RIGHT VENTRICULAR MYXOMA SIMULATING PULMONARY STENOSIS

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SUMMARY : A case of right ventricular myxoma simulating pulmonary stenosis is reported. The ultimate diagnosis was reached by echocardiography and magnetic resonance imaging which revealed large filling defects in the right ventricle. The tumor was successfully excised through a right atrial approach. This approach provides adequate surgical exposure, avoids undue trauma to the ventricular myocardium, and offers an easy way for inspection of the cardiac chambers.

Key Words : *Right Ventricle, Myxoma, Magnetic Resonance Imaging.*

INTRODUCTION

Myxomas are the most common cardiac tumors in adult patients, but their localization in the right ventricle is quite unusual. Currently tumors of the right ventricular cavity can be clearly identified by new diagnostic tools such as echocardiography and nuclear magnetic resonance. A case with right ventricular myxoma simulating pulmonary stenosis diagnosed noninvasively is presented.

CASE REPORT

A 26 year old soldier was admitted to the Gülhane Military Medical Academy in May 1995, for evaluation of marked exertional dyspnea. Three years previously, a cardiac murmur had been detected, but no therapy or further investigation was contemplated, since the murmur was considered to be functional. On admission, physical examination revealed a blood pressure of 120/70 mmHg and a regular pulse rate of 80 beats per minute. Noteworthy auscultatory findings were the attenuated second heart sound and a protosystolic click followed by a harsh 3/6 ejection murmur on the pulmonary area. The electrocardiogram showed sinus rhythm and incomplete right bundle branch block. On the chest roentgenogram, only mild enlargement of the pulmonary artery shadow was noted. Routine laboratory findings were within normal limits. 2-D echocardiogram showed clouds of diastolic echoes in the right ventricular cavity and the right ventricular outflow tract. There was a pulmonary infundibular gradient of 45 mmHg. Magnetic resonance imaging (MRI) demonstrated a large mass in the right ventricle and extending into the pulmonary artery (Fig. 1a, b). Operation was performed through a median sternotomy with the aid of cardiopulmonary bypass, mild systemic hypothermia, and cold cardioplegic arrest. The right atrium was opened, the tricuspid valve was gently retracted, and part of the tumor was visualized. It was inserted by a short stalk to the free wall of the right ventricle and obstructed the right ventricular outflow tract almost completely. By investigating the infundibular free

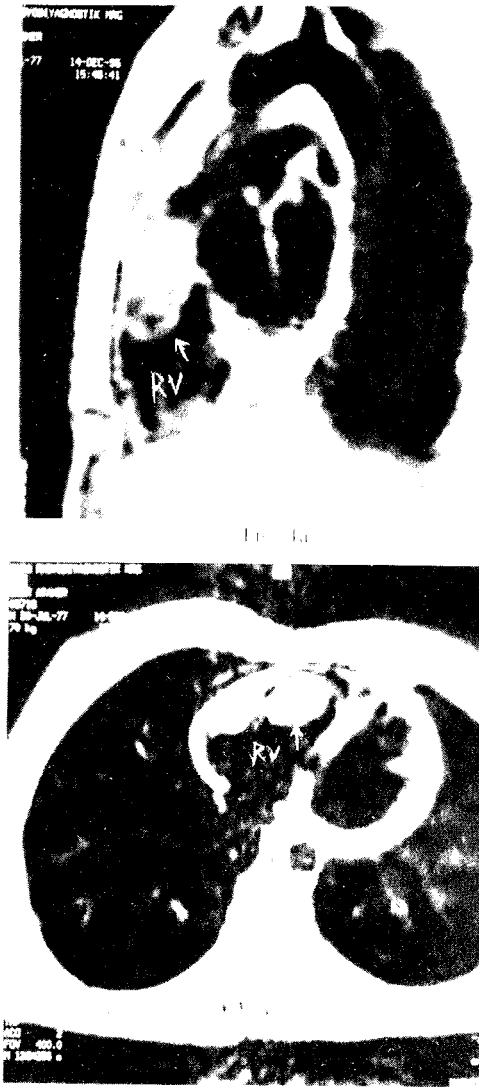

Fig - 1a, b : Preoperative magnetic resonance imaging (a: sagitall b: horizantal section) showed the large mass (arrowheads) in the right ventricle and prolapsing through the pulmonary valve. RV : right ventricle.

wall with external finger compression, the tumor was easily dislocated inside the right atrium and completely excised together with its base of implant. Subsequently, the fossa ovalis was opened and, on retraction of the interatrial septum, the left atrium and the mitral valve was inspected in order to rule out the coexistence of left-sided myxomas. The postoperative course was uneventful. To day, five months after operation, the patient is doing well.

The tumor was a sessile, lobuled, and polypoid mass with a smooth surface; it weighed 20 gm and measured 3 x 3,5 cm. Ultrastructural examination revealed the classic, very undifferentiated "myxoma cells" with rare cytoplasmic organelles. Cells surrounding the vessellike structures had cytoplasmic membranes with microvilli-like projections; the tightly juxtaposed cells had distinct cytoplasmic membranes.

Written informed consent was obtained from the patient and this status was approved by the ethical committee of our institution.

DISCUSSION

Right ventricular myxomas are extremely rare. They are inserted mainly on the right ventricle free wall or interventricular septum; in rare instances these involve the tricuspid valve (5). Rarely, right ventricular myxomas are associated with other intracardiac myxomas and with extracardiac anomalies (6).

Right ventricular myxomas may simply present as an indefinite illness, but frequently are associated with syncopal episodes due to the transient occlusion of the tricuspid or pulmonary valves or with exertional dyspnea or chest discomfort (3). A harsh ejection murmur on the pulmonary area is a frequent finding, and pulmonary valve stenosis must be ruled out in this stiuation (3, 5). Electrocardiogram and chest X-ray are often nondiagnostic; although, sometimes an incomplete right bundle branch block may be present as in our case, or calcific deposits in the heart on standard radiographs may raise the suspicion of a tumor (4). Since cardiac catheterization can fail to demonstrate a transpulmonary gradient, and may be harmful because of the risk of pulmonary embolism, visualization of the mass by noninvasive techniques such as echocardiography and magnetic resonance imaging has been considered to be the final step to a correct diagnosis. Recent reports have emphasized the importance and reliability of noninvasive techniques in the detection of intracardiac masses (1, 8). The usefulness of MRI in assessing cardiac and paracardiac masses has been reviewed by some authors (2, 7).

Excision of the tumor can be performed through the right atrium. Right atrial approach for right ventricular myxomas is successful in achieving a satisfactory exposure and complete excision of the tumor and its insertion. Furthermore, it offers an easy way for inspection of the left cardiac cavities through the foramen ovale in order to rule out the presence of additional small tumors not detected preoperatively. In our patient with right ventricular myxoma, complete excision of the tumor was achi-

eved by limiting the pedicle. Full-thickness excision of the right ventricle wall is not justified according to the literature, since there are no reports of recurrence of such neoplasm; however, it would be necessary in the case of obvious or even suspected local infiltration.

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