Severe quadricuspid aortic valve stenosis after mediastinal irradiation

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Quadricuspid aortic valve is an uncommon cause of valve dysfunction, because it is usually found at necropsy or incidentally recognized at operation for severe aortic regurgitation.[1-3] Severe aortic stenosis requiring valve replacement involving a quadricuspid aortic valve has not been reported. In the case presented here, stenosis in a quadricuspid aortic valve is likely to have been favored by previous mediastinal irradiation.

Clinical Summary

A 48-year-old man began having exertional angina 1 year before admission. Risk factors for cardiovascular disease included hypercholesterolemia and smoking. Approximately 30 years previously, he had received mediastinal irradiation for a metastatic seminoma with a total radiation dose of 3000 cCy without further sequelae. A stress test was negative for myocardial ischemia, but a transthoracic 2-dimensional echocardiogram disclosed severe calcific aortic stenosis with a peak transvalvular gradient of 90 mm Hg and trivial aortic regurgitation. There was also evidence of left ventricular hypertrophy with preserved function. After recurrent episodes of chest pain, he was admitted for further evaluation and treatment. Physical examination showed a systolic ejection murmur in the aortic area. The electrocardiogram disclosed sinus rhythm and signs of left ventricular hypertrophy, whereas the chest x-ray film showed no abnormalities. Angiography showed occlusion of the right coronary artery and stenosis of a small marginal branch of the left circumflex; fluoroscopy revealed diffuse calcification of the aortic root. At operation in September 2002, calcifications appeared confined to the aortic root. The aortic arch was cannulated, and on moderately hypothermic cardiopulmonary bypass, the heart arrested with cold blood cardioplegic solution infused into the aortic root. A segment of saphenous vein was grafted to the right coronary artery. The aortic valve appeared formed by 3 cusps of equal size and a small accessory cusp between the right and noncoronary cusps; all cusps were fibrotic and grossly calcified (Figure 1). Because of the coarse calcification of the aorta, the aortic valve and root were replaced with a 23-mm cryopreserved aortic homograft with coronary ostia reimplantation. The proximal end of the venous graft was then anastomosed to the donor aorta before release of the aortic crossclamp. The patient recovered uneventfully and was discharged on postoperative day 7.

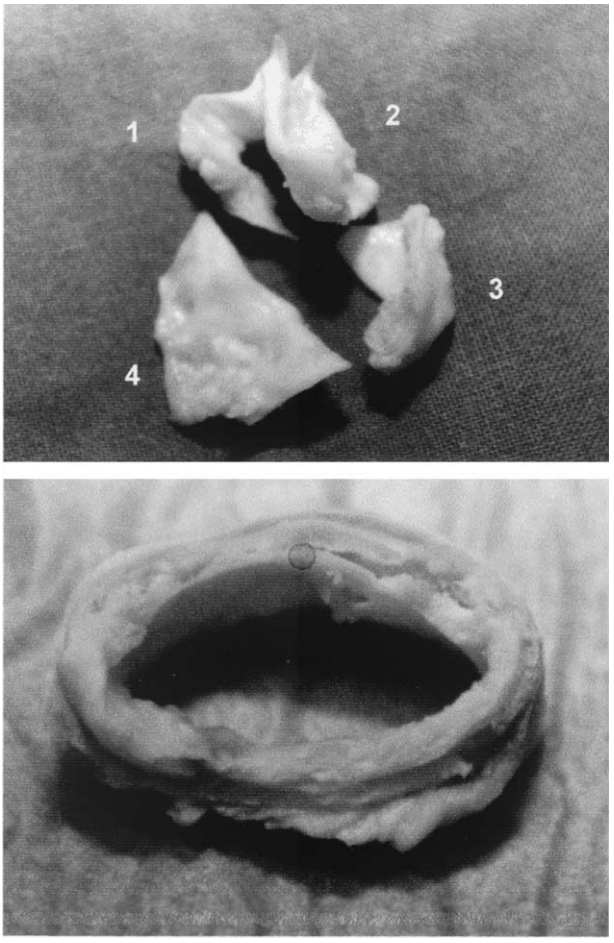

Figure 1. Gross view of the explanted valve *(upper)* showing 4 calcified cusps indicated by numbers. Gross view of a segment of ascending aorta *(lower)* with thickened and calcified wall.

Discussion

The adverse effects of mediastinal radiation therapy on the cardiovascular system are well documented. Pericardial disease is clinically evident in 60% to 70% of patients receiving mediastinal irradiation followed by valvular and coronary disease.[4] In a postmortem series, Brosius and colleagues[5] found that 80% of the patients studied showed evidence of valve damage after mediastinal irradiation. Veinot and Edwards[6] reported a pathologic study of 27 patients exhibiting radiation-related cardiac injuries involving

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25 valves with various degrees of fibrosis and calcification. Surgical treatment of postradiation valvular disease has been reported, predominantly in patients with aortic stenosis.[4] The largest surgical series for radiation-induced valvular heart disease was recently performed by Handa and associates.[7] In this single-center retrospective study of 60 patients, they reported a 43% incidence of isolated aortic valve replacements.

Valvular incompetence is the most frequent hemodynamic abnormality in patients with a quadricuspid aortic valve.[1-3] The occurrence of a severe aortic stenosis in the setting of a quadricuspid aortic valve is an exceptional finding and has not been reported. No such cases were recorded by Turri and colleagues[8] among 140 aortic valve specimens retrieved at surgery for aortic stenosis, and Passik and colleagues[9] did not mention any case of quadricuspid stenotic valve in 646 aortic valves explanted for pure stenosis. Our patient had severe aortic stenosis and coronary artery disease approximately 30 years after mediastinal irradiation for a metastatic seminoma. At operation, a calcified quadricuspid aortic valve was found characterized by a small accessory cusp between the right and noncoronary cusps which, according to Hurwitz and Roberts,[1] is the most frequent variety of this malformation.

Quadricuspid aortic valves are prone to develop fibrosis, causing valve incompetence.[3] In the present case, previous mediastinal irradiation is likely to have contributed to cusp calcification, allowing this malformation to develop an unusual pattern of valve dysfunction. Concomitant severe calcification of the ascending aorta and coronary artery disease indicate an extensive cardiac involvement from the effects of radiation.

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Chylous leak after cervical mediastinoscopy

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Chylous leak after cervical mediastinoscopy is a rare complication.[1] To our knowledge, 3 cases have been reported in the literature.[2-4] Three consecutive cases observed at our institution drew our attention to its possibly underrated occurrence.

Clinical Summary

PATIENT 1. A 16-year-old girl reported having weight loss and asthenia after an episode of erythema nodosum. On physical examination, there was no lymphadenopathy or hepatomegaly. The chest radiograph demonstrated bulky paratracheal lymph nodes. Tuberculosis was suspected but could not be confirmed. Diagnosis was assessed by cervical mediastinoscopy, so as to rule out a lymphoma. During the procedure, active suction drainage of the mediastinal bed was used, yielding 300 mL of a milky fluid on postoperative day 1. A chylous leak was confirmed by the presence of chylomicrons and triglycerides (14.6 mmol/L) in the fluid. Suction was discontinued, and a medium-chain triglyceride (MCT) diet was started. Lymphangiography demonstrated backflow of the contrast medium from the thoracic duct into the paratracheal lymph nodes (Figure 1). The chylous leak stopped, the mediastinum remained unchanged on successive chest radiographs, and the drain was removed on the fourth postoperative day.

PATIENT 2. A 56-year-old man had a superior vena caval obstruction syndrome. On physical examination, there was no lymphadenopathy or hepatomegaly. Chest tomodensitometry demonstrated a huge right lung tumor and large paratracheal lymph nodes. Fibrobronchoscopy failed to provide a diagnosis. A mediastinoscopy was performed. The mediastinal bed was not drained. The diagnosis of squamous cell carcinoma was confirmed. On postoperative day 3, the mediastinoscopy incision leaked a milky

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