Case report

Obstructive right ventricular cardiac fibroma in an adult

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Abstract

This study reports the case of a large intramural right ventricular cardiac fibroma, causing a right medio-ventricular stenosis and full loss of consciousness in a 31-year-old female patient, which was successfully treated by enucleation. © 1998 Elsevier Science B.V. All rights reserved.

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1. Case report

In April, 1994, a 31-year-old woman was referred to the Marie Lannelongue institute for treatment of a tumor of the right ventricle. The tumor was discovered because of a complaint of loss of consciousness while walking.

This patient had been in good health. Family history was not contributory and her past medical history was unremarkable. On physical examination, the patient looked healthy and did not have any obvious manifestations of a cardiovascular disorder. A grade 2/6 diastolic rumble was found at the apex. The electrocardiogram showed normal sinus rhythm except for an ST-segment elevation in Leads III, aVR, and the right precordial leads. The chest X-ray film showed cardiomegaly (cardiothoracic ratio, 0.6) without abnormal contours, or calcification. Two dimensional echography revealed a nodular mass (6 × 4 cm) developed in the free right ventricular wall, obstructing the right outflow and causing a pressure gradient across the right ventricular outflow tract of 35 mmHG during the doppler examination. Magnetic resonance imaging (MRI) showed an homogeneous signal intensity, isointense to the skeletal muscle. Angiography showed a filling defect in the right ventricular cavity, without coronary displacement.

The operation was performed under extracorporeal circulation in April, 1994. A 6 × 3.5 × 2.5-cm nodular, gray and white, firm tumor was found occupying the quasi-totality of the right anterior myocardium wall, extending from the left anterior descending artery to the right side of the heart (Fig. 1). An incision was made over the apical aspect of the tumor, which had no real capsule, but was surrounded by compressed muscles. Sharp dissection was used to remove this fibrous tumor from the myocardium wall. During the dissection, the right ventricular cavity was not entered and no coronary damage occurred. Right atriotomy was performed in order to control the integrity of the tricuspid valve and the sub-valvular apparatus. The ventricular wall was closed with three layers of silk and a continuous epicardial suture. The patient was weaned from bypass without difficulty and recovery from the operation was uneventful. The postoperative echocardiogram revealed a thickening of the ventricular wall in the resected area. The immunohistochemic examination showed fibrous conjunctive tissue Fig. 2 with rare and regular fibroblastic cells, which did not express either

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CD34 antigen or S-100 protein, and diagnosis was confirmed to be fibroma.

The patient continues to do well 3 years postoperatively. Echocardiographic examination shows no inflow or outflow obstruction and no significant modification in the resected area.

2. Comment

Cardiac fibroma are rare benign tumors. They are the second most common benign primary cardiac tumor after myxoma. Fewer than 100 cases have been reported [1]. In the present case, the tumor was discovered in a 31-year-old patient, although cardiac fibroma are considered as tumor of infancy and childhood. More than 80% of cardiac fibroma occur in children, one third of whom are under 1 year old. Fibroma are the first resected cardiac tumor of children and the second after rhabdomyoma in autopsy series of childhood [1]. There is no sex predilection or familial incidence noted in the literature [1–3].

Cardiac fibroma are benign histologically and do not metastasize or infiltrate but because of their location may cause severe cardiac dysfunction or sudden death. Their recognition is obviously difficult—most clinical signs being non-specific. Congestive heart failure, heart murmurs, and sudden death are the most common presenting features [4]. Depending on their location, cardiac fibromas may mimic hypertrophic sub-aortic stenosis, pulmonary stenosis, tricuspid stenosis, coronary disease with left ventricular aneurysm, and endocardial fibroelastosis [1,3]. Arrhythmia is found in association with cardiac fibroma in approximately 30% of cases [4,5]. Ventricular arrhythmia, explains the high incidence rate of sudden death encountered in these patients. A large variety of non-specific abnormalities could be found on the ECG, including ventricular hypertrophy, rhythm or conduction disturbance of pattern of ischemia, or infarction [5]. Radiological examination may reveal diffuse cardiomegalia with or without a focal cardiac bulge, frequently associated with the discovery of congestive heart failure. Calcification is non-specific and not frequent, being found in 15% of cases [3,4]. Although, associated with irregular cardiomegalia, particularly in children, it is often the one finding which points toward the diagnosis [4]. The role of echocardiography is well established. Two-dimensional echocardiography can characterize the tumor location and extend its echogenicity, as well as its connection with other cardiac structures, elucidating their physiopathological repercussions. The majority of the fibromas are found in the interventricular septum or in the left ventricular free wall [1,2,5]. Less than 10% are located in the right ventricular free wall, and occasionally a tumor may arise from the right or the left atrium [1,2,4,6]. Chest CT and MRI may prove to be of benefit in delimiting cardiac tumors and could provide good prediction of resectability [7,8]. At angiography,
tumors usually produce significant filling defects in the ventricular cavities. Although cardiac catheterization is less useful for diagnosis, it could be of help in looking for other cardiac diseases and coronary arteries displacement.

Since the first successful surgical treatment in 1962 prognosis of cardiac fibroma is closely related to resectability [6–8]. Operative resection should be complete because of the propensity of recurrence of this tumor [7]. After complete resection excellent long-term survival (up to 17 years after operation) has been reported [4]. In order to perform complete resection, wide excision with extensive reconstruction such as closure of inter-ventricular septal defect, dacron patch for ventricular remodelling, coronary artery bypass grafting, valve repairs or valve replacement, were necessary [6]. Even a cardiac transplantation was found to be necessary in one reported case [6]. Although complete resection still remains the best treatment, in particular cases, a careful partial resection [9,10] may provide a satisfactory palliative solution for such a poor evolutive benign tumor.

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References