Acquired aneurysm of the left sinus of Valsalva and coexistence of coronary artery disease with hypereosinophilia in a young menstruating woman: A case report

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A 35-year-old menstruating woman presented with exertional angina, and was diagnosed with an unruptured left sinus of

Valsalva and a noncoronary sinus of Valsalva aneurysm. Hypereosinophilia and coronary artery occlusion was also observed; this association is extremely rare.

Key Words: Hypereosinophilia; Sinus of Valsalva aneurysm

Aneurysms of the sinus of Valsalva are rare cardiac anomalies. Most of these aneurysms (approximately 95%) originate from the right and noncoronary sinuses. Left sinus aneurysms are rare (less than 5%). Congenital aneurysms are more prevalent than acquired aneurysms, and most of these involve the noncoronary and right sinus of Valsalva (99%) (1). Acquired sinus of Valsalva aneurysms may arise from any one of or involve all three sinuses (2). Sinus of Valsalva aneurysms are associated with Marfan syndrome, atherosclerosis, syphilis, infective endocarditis, cystic medial sclerosis, chest trauma and hypereosinophilic syndrome (3,4). Sinus of Valsalva aneurysms are usually found incidentally during diagnostic procedures. Some patients may have compression symptoms or thromboembolic phenomena (5).

CASE PRESENTATION

A 35-year-old, menstruating woman was admitted to the Kasturba Hospital, Manipal, India, with a six-month history of anginal chest pain and dyspnea during exertion (New York Heart Association class II). There was no history of dyslipidemia, diabetes or hypertension, or any conventional coronary risk factors. Physical examination revealed a thin, normotensive woman. Cardiac examination revealed a palpable pulsation in the second, third and fourth left intercostal spaces. Grade I/III left parasternal heave was present. Auscultation revealed normal first and second heart sounds, and a grade 2/6 high-pitched pansystolic murmur at the apex, radiating to axilla. Respiratory system and abdominal examination did not reveal any abnormality.

Figure 1) Peripheral blood smear (original magnification $\times 100$) showing high eosinophil count

Hematological evaluation showed hemoglobin 137 g/L, total white blood cell count $25 \times 10^9/L$, differential count (polymorph 23.71%, lymphocytes 12.5%, monocytes 2.8%, eosinophil 61.3%) and erythrocyte sedimentation rate 30 mm/h. Absolute eosinophil count was $12.98 \times 10^9/L$ and a peripheral blood smear revealed 59% eosinophils, 30% polymorphs and 10% lymphocytes (Figure 1). Electrocardiogram showed sinus rhythm and left ventricular hypertrophy with strain pattern (Figure 2). A posteroanterior chest x-ray showed a prominent

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Figure 2) Electrocardiogram showing left ventricular hypertrophy with strain pattern

Figure 3) Chest x-ray (posteroanterior view) showing prominent left hilum and straightening of the left heart border

left hilum and straightening of the left heart border (Figure 3). In view of the patient's chest pain and left ventricular hypertrophy on electrocardiogram, a transthoracic echocardiogram was performed, and revealed an unruptured left sinus of Valsalva aneurysm (5.8 cm × 3.8 cm) in the short-axis view, as well as trivial aortic regurgitation, moderate mitral regurgitation, mild pulmonary arterial hypertension and fair left ventricular systolic function (ejection fraction 51%) (Figure 4). Transesophageal echocardiogram confirmed these findings, and dilation of the aortic root (Figure 5).

Aortic root angiogram in anteroposterior lateral, right anterior oblique and left anterior oblique views (Figures 6 to 8) showed a hugely dilated left sinus of Valsalva with blood stasis. The noncoronary sinus was also dilated and no significant aortic regurgitation was observed. The right coronary artery appeared nondominant with proximal total occlusion. The left

Figure 4) Transthoracic echocardiogram in parasternal short-axis view shows an aneurysm of left sinus of Valsalva, a mildly dilated noncoronary sinus and a normal right sinus of Valsalva

Figure 5) Transesophageal echocardiogram confirms aortic root dilation with an unruptured left sinus of Valsalva aneurysm

coronary artery was not visualized due to the distorted and dilated left sinus of Valsalva.

To rule out associated vasculitis or infectious etiology, C-reactive protein, rheumatoid factor, antinuclear antibody, anti-double-stranded DNA antibody, anti-Sm antibody, anti-SS-A/SS-B antibody, anti-U1 RNP antibody and *Treponema pallidum* hemagglutination assays were performed, and all were negative. The cytoplasmic-staining antineutrophil cytoplasmic antibodies (cANCA) test was weakly positive. Because the left coronary artery was not clearly delineated, computed tomography angiography was planned to assess this before operative intervention, but the patient was subsequently lost to follow-up.

DISCUSSION

In young women, coronary artery disease in association with left sinus of Valsalva aneurysm is extremely rare. Our patient also had a high eosinophil count (absolute eosinophil count

Figure 6) Aortic root angiogram in anteroposterior view with pigtail catheter showing left sinus of Valsalva aneurysm on the left side and the noncoronary sinus aneurysm on the right side of the ascending aorta

12.98 $\times 10^9$ /L). In patients with hypereosinophilic syndrome, cardiovascular manifestation is severe, and results in high morbidity and mortality. Typical cardiovascular manifestations are endomyocardial fibrosis with subsequent cardiac failure, and thrombosis. Eosinophilic aortitis and valvulitis are also observed (6), and may affect the coronary sinuses of Valsalva and the coronary arteries.

A report by Okinaka et al (3) observed a left sinus of Valsalva aneurysm and coronary artery aneurysm associated with hypereosinophilic syndrome. Histology of the left sinus of Valsalva aneurysm revealed eosinophil infiltration along the initial and inner part of the aortic media. An aortic root angiogram of our patient revealed a total occlusion of the right coronary artery. Total right coronary artery occlusion in association with left sinus of Valsalva aneurysm and hypereosinophilic syndrome has not previously been reported. The aortic valve is typically spared in hypereosinophilic syndrome despite the root pathology, as was observed in our case.

Because our patient was a young, menstruating woman without conventional coronary artery disease risk factors, an atherosclerotic cause was unlikely for her right coronary artery occlusion; therefore, hypereosinophilia is the most likely cause. The cANCA was weakly positive, but this patient did not have symptoms suggestive of disorders that are associated with eosinophilic vasculitis (eg, Churg-Strauss syndrome, polyarteritis nodosa, temporal arteritis). Forty-eight per cent of patients with Churg-Strauss syndrome are positive for ANCA, usually perinuclear ANCA. Rarely, patients with polyarteritis nodosa may be positive for perinuclear ANCA

Figure 7) Aortic root angiogram in right anterior oblique view with pigtail catheter showing an enlarged aneurysmal left sinus of Valsalva (visualized anteriorly) and noncoronary sinus (visualized posteriorly)

Figure 8) Aortic root angiogram in left anterior oblique view with pigtail catheter showing an enlarged aneurysmal left sinus of Valsalva (visualized anteriorly) and noncoronary sinus. Left coronary sinus is visualized anteriorly and noncoronary sinus is visualized inferiorly. The right sinus of Valsalva is normal, and the occluded right coronary artery stump can be seen

or cANCA. Vasculitis is not a prominent feature of hypereosinophilic syndrome. Hence, we can reasonably conclude that hypereosinophilia was responsible for our patient's sinus of Valsalva aneurysm and right coronary artery occlusion.

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