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Actinic Papillary Fibroelastoma of the Left Ventricle

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Abstract

We present the case of a 69-year-old woman with a history of endometrial carcinoma in 1996, who underwent a total hysterectomy and bilateral adnexectomy. The patient also received chemotherapy (doxorubicin and cisplatinum) and local radiotherapy (50 Gy) because of a single lung metastasis, with total remission during later follow-up. During follow-up, 10 years later following radiotherapy, a transthoracic echocardiogram (TTE) revealed an image consistent with a primary cardiac tumor (papillary fibroelastoma) or metastatic cardiac tumor on the posteromedial papillary muscle. Cardiac magnetic resonance imaging (MRI) revealed a solid mass on the posteromedial papillary muscle with late enhancement, consistent with a primary cardiac tumor. During surgery, the tumor located in the posteromedial papillary muscle was resected. A pathological examination revealed the presence of a tumor mass with a core of dense connective tissue surrounded by a layer of hyperplastic endocardial cells characteristic of a papillary fibroelastoma. After 8 years of follow-up, the patient remains asymptomatic.

Keywords: cardiac tumor, radiotherapy, actinic papillary fibroelastoma, echocardiography, left ventricular mass, malignancy

1. Background

Radiotherapy can affect the heart and lead to effusive and constrictive pericarditis, coronary artery disease, myocardial fibrosis, and valvular disease. Radiation-associated valvular disease can progress over several years from asymptomatic valvar thickening to symptomatic valvular dysfunction. We report the case of a woman, who developed severe mitral regurgitation and an endocardial left ventricular papillary fibroelastoma (PFE) a long time after chest radiotherapy. The mechanism for this pathology is demonstrated, and histological findings are shown.

2. Case report

We present the case of a 69-year-old woman with a history of endometrial carcinoma in 1996, who underwent a total hysterectomy and bilateral adnexectomy. The patient also received chemotherapy (doxorubicin and cisplatin) and local radiotherapy (50 Gy) because of a single lung metastasis, with total remission during later follow-up. About 10 years later following radiotherapy, she had increasing breathlessness and a new pansystolic murmur at the left sternal edge and progressed insidiously to retrograde left heart failure [1].

An ECG showed a sinus rhythm and complete right bundle branch block. A chest X-ray revealed a normal cardiac contour with upper right pleural thickening with a sequela appearance, and blood flow redistribution consistent with pulmonary edema. A transthoracic and transesophageal echocardiogram showed thickening and retraction of the mitral valve, severe mitral regurgitation, and severe pulmonary hypertension. The left ventricular function was normal. An assessment of myocardial perfusion with SPECT and coronary angiography were normal. During follow-up, a transthoracic echocardiogram (TTE) revealed an image consistent with a primary or metastatic cardiac tumor on the posteromedial papillary muscle (**Figure 1A**, <https://www.dropbox.com/s/q9mtt67rcvps82u/Movies%201%20and%202.rar?dl=1>). Cardiac magnetic resonance imaging (MRI) revealed a solid mass on the posteromedial papillary muscle with late enhancement, consistent with a primary cardiac tumor (**Figure 1B**).

Parasternal short-axis view from a transthoracic echocardiogram showing a solid mass on the posteromedial papillary muscle consistent with a primary cardiac tumor (papillary fibroelastoma). A mild pericardial effusion and large left pleural effusion are also visible.

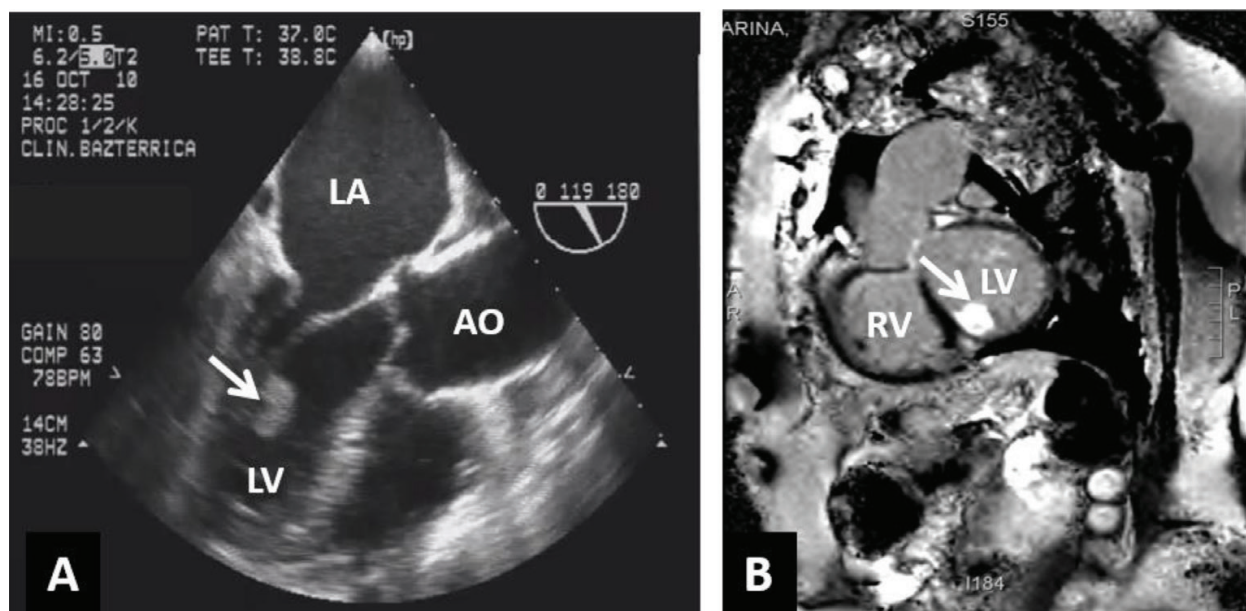


Figure 1. Transesophageal echocardiography. Longitudinal view of the left ventricle at 119° (A) and cardiac MRI (B) shows a solid mass on the posteromedial papillary muscle (arrows), measuring 21.9 × 14.6 mm with gadolinium enhancement consistent with a primary cardiac tumor. LA = left atrium; LV = left ventricle; RV = right atrium; Ao = ascending aorta.

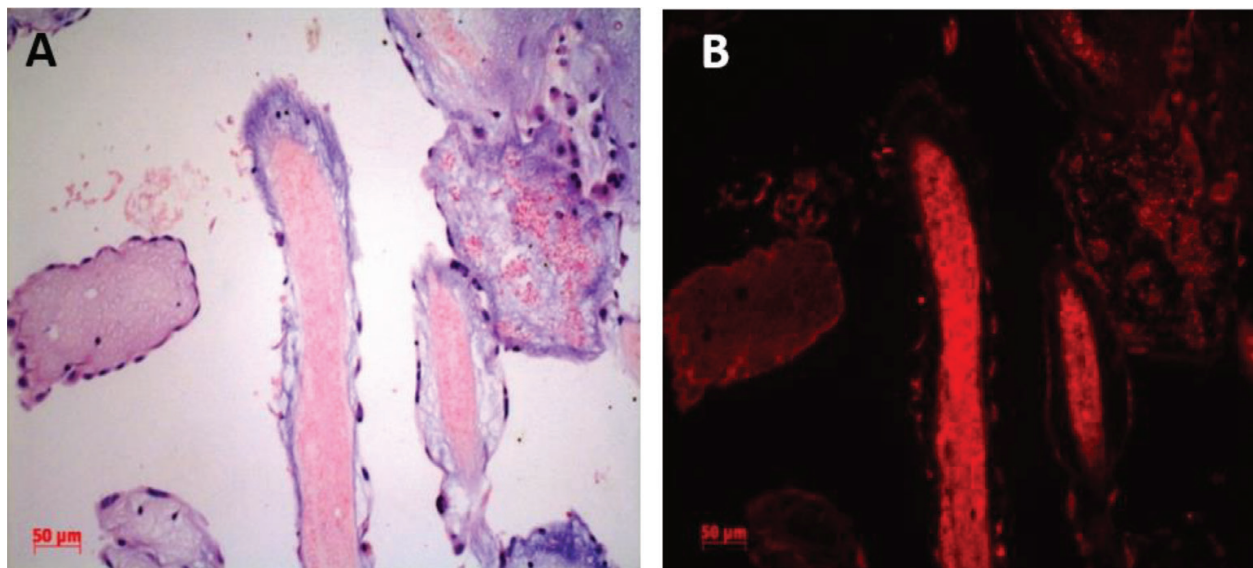


Figure 2. Pathology examination. A: Panoramic image of the papillary fibroelastoma and the villi. B: With fluorescence technique, the same papilla shows an axis of elastic fibers.

During surgery, the mitral valve was thickened, and the patient underwent mitral valve replacement with an SJM #29 biological prosthesis with resection of the tumor located in the posteromedial papillary muscle. During the postoperative period, the retrograde heart failure regressed, pulmonary pressures were normalized, and the patient remained symptom-free. A pathological examination revealed the presence of a tumor mass with a core of dense connective tissue surrounded by a layer of hyperplastic endocardial cells characteristic of a papillary fibroelastoma (**Figure 2**); the mitral valve had signs of radiotherapy-induced damage with increased collagen and dysmorphic nuclei. After 8 years of follow-up, the patient remains asymptomatic.

3. Discussion

Radiosensitivity of the myocardium, pericardium, and great vessels is an issue of great concern. Previously, the heart was thought to be a radio-resistant organ; however, this theory was subsequently abandoned when late cardiac involvement was found in young patients, who had received radiotherapy treatment due to Hodgkin's disease [2].

The prevalence of cardiac disease associated with radiation may clinically manifest after a very prolonged time period, and its incidence is on the rise because of the increasing survival of cancer patients. Modern treatment techniques seem to have decreased the toxicity, but long-term results that allow an assessment of its effects are still lacking. It is worth noting that survivors who do not experience recurrence of the original tumor or develop a second malignant tumor have a greater risk of cardiovascular death than the general population, and in some series that risk is fivefold [3].

Radiation causes progressive and irreversible damage; acute and chronic pericardial manifestations are the most frequent manifestations seen in daily practice. However, valvular lesions,

conduction abnormalities, and cardiomyopathies due to myocardial fibrosis or coronary disease have also been described. International guidelines on cancer management recommend aggressive management of cardiovascular risk factors and performing perfusion exams or echocardiograms at baseline and 10 years after radiation therapy to detect coronary disease and valvular lesions in these patients [4].

In terms of the additional incidental finding of a PFE on the papillary muscle, it should be noted that PFEs are the second most common primary benign cardiac tumor after myxoma, and it most often affects the heart valves, as opposed to myxomas, which are most often found in the atria.

A cardiac MRI with gadolinium showed that the solid mass on the papillary muscle exhibited late enhancement, suggesting that the mass was consistent with a primary cardiac tumor rather than a metastatic lesion. The characteristic irregular surface with mobile papillae observed on the TEE suggested that the tumor was a PFE.

The valves most frequently affected by PFEs are the mitral and aortic valves. In our experience with a total of 54 papillary fibroelastomas [5], in 29.6% (16/54) of patients, they were located in the endocardium of the left ventricle (LV), as in this case. The most common symptoms are due to a peripheral embolism, and the following symptoms have been described: stroke, acute myocardial infarction, and sudden death due to an embolism or obstruction of the coronary ostium. This type of cardiac tumor is not associated with valvular regurgitation. Several authors have reported that this type of tumor is more often seen in elderly patients with longstanding heart disease; therefore, this suggests that they occur due to mechanical damage and involve a degenerative process [6]. In accordance with this theory, in a series of 17 patients with a diagnosis of fibroelastoma, echocardiography was shown to correlate well with pathological findings, and these tumors were usually found in areas of cardiac irritation, such as the aortic and mitral valves [5]. This information proved to be very important in our case, since the damage caused by radiotherapy might have affected the valvular and subvalvular mitral apparatus, thus enhancing the potential development of this type of tumor.

4. Conclusion

We report the case of a patient with radiotherapy-induced symptomatic severe mitral regurgitation and an asymptomatic papillary fibroelastoma, which is a rare complication of thoracic radiotherapy that required valve replacement and resection of the tumor. The latent period between radiotherapy and these complications was 10 years and was related to the dose of radiation received. The increase in survival seen in cancer patients has resulted in an increased frequency of these types of complications. This unusual mechanism of formation of an actinic papillary fibroelastoma should be disseminated among cardiologists. In patients who have survived for long periods of time after radiotherapy, it is important to remember that cardiac complications may indeed occur, and the treating physician is responsible for detecting them.

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