Case Reports

Ruptured Sinus of a Valsalva Aneurysm Associated with Autosomal-Dominant Polycystic Kidney Disease in an Elderly Patient: Report of a Case

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Abstract We report herein the case of a 71-year-old woman with autosomal-dominant polycystic kidney disease (ADPKD), who was referred to our hospital for investigation of facial edema. Echocardiography demonstrated a large aneurysm arising from the non-coronary sinus of Valsalva, with a left to right shunt and jets of blood passing from the aneurysm toward the septal leaflet of the tricuspid valve. Surgical treatment was successfully carried out by resecting the aneurysmal wall and performing a patch closure of the orifice. If is well known that ADPKD predisposes patients to cardiovascular disease, and this case report serves to demonstrate that when a patient with ADPKD presents with progressive heart failure, the possibility of a ruptured sinus of a Valsalva aneurysm must be considered.

Key Words Rupture · Sinus of Valsalva · Aneurysm · Autosomal-dominant polycystic kidney disease

Introduction

Autosomal-dominant polycystic kidney disease (ADPKD) is a systemic hereditary disorder with both renal and extrarenal manifestations, characterized by localized cellular proliferation and extracellular matrix abnormalities. An increasing number of patients without any symptoms of kidney disease are now being diagnosed when investigated for hypertension, or incidentally during radiological studies of the abdomen.

This case report describes our experience with an elderly patient diagnosed as having ADPKD during detailed examinations for a ruptured sinus of Valsalva aneurysm (SVA). This combination is so rare that we were only able to find one other such case documented so far.1

Case Report

A 71-year-old Japanese woman who was receiving treatment for hypertension was referred to our hospital for investigation of facial edema and general fatigue. Systemic edema had gradually developed since she had experienced transient chest pain 7 months prior to the examination. On admission, a grade 3/6 systolic murmur was heard, which was loudest over the apex. Ascites and hepatomegaly were noted on palpation. A chest roentgenogram demonstrated cardiomegaly with a cardiothoracic ratio of 70%, along with pleural effusion. Her blood pressure was 120/40 mmHg, and her pulse rate was 78 beats/min and regular. The blood urea nitrogen level was 44 mg/dl, the serum creatinine level was 1.8 mg/dl, and creatinine clearance was 21 ml/min. Precordial echocardiography showed a large aneurysm arising from the noncoronary sinus of Valsalva (Fig. 1), with a left to right shunt and jets of blood passing from the aneurysm toward the septal leaflet of the tricuspid valve. No aortic regurgitated flow or any ventricular septal defects were detected. Right and left cardiac catheterization was performed and oximetric recordings showed an oxygen saturation increase of 13% at the level of the right atrium. Hemodynamic measurements demonstrated a mean right atrial pressure of 19 mmHg. The right ventricular systolic pressure, the end-diastolic pressure, and the pulmonary arterial systolic pressure were 58, 28, and 61 mmHg, respectively. The calculated pulmonary to systemic flow rate was 4.0. An aortogram demonstrated a large fistula connecting the noncoronary sinus of Valsalva to the right atrium (Fig. 2), with a normal-sized aortic root and no aortic regurgitation. A left ventriculogram showed normal wall motion and no signs of mitral regurgitation. As the patient had a
familial history of polycystic kidney disease, further detailed examinations were undertaken. Abdominal computed tomography showed multiple cysts in the liver and kidneys (Fig. 3) and brain magnetic resonance angiography revealed multiple intracranial aneurysms. Finally, a diagnosis of a ruptured SVA associated with ADPKD was made. She was initially treated with diuretics and dopamine, which achieved some symptomatic improvement.

Surgery was performed through a median sternotomy. After inducing cardiac arrest by the retrograde infusion of blood cardioplegic solution, the ascending aorta and right atrium were incised, revealing a large fistula originating in the noncoronary sinus of Valsalva. A windsock-like aneurysm 15 × 10 mm in size was also found, located just above the anteroseptal commissure of the tricuspid valve with a 5-mm opening at its distal end. After it was confirmed that the fistula was not connected to the other chamber, the aneurysmal wall was resected and the defect was closed through the right atrium with a 2-ply bovine pericardial patch using pledgeted polypropylene sutures, avoiding any distortion of the valves. The patient was easily weaned from cardiopulmonary bypass into sinus rhythm. Her postoperative course was uneventful and renal function was well maintained. Histological analysis of the aneurysm showed myxomatous degeneration with loss and disruption of collagen.

**Discussion**

ADPKD is the most common cystic disease of the kidney, with an estimated prevalence ranging from 1:400 to 1:1000. The systemic manifestations include cystic and noncystic abnormalities in both renal and extrarenal sites. Two, or possibly three, causative genes located on chromosomes 4 and 16 have been identified. A gene defect mapped to chromosome 16p13.3 accounts for 90% of cases of the disease and the abnormal proteins produced lead to decreased tensile strength of the basement membrane.