Radiation induced valvulitis with late leaflet rupture

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Abstract
Various cardiac sequelae of mediastinal irradiation have been reported, from pericarditis to conduction defects. Despite the potentially fatal nature of some of these abnormalities, many may present with few or no symptoms. In this case report, the patient, who had received 4000 rads to the mediastinum 24 years previously, presented with worsening shortness of breath and two episodes of lightheadedness. Subsequently, he was found to have aortic valve rupture associated with fibrosis. A review of the literature indicates that valve rupture is a novel consequence of mediastinal radiation.

Keywords: mediastinal radiation; valvulitis; aortic valve; rupture

Heart disease is a well documented result of mediastinal radiation. Various manifestations have been reported, from pericarditis as the most common,\(^1\) to valvar stenosis,\(^2\) valvar regurgitation,\(^1\) and conduction defects.\(^4,5\) Stenosis and regurgitation secondary to radiation have been described in all heart valves; however, left sided valvar radiation disease is predominant.\(^2,6\) Many of these abnormalities may present with few or no symptoms,\(^4,7\) despite potentially fatal sequelae. In this case report, the patient, who had received 4000 rads to the mediastinum, presented with worsening shortness of breath and two episodes of lightheadedness. Subsequently, he was found to have aortic valve rupture associated with fibrosis. To our knowledge, valve rupture has not been reported as a consequence of mediastinal radiation.

Case report
A 54 year old man, a former military helicopter pilot with a history of Hodgkin’s lymphoma, had received 4000 rads to the mediastinum 24 years previously. He had a history of hypertension, well controlled with enalapril and furosemide. Restrictive pulmonary disease had been diagnosed and was thought to be related to the previous irradiation. The patient had known stable aortic stenosis without significant regurgitation, six weeks of progressive dyspnoea on exertion, and two episodes of lightheadedness at rest. On physical examination, his blood pressure was 142/44 mm Hg. There was a grade 4/6 systolic ejection murmur radiating to the carotids and a grade 3/6 early diastolic decrescendo murmur. A transthoracic echocardiogram showed aortic stenosis and new severe aortic regurgitation. The estimated ejection fraction was 50% (decreased from 60% one year previously). Cardiac catheterisation showed a pulmonary artery pressure of 47/23 mm Hg, a left ventricular end diastolic pressure of 26 mm Hg, and a 95% stenosis of the right coronary artery.

At operation, several findings indicated previous radiation: the sternal marrow had decreased density and a white, fibrous tissue enshewed portions of the aorta, right atrium, and right ventricle. Particularly dense adhesions surrounded the proximal ascending aorta, with scattered adhesions between the aorta and the pericardium. The transoesophageal echocardiogram showed severe aortic regurgitation with a tricuspid aortic valve. On direct visualisation, the three leaflets had calcium deposits and fibrotic changes, with areas of thickening. The non-coronary leaflet had ruptured near its base (as indicated by the arrow in fig 1). The valve was replaced with a 25 mm St Jude mechanical prosthesis.

Figure 1 Rupture of the non-coronary leaflet of the aortic valve in a patient with radiation induced valvulitis.
artery, as the findings at surgery suggested that the internal thoracic arteries may well have developed radiation damage themselves. The patient’s hospital course was uneventful. Five months postoperatively, he was feeling well and fully active, noting limitation only with heavy exercise.

**Discussion**

Hodgkin’s lymphoma is newly diagnosed in approximately 7500 patients every year in the United States, with radiation therapy being the mainstay of treatment in early stage disease. Consequently, Hodgkin’s lymphoma is the predominant cause of the reported cases of radiation induced heart disease; however, a number of other diseases can result in similar pathologies. Pericarditis has been noted after radiation therapy for breast cancer,\(^1\) cystic hygroma,\(^1\) carcinoid tumour,\(^1\) thymoma,\(^2\) squamous cell carcinoma, non-Hodgkin’s lymphoma, and neurofibrosarcoma.\(^2\) Estimates of the incidence of postradiation heart disease range widely, from 0% at low doses (<3000 rads)\(^3\) to 96% at high doses.\(^7\) However, disease has been found in patients receiving as little as 1530 rads.\(^3\) Important variables are the age at which the patient received the radiation therapy, the extent of direct radiation to the heart, and, possibly more important for our patient, the time since treatment.\(^3\)\(^,\)\(^7\) This case extends the list of pathological cardiac sequelae that may follow chest radiation.

The prognosis for many of the sequelae, such as pericarditis or valvulitis, is good, and the conditions tend to respond well to surgical intervention.\(^6\) Nevertheless, the disease may progress in the asymptomatic patient with severe late manifestations. Consequently, mediastinal radiation is an important cardiac risk factor to identify. Time since treatment, location of the radiation portal, and radiation dose are important factors to elicit in such a history. Regular cardiac evaluation is important for all patients who have undergone mediastinal radiation.