncommon and adults are most commonly affected. Aneurysm of the noncoronary sinus is less frequent. Congenital failure of fusion of the

The ORION Vol. 22 Sep 2005
aortic media with the fibrous skeleton of the heart at the aortic valve ring provides a point of weakness through which a sinus of valsalva aneurysm may develop. When aneurysm of the noncoronary sinus rupture, a fistulous connection between the aorta and right atrium results. Rupture of a sinus of valsalva aneurysm usually results in a large shunt from the aorta to the right heart chambers. The patient presents with a continuous murmur, bounding arterial pulses and high JVP. Often severe heart failure is present. In this case the murmur was loud and continuous, heard over second and third intercostal spaces on left sternal border. The apical impulse was hyperdynamic. Pulse was large volume and bounding.

In this case diagnosis was readily made from the clinical picture and color Doppler echocardiography. Cardiac catheterization confirmed the level of the shunt. In this case, oxymetry showed oxygen saturation in the mid right atrium was 76% with significant step up (15%) in the right atrium. Pulmonary and systemic blood flow ratio was 2.1 7:1. Left to right shunt was 54.11%. Pulmonary vascular resistance was 3.16 wood units. Right atrial systolic, diastolic and mean pressures were 25, 13 and 19 mmHg respectively. Root aortography showed rupture sinus valsalva into right atrium (Figure-2). Coronary angiogram showed normal coronary arteries with LVEDP 15 mm Hg. Peripheral angiogram showed 90% stenosis in the proximal part of right common iliac artery (Figure-3). With slow rupture and a small shunt, the major risk is infective endocarditis or extension of the rupture with an increasing shunt. With a large shunt, the heart failure is usually rapidly progressive and may result in death very quickly. A few patients seem to stabilize in this situation.

Because of the natural history, all patients should have this condition corrected surgically. Surgical results are usually quite good. In this patient for right common iliac artery surgical revascularization may be considered. Large vessel bypass surgery with synthetic graft material is well established and durable. More recently, in situ distal bypass utilizing reversed or intact saphenous vein has shown promising long-term patency. Percutaneous balloon angioplasty with or without stent placement is often useful for lesions of the proximal renal and iliac arteries.

Conclusion
Rupture sinus of valsalva is a rare disease usually present in young and peripheral arterial disease is a disease of elderly people usually associated with coronary artery disease. In this young patient presents with rupture sinus of valsalva also have an isolated single peripheral arterial disease, a rare combination, may be missed without meticulous clinical examination.

References