COMUNICACIONES BREVES

Spontaneous rupture of an aneurysm of the sinus of Valsalva into the right atrium, associated with an atrial septal aneurysm

Pedro López-Velarde B,1 Nydia Avila-Vanzzini,2 Tatiana López-Velarde Peña,4 Alejandro Dabaghi-Richerand,4 Fernando López-Soriano,3 Nilda Espinola-Zavaleta.2

1Cardiovascular Hemodynamic Department, American British Cowdray Medical Center, Mexico City.
2Cardiovascular Echocardiography Department, Instituto Nacional de Cardiología Ignacio Chávez and The American British Cowdray Medical Center, Mexico City.
3Cardiothoracic Surgery Department, Instituto Nacional de Cardiología Ignacio Chávez and The American British Cowdray Medical Center, Mexico City.
4Medicine School, Universidad La Salle, Mexico City.

Received on November 26, 2008; accepted on March 29, 2010.

KEYWORDS
Sinus of Valsalva; Atrial septal aneurysm; Spontaneous rupture; Association of two isolated malformations; Echocardiography; Aortography; Mexico.

Abstract
Aneurysms of the sinus of Valsalva (SV) and the atrial septum are a rare association. We report the case of a 28-year-old woman, who was admitted to our department complaining of progressive dyspnea of 10 days of evolution, five hours previous to her admission to the hospital; she presented sudden oppressive anterior chest pain, accompanied by palpitations. The presence of rupture of the right SV to the right atrium was clinically confirmed, by echocardiography and hemodynamic studies. In addition, an associated atrial septal aneurysm was found. She underwent surgical correction through sinusplasty without requiring aortic valve replacement. The patient presented persistent postoperative atrioventricular block, which required a permanent pacemaker. Clinical evolution was satisfactory. To our knowledge, this case is a rare combination of two isolated malformations, without previous events that could explain the rupture of the right SV.
Spontaneous rupture of sinus of Valsalva aneurysm to right atrium

Ruptura espontánea de un aneurisma del seno de Valsalva a la aurícula derecha en una paciente con aneurisma del septum interatrial

Resumen
Los aneurismas del seno de Valsalva y del septum interauricular son una asociación rara. Informamos el caso de una mujer de 28 años de edad con cuadro de disnea progresiva en los últimos 10 días, al cual se agregó dolor precordial opresivo, cinco horas previas a su ingreso. Se comprobó clínicamente, por ecocardiografía y hemodinamia la presencia de ruptura del seno de Valsalva derecho hacia el atri derecho. Un hallazgo interesante fue la presencia de un aneurisma del septum interauricular asociado. La paciente fue sometida a corrección quirúrgica con plástia del seno de Valsalva, sin requerir reemplazo valvular aórtico. En el postoperatorio presentó bloqueo auriculo-ventricular persistente, requiriendo implante de marcapaso definitivo. Su evolución fue satisfactoria. Este es un caso de una rara asociación de dos malformaciones aisladas.

Case report
A 28-year-old woman was referred to our department, she had a history of 10-day progressive dyspnea, five hours previous to her admission at the hospital, and she presented suddenly oppressive anterior chest pain without irradiations, accompanied by palpitations at rest and orthopnea. Physical examination showed heart rate (HR) of 110 beats/min, blood pressure (BP) of 120/40 mmHg and the jugulars veins were distended. There was a grade 4/6 continuous murmur along the left parasternal line without thrill. There was also hepatomegaly 2-2-1. The electrocardiogram showed tachycardia and incomplete right bundle branch block. An increase of pulmonary vascula-

The two-dimensional Doppler echocardiogram documented a short aneurysmal dilatation of the right sinus of Valsalva, protruding into the right atrium. The place of rupture was shown with color flow imaging, revealing a unidirectional continuous mosaic jet from the aorta to the right heart chamber, (Figure 1A, 1B). In a four chamber view with posterior angulation, an interatrial septal aneu-

Aneurysm of the sinus of Valsalva (ASV) is a rare cardiac abnormality, occurring in 0.15% to 1.5% of patients who undergo cardiopulmonary bypass. They may be congenital or acquired. A congenital aneurysm is caused by separation or failed fusion of the aortic media layer with the fibrous annulus of the aortic valve. Acquired aneurysm of sinus of Valsalva can develop as the result of traumatic injury; endocarditis, syphilis, Behcet’s disease or Marfan’s syndrome. The first reports of ASV appeared in the 19th century and Lillehei et al. reported the first successful surgical repair in 1957.

Aneurysms are generally silent for prolonged periods of time. Regarding complications, the most frequent occurrence is rupture. It occurs in most cases between the third and fourth decades of life. The clinical presentation of this entity varies considerably due to the fact that it can be asymptomatic. It can be found in a postmortem or angiographic study, or they present with cardiogenic shock and death. This wide variety in initial clinical presentations can be due to the size of the shunt; little shunts are asymptomatic, and big aorticcardiac fistulas cause a clinical presentation similar to an acute aortic regurgitation.

There are numerous complications that can originate from a Valsalva aneurysm, including obstruction of the right ventricular outflow tract, infectious endocarditis, and thrombus formation, with systemic or local embolic events. The compression of the origin of the coronaries or the obstruction of their ostia can cause ischemia or necrosis. Ischemia has been reported as a conditioning factor for ventricular fibrillation in some patients. Whereas, in others, ventricular fibrillation is secondary to the dissection of the interventricular septum due to a ruptured aneurysm at this site. In this case, complete atrioventricular block usually accompanies the clinical presentation.
Aneurysms are most frequently localized at the right sinus of Valsalva (76.8%) and at the non-coronary sinus (20.2%). Left sinus of Valsalva localization is not very frequent (3%). The least frequent sites are the pericardium, pleural space, interventricular septum, or the left ventricle chambers.

Eventhough the first report of a Valsalva aneurysm diagnosed by echocardiography was in 1974, currently the gold standard for the diagnosis of these lesions continues to be cardiac catheterization with aortography. Development of new generation ultrasonography machines has made transthoracic and, especially, the transesophageal echocardiography a useful tool in the confirmation of the diagnosis. Additionally, it may help in the differential diagnosis of other abnormalities causing continuous murmurs like patent ductus arteriosus, aorto-pulmonary window, or coronary fistulas. Nuclear magnetic resonance imaging is equally useful, but more expensive and less available than echocardiography. The latter is a better and faster diagnostic tool, especially when dealing with critically ill patients.

Congenital sinus of Valsalva aneurysms can coexist with other malformations; the most common of which is the association with subaortic ventricular septal defect (25-55%) and aortic regurgitation. Other associated abnormalities less frequently found are pulmonary stenosis, patent ductus arteriosus, atrial septal defect, subaortic stenosis, and Fallot’s Tetralogy.

Regarding the atrial septal aneurysm, this is an infrequent finding in adult patients. Its formation may be secondary to raised interatrial pressure gradients, producing a bulging septal shift toward the low-pressure side.
Spontaneous rupture of sinus of Valsalva aneurysm to right atrium

However, it has been found also in patients with normal atrial pressure,\(^1\) suggesting a primary malformation.\(^1\)

The coexistence of sinus of Valsalva aneurysms and atrial septal aneurysms is very rare. Atrial septal aneurysm is believed to be a disorder of connective tissue,\(^1\) which is manifested along time in adults. In fact, connective tissue abnormalities must be suspected when both defects coexist together. Replacement of the valve was not considered necessary in the absence of aortic valve dysfunction. Regarding the atrioventricular block, this was temporal, because in a follow-up revision of the patient, she had sinus rhythm, alternating with pacemaker rhythm (data not shown). We consider that the presence of this block was secondary to the inflammatory process of the surgery.

The present case illustrates the rare coexistence of two congenital malformations, as are sinus of Valsalva aneurysm and aneurysm of the interatrial septum.

References