Malignant fibrous histiocytoma of the heart

A case report

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Summary

Malignant fibrous histiocytoma is a recently recognised common sarcoma of late adult life which, rarely, may arise in the heart. Unlike the atrial myxoma which it may mimic clinically, malignant fibrous histiocytoma is a far more aggressive tumour with a greater tendency for local recurrence and infiltration. A case of a primary intracardiac malignant fibrous histiocytoma is reported and relevant published reports reviewed.

Malignant fibrous histiocytoma is a relatively newly described entity, which is widely recognised as occurring in the soft tissues and bone but may occasionally involve the heart or great vessels. In the patient described below the malignant fibrous histiocytoma arose in the left atrium, mimicked a left atrial myxoma clinically and recurred after attempted surgical removal.

Case report

A 76-year-old woman doctor presented to hospital with sudden onset of acute dyspnoea, palpitations, atypical chest pain and paroxysmal nocturnal dyspnoea of 3 weeks' duration. Ankle oedema also developed and she had lost about 15 kg in weight over 6 months. Examination revealed a thin, apyrexial woman with ankle oedema, blood pressure of 120/60 mmHg, and a loud first heart sound with a short systolic murmur. The ECG showed a sinus tachycardia of 100/min, a P-R interval of 0.12 s with an axis of +90 and evidence of incomplete right bundle-branch block. Treatment with digitalis and diuretics led to transient improvement. Chest radiography showed borderline cardiomegaly with interstitial pulmonary oedema, Kerley B lines and a small right pleural effusion. The blood chemistry was normal. Echocardiography revealed a sessile left atrial tumour attached to the interatrial septum. Cardiac catheterisation (Fig. 1) revealed a filling defect in the left atrium, pulmonary hypertension and a pulmonary capillary wedge pressure elevated to 29 mmHg. A diagnosis of a left atrial myxoma was made and she was referred for urgent surgery.

The operation, performed on 24 November 1984, revealed a large, polypoid tumour attached to the left side of the atrial septum. Cardiac catheterisation (Fig. 1) revealed a filling defect in the left atrium, pulmonic hypertension and a pulmonary capillary wedge pressure elevated to 29 mmHg. A diagnosis of a left atrial myxoma was made and she was referred for urgent surgery.

The operation, performed on 24 November 1984, revealed a large, polypoid tumour attached to the left side of the atrial septum by a short pedicle. The tumour filled the entire left atrium as well as the left atrial appendage and completely occluded both right pulmonary veins. The neoplasm obstructed the mitral valve and it was attached to the valve leaflets by adhesions which were easily broken down. The tumour and its pedicle together with a portion of the atrial septum as well as the superior wall of the left atrium were excised and the defect was closed with a pericardial patch graft. Moderate mitral regurgitation was corrected by a posteromedial annuloplasty.

The excised greyish-coloured tumour, which measured 10 x 7 x 2 cm, weighed 130 g and had a 3 cm long pedicle. Histological examination (Fig. 2) revealed plump spindle-like fibrohistiocytic cells.
shaped cells arranged in a storiform pattern with an admixture of pleomorphic Touton-type giant cells. One mitosis per high-power field was observed. Areas of myxomatous change and necrosis were also seen. The tumour extended to the resection line and intravascular tumour was also observed. A malignant fibrous histiocytoma was diagnosed.

Postoperatively, the patient remained well for 6 months. Symptoms recurred abruptly in July 1985 and she was admitted to hospital with pulmonary oedema. Cardiac catheterisation revealed an elevated wedge pressure and angiography demonstrated a large left atrial tumour filling the whole of the left atrium and obstructing the pulmonary veins.

Re-operation was performed on 1 August 1985 to remove the recurrent tumour. At operation a large dumb-bell-shaped tumour filled both the left and right atria. The tumour was resected. The interatrial septum and the left atrial wall were reconstructed with prosthetic material. After a protracted operation the heart failed to sustain the circulation and the patient died.

At autopsy the recurrent, dumb-bell-shaped tumour measured 7 x 5 x 4 cm and consisted of a lobulated mass of pale tumour tissue exhibiting extensive haemorrhage and necrosis. The malignant fibrous histiocytoma was confined to the heart, but externally the tumour was invading the adventitia of the aorta and pulmonary artery (Fig. 3). On histological examination the appearance was identical to that of the original tumour.

Within an abundant collagenous stroma. While some atypism may be seen, mitoses are rare. A benign fibrous histiocytoma of the heart has previously been reported from our laboratory. A malignant fibrous histiocytoma is a sarcoma composed of malignant oval and spindle-shaped cells arranged in a characteristic storiform pattern. Malignant fibrous histiocytoma most commonly occurs in the limbs or less frequently in the deep soft tissues and it has been divided into fibrous, giant cell or inflammatory variants. Its histogenesis is uncertain and postulated cells of origin include histiocytes or primitive mesenchymal cells which may give rise to both fibroblastic and histiocytic cells. As far as we are aware, only 16 malignant fibrous histiocytomas arising primarily in the heart have been documented. All these cases have been reported since 1978 and 8 out of the 16 have come from Japan. A malignant fibrous histiocytoma of the pericardium has also been documented. In the case reported by Holtzman et al. the malignant fibrous histiocytoma arose in the left atrium adjacent to a valve prosthesis and the possibility of a causal relationship between the tumour and the prosthesis cannot be excluded. This possibility is strengthened by reports of malignant fibrous histiocytoma arising elsewhere at sites of previous surgery, e.g. alongside a metal plate in the femur, in a scar or at the site of excision of a lipoma or even in the vicinity of a bone infant.

In our patient the extent of the tumour, which involved the superior vena cava, pulmonary artery and aorta, had not been appreciated pre-operatively and rendered successful surgery impossible.

**Discussion**

Tumours of fibrohistiocytic origin are rare in the heart. Cardiac fibromas are most frequently located in the ventricular septum and have a bland appearance of spindle-shaped cells lying amongst collagen and elastin. The malignant fibrous histiocytoma is a tumour that may arise from any site, especially from the limbs. The clinical presentation may be caused by local infiltration or invasion of adjacent structures. The tumour may be benign or malignant, depending on its histological features. The malignant form is characterized by a high mitotic rate and evidence of nuclear pleomorphism. The tumour is often associated with necrosis and haemorrhage.

**REFERENCES**


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Anomalous origin of the right coronary artery from the left sinus of Valsalva

A report of 2 cases

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Summary

Two cases of inferior myocardial infarction associated with anomalous origin of the right coronary artery from the left sinus of Valsalva are described.

Anomalous origin of the right coronary artery from the left sinus of Valsalva is an uncommon congenital anomaly. Although previously considered benign, several reports have now shown that this anomaly may cause angina, myocardial infarction and sudden death. We report 2 further cases of inferior myocardial infarction associated with this anomaly.

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Case reports

Case 1

A 36-year-old Indian man was admitted to hospital with severe chest pain unrelated to physical activity. He had no history of angina and had smoked for 20 years. There were no other coronary risk factors. Clinical examination was normal. Serial cardiac enzyme values and electrocardiography confirmed the presence of inferior myocardial infarction. An exercise stress test performed 6 weeks later was normal. Because of his age, coronary angiography was performed to determine the extent of coronary artery disease. Left ventriculography demonstrated akinesis of the inferolateral segment of the left ventricle. The left coronary artery was normal. Despite the use of several catheters the right coronary artery could not be selectively engaged. Injection of contrast into the sinus of Valsalva demonstrated that the right coronary artery originated from the left sinus of Valsalva (Fig. 1). There was no evidence of atherosclerosis of this vessel. At follow-up 6 months later the patient remained asymptomatic.

Case 2

A 32-year-old Indian man with a 6-month history of effort-induced angina was admitted to hospital in December 1983 with inferior myocardial infarction. Apart from a family history of coronary artery disease, there were no other risk factors. On admission to the coronary care unit he was comfortable with a blood pressure of 120/80 mmHg and a regular radial pulse of 120/80 mmHg and a regular radial pulse of 80/min. Serial cardiac enzyme values confirmed the diagnosis of acute myocardial infarction and an ECG displayed a fully evolved Q-wave inferior infarct. The patient had an uneventful course and was discharged from hospital on day 8. At coronary angiography there was akinesis of the diaphragmatic segment. Several attempts to cannulate the right coronary artery failed and a sinus injection demonstrated its origin close to the ostium of the left coronary artery in the left sinus of Valsalva. There was no evidence of atherosclerotic disease in this vessel (Fig. 2).