Haemangiosarcoma of the Pulmonary Valve Presenting as a Pulmonary Stenosis
A Case Report

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SUMMARY

Intractable congestive cardiac failure, unexplained arrhythmias, changes in the cardiac silhouette and murmurs which change with position are all features which should arouse suspicion of a cardiac tumour.

A case of intracardiac haemangiosarcoma of endothelial origin is discussed. The tumour presented as a pulmonary stenosis and extended into the right ventricular myocardium, causing a right bundle-branch block. At first the tumour seemed to be restricted to the endothelium of the pulmonary vascular bed, but it later infiltrated the interstitium and bronchi, causing dyspnoea and haemoptysis.

After surgical removal of the tumour and reconstruction of the pulmonary outflow tract, the patient was free of symptoms for 14 months. However, rapid tumour growth in the pulmonary arterial system, lung interstitium and right ventricle subsequently recurred. Deep radiotherapy did not cause the tumour to regress.


Antemortem diagnosis of intracardiac tumours is usually limited to myxomas. Primary intracardiac tumours are rare, comprising only 0.05% or less of autopsy material. Myxomas comprise over half the total number of primary intracardiac tumours and sarcomas comprise one-third. Benign tumours make up the remainder.

A primary haemangiosarcoma originating from the endothelium of the right ventricle and extending into the pulmonary vascular bed, with destruction of the pulmonic leaflets, has, to our knowledge, not been reported before.

CASE REPORT

A 56-year-old White woman was referred to us with a 6-month history of rapidly increasing cardiac failure unresponsive to any form of medical therapy. She had been well until 6 months previously, when she became orthopnoeic and dyspnoeic. One month before admission to hospital, she noticed that her legs were swollen, her abdomen was distended and the previous symptoms were increasing alarmingly.

On clinical examination she was distressed, and severe neck vein congestion, mitral facies and slight peripheral oedema were present. There was no hepatomegaly. The heart was not enlarged but a prominent heave was present over the right ventricular outflow tract. The S1 was blunt and S2(P) was of normal intensity and single. A grade 3/6 systolic ejection murmur was heard maximally over the pulmonary area, but radiated widely towards the apex and the left shoulder. Chest radiographs showed no main pulmonary artery dilatation and right ventricular enlargement (Fig. 1). An ECG showed intraventricular conduction delay with a right bundle-branch block (Fig. 2). Echocardiography confirmed the right ventricular enlargement.

Cardiac catheterization was performed and a 70-mmHg pressure gradient was found between the main pulmonary artery and right ventricular cavity (Fig. 3). Angiography demonstrated filling defects in the right ventricular cavity.
Fig. 4. Right ventriculogram showing various large oval-filling defects in the right ventricular cavity and an absence of post-stenotic dilatation of the main pulmonary artery.

At operation, light-brown, soft, fragile tumour tissue was present in the main pulmonary artery and extended into the right ventricular outflow tract. The sinuses of Valsalva were completely filled with, and the pulmonary valve slips partially destroyed and replaced by, tumour tissue (Figs 5 and 6). The tumour tissue could be removed like a cast out of the main pulmonary artery. All the tumour tissue was excised, and the right ventricular outflow tract was reconstructed with a pericardial flap that extended up to the bifurcation of the pulmonary artery. A No. 23 Björk-Shiley disc prosthesis was inserted at an angle.

Histological examination of the tumour tissue showed an abundance of plasma-like cells, and at that stage, the tumour could not be identified.

The postoperative course was uneventful and the patient remained asymptomatic until 1 year later when haemoptysis occurred. Chest radiographs showed extensive infiltration in the hilar regions, with extension into the periphery of the lungs (Fig. 7). Angiography confirmed the suspicion of tumour recurrence in the right ventricle (Fig. 8). On bronchoscopy, infiltration of the tumour into the left main bronchus could be seen. Biopsy showed a haemangiosarcoma of endothelial origin.

The patient received a course of deep radiotherapy to a total of 4000 rad. The tumour appeared to be wholly unresponsive to radiotherapy and on follow-up chest radiographs, diffuse, bilateral pulmonary extension could be seen. Chemotherapy was not thought to be of any additional value.

The patient died 17 months after the diagnosis had been made.
Fig. 5. Section of main pulmonary artery and pulmonary valve. The tumour tissue has obliterated the sinuses of Valsalva, causing them to bulge.

DISCUSSION

In a patient presenting with cardiac debility of short duration, and relentless progression of congestive cardiac failure in spite of adequate therapy, the possibility of an intracardiac tumour should be considered.

Cardiac tumours cause symptoms and signs which depend on their location in and around the heart. These tumours are to be found in the pericardium, myocardium and endocardium.

Pericardial Tumours

Pericardial tumours usually present with symptoms of superior or inferior vena-caval obstruction, pericardial effusion or constrictive pericarditis when excessive epicardial deposits are present.

Myocardial Tumours

Myocardial tumours usually manifest in childhood as paroxysmal tachycardias, heart failure, and, in some cases, a murmur, due to partial obstruction of flow within the ventricle. Intraventricular conduction delay (correlating well with the extent of tumour infiltration), suggests invasion of the His bundle and Purkinje system.

A benign hamartoma is inclined to contain calcifications, and on chest radiography, an abnormal bulge may be seen on the cardiac border. Benign tumours usually comprise fibromas, hamartomas or, rarely, myxomas, whereas sarcomas, malignant lymphomas or metastases make up the malignant group.

Endocardial Tumours

Endocardial myxomas are the most common intracardiac tumours. Seventy-five per cent occur on the left side and 25% on the right side, mostly near the foramen ovale. Obstruction of the mitral or tricuspid valves, and emboli from tumour fragmentation are common complications. Reports on tumours of the right ventricle or pulmonary artery are few. Green et al. reported a fibromyxosarcoma of the pulmonary artery, Catton et al. a myxoma of the pulmonary valve, and Nicks a hamartoma of the right ventricle. Two fibromas of the right ventricle and
Fig. 7. Chest radiograph taken on second admission. Diffuse infiltrations into the hilar regions and right lung base, as well as scattered small irregular densities throughout both lung fields, are visible.

4 cases of myxoma of the right ventricle\(^1\),\(^2\),\(^3\) have also been reported.

The pointers to an intracardiac tumour in our patient were intractable cardiac failure, absence of post-stenotic dilatation of the main pulmonary artery, a loud systolic ejection murmur over the right ventricular outflow tract and pulmonary valve area, and an intraventricular conduction delay. Angiography should be done in any problematic case to confirm the diagnosis. Once the diagnosis is made, surgery is the obvious treatment — even if only for symptomatic relief.

The question whether chemotherapy or radiotherapy is of any benefit is still an open one. The experience with chemotherapy in haemangiosarcomas has been extremely limited and the results have been poor. Radiotherapy in general is thought to be elective in rapidly growing tumours. Our patient showed no signs of tumour regression at all, although there was an initial decrease in haemoptysis.

REFERENCES