AN UNUSUAL CASE OF RUPTURED ANEURYSM OF SINUS OF VALSALVA

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ABSTRACT

Ruptured congenital aneurysms of the noncoronary aortic sinus shunting into both the right atrium and right ventricle are extremely rare. We present here such an anomaly in a 40-year-old man, focusing on the diagnostic reliability of echocardiography and the unusual angiographic features of the aortic sinus aneurysm in this patient.

Keywords: *fistula, sinus of Valsalva aneurysm, right atrium, right ventricle*

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INTRODUCTION

Congenital aneurysm of sinus of Valsalva is a rare entity due to an intrinsic weakness and deficiency of the media at the junction of the ascending aorta and the heart, with subsequent aneurysmal formation[1,2]. The aneurysm may then rupture into any of the cardiac chambers[1-5]; this is commonly from the aneurysmal right coronary sinus into the right ventricle in Orientals[3,4]. We report here an unusual case of aneurysm of the noncoronary sinus rupturing into both the right atrium and right ventricle.

CASE REPORT

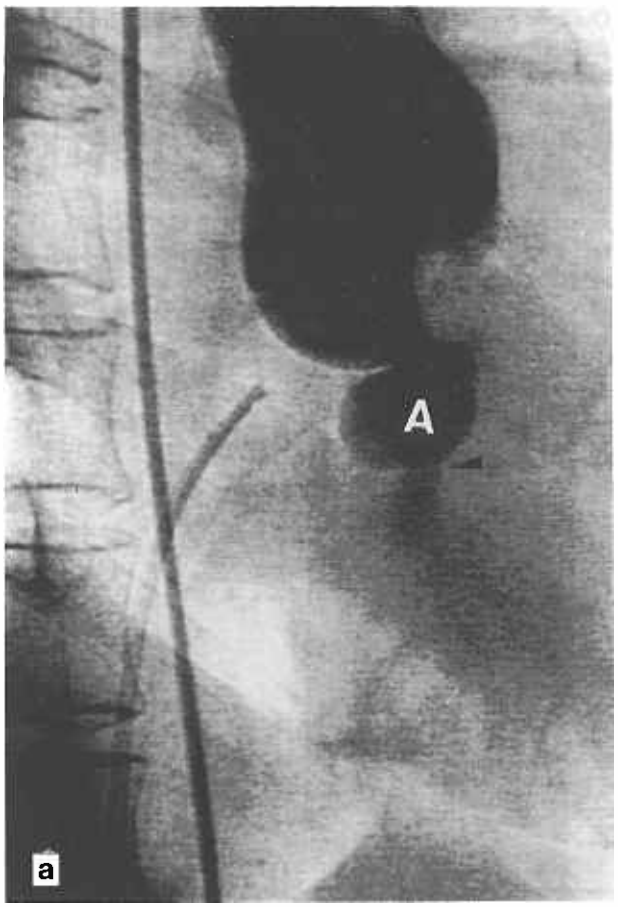
A previously asymptomatic fit-looking 40-year-old Chinese man presented with exertional dyspnoea and decreased effort tolerance of recent onset. There was no history of chest discomfort.

Physical examination revealed a widened pulse pressure as evidenced by a blood pressure of 120/55 mmHg and collapsing pulse. The jugular venous pressure was not elevated. The apical impulse was noted to be hyperdynamic but not displaced. A grade 4/6 continuous murmur accompanied by a thrill along the left sternal edge was detected.

The baseline ECG showed a right ventricular hypertrophy and strain pattern and the chest X-ray revealed mild cardiomegaly with pulmonary plethora. Transthoracic echocardiographic evaluation demonstrated a tricuspid aortic valve with a large "wind-sock" abnormality of the noncoronary sinus of Valsalva projecting into the right atrium. Doppler echocardiography showed a continuous flow pattern emanating from a defect (representing the site of rupture) at the tip of aneurysm into the right atrium. There were no other significant echocardiographic findings, in particular, no associated congenital heart defect was observed.

During right heart oximetry study, a significant step-up in oxygen saturation localised at the midatrial level was detected. The calculated Qp:Qs ratio was 3:1. Aortography supported previous echocardiographic findings; a large aneurysmal sac arising from the noncoronary sinus and bulging into the right atrium was clearly visualised. In addition, the aneurysm with its jet of contrast medium was observed to swing back and forth between the right atrium and the right ventricle across the tricuspid valve (Fig 1a, b). Coronary angiography showed no evidence of coronary artery disease. The aneurysm of the noncoronary sinus of Valsalva with fistulisation into the right cardiac chambers was verified during surgery and excised.

Fig 1 - Freeze frame angiographic views in the right anterior oblique projection displaying the aneurysm of noncoronary sinus of Valsalva (A) rupturing (arrow head) into the right atrium in systole (a) and prolapsing across the tricuspid valve into the right ventricle during diastole (b).



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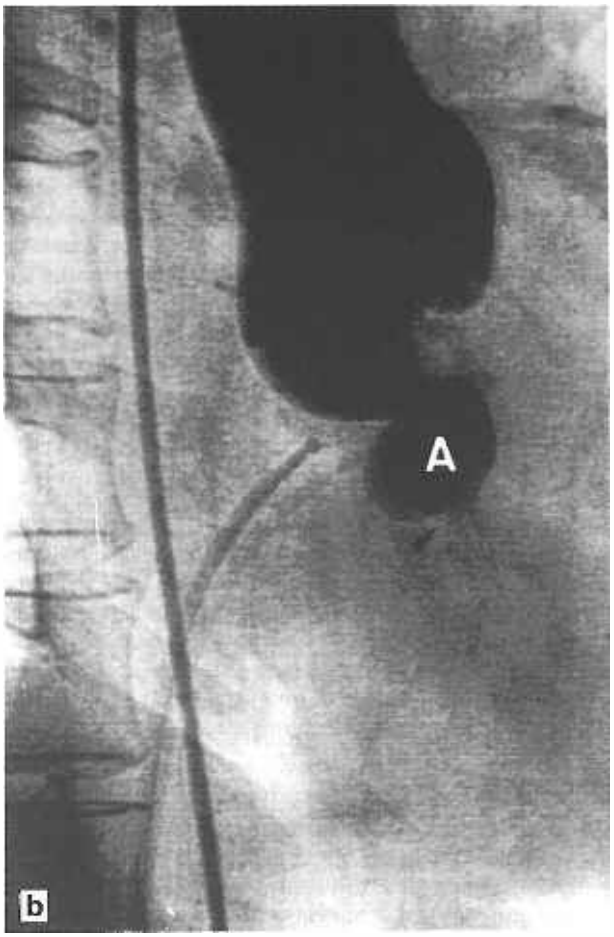

DISCUSSION

Our case highlights (1) the pivotal role of echocardiography in the diagnosis and treatment strategy of patients with ruptured aneurysm of sinus of Valsalva (RASV), and (2) certain unusual angiographic features of the RASV in our patient.

In the past, cardiac catheterisation was considered mandatory for the accurate diagnosis of RASV and the identification of potential associated congenital cardiac lesions such as ventricular septal defect, aortic regurgitation from aortic valve prolapse or bicuspid aortic valve, and pulmonary stenosis. However, the advent of 2-dimensional and Doppler echocardiography has allowed a noninvasive, reliable and more widespread means of diagnosing RASV (as demonstrated in our patient) and its associated cardiac anomalies [5]. These capabilities, together with increased operator experience and confidence in echocardiography have obviated the need for routine cardiac catheterisation as a diagnostic tool in this condition. Thus, the current role of the latter investigatory procedure should be limited to selected patients requiring further evaluation of other associated cardiac anomalies when these are not well-defined by echocardiography alone [5], and/or importantly, for the exclusion of concomitant significant coronary artery disease which may require bypass grafting during surgical repair of the RASV. In our patient, because of the increased risk of possible underlying coronary artery disease due to his age and gender, cardiac catheterisation, particularly coronary arteriography, was performed for delineation of the coronary anatomy.

Isolated RASV, in itself a rare anomaly, usually involves the right coronary sinus shunting into the right ventricle [1-4]. This is more true in Orientals compared with the Western population [3,4]. In a large comparative analysis of RASV in Oriental and Western patients, Chu et al [4], found in Orientals a significantly higher incidence of RASV involving the right aortic sinus (88% vs 63%) and an extremely low incidence of aneurysms originating from the noncoronary sinus or rupturing into the right atrium or right ventricle. Our patient had a ruptured aneurysm of the noncoronary sinus of Valsalva which, interestingly, was so large that it was able to swing from the right atrium across the tricuspid valve into the right ventricle and back, in syncrony with the cardiac cycle (Fig 1). This latter phenomenon is even more unusual[1-5].

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