

## Intra-myocardial right ventricular haemangioma in combination with a coronary fistula

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### Abstract

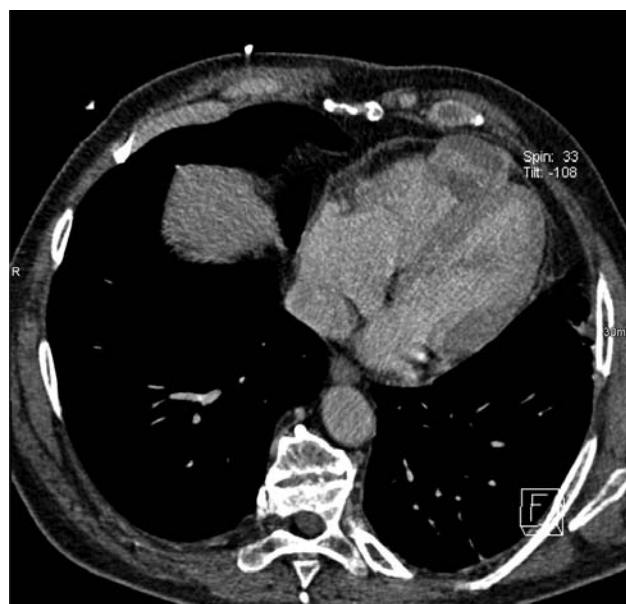
Cardiac haemangiomas are rare benign neoplasms of the heart. Though co-existing lesions have been described before, the present case is the first to report an intra-myocardial right ventricular haemangioma in combination with a coronary fistula and tricuspid regurgitation.

**Key words:** haemangioma, intra-myocardial.

### Case report

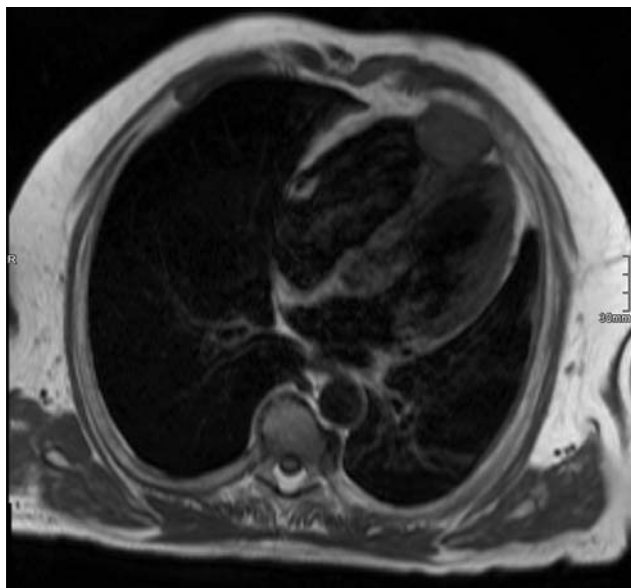
A 52-year old patient presented with dyspnoea. A chest x-ray revealed a pleural effusion which was treated with pleural drainage. After evacuation of a 2 000 ml empyema and antibiotic treatment for 21 days the patient recovered and was clinically asymptomatic. In an echocardiographic investigation a 41 mm pericardial mass was detected at the right ventricular wall. The patient was transferred to our clinic with suspected intrapericardial abscess formation for further evaluation and treatment. Coronary angiography revealed a stenosis of the LAD and a fistula from the LAD to the pulmonary artery. Tricuspid valve regurgitation was detected by echocardiography. In a CT scan, the pericardial mass was confirmed and a cardiac haemangioma suspected due to diffuse but vessel-like high central contrast enhancement. The tumor presented as intra-pericardial but extra-myocardial (Fig. 1). In addition an MRI of the heart was performed, showing intermediate signal intensity in T1 (Fig. 2) and high intensity in T2. After contrast medium administration a high signal was observed (Fig. 3). In the “true fisp” cine sequences a slight flow was observed inside the tumour. A discrete compression but no constriction of the right ventricle was given. The operation was performed with bi-caval cannulation in total bypass. After completing an IMA bypass to LAD, a tricuspid reconstruction by using a 30 mm Carpentier Edwards Ring was performed. The excision of the fistula was achieved by complete dissection of the adventitia of the pulmonary artery and ligation of the vessels at its coronary side and direct closure of the fistula after longitudinal opening of the pulmonary artery.

Intra-operatively it became obvious that the tumour was localized within the right ventricular wall and not intra-pericardially. Complete extirpation of the tumor required opening up the right ventricle and dissecting the tumour from within the right ventricular wall and the intra-ventricular septum. The incision was closed directly with two 5/0 running sutures. Histological examination confirmed a ca-

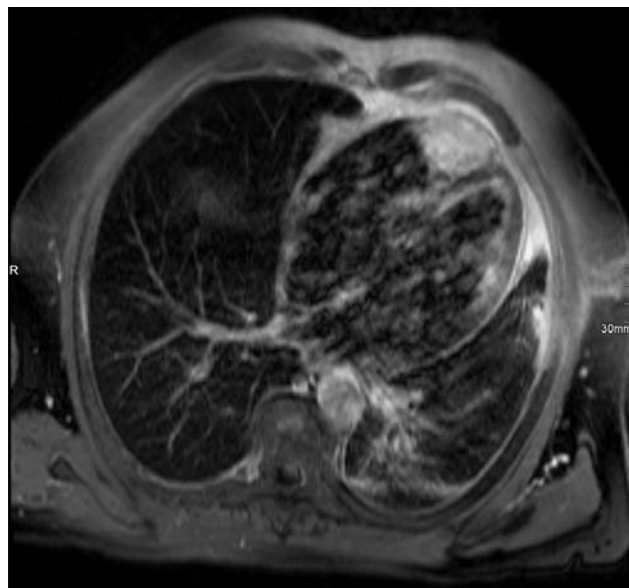


**Fig. 1.** Four chamber view reconstructed out of an axial CT data set showing the apical mass with central high density contrast enhancement

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**Fig. 2.** Four chamber view in MRI, T1 weighted, the mass presents intra-pericardial with intermediate signal intensity



**Fig. 3.** Four chamber view in MRI: massive contrast medium enhancement in the apical mass

venous haemangioma with an extension of 4 × 3.5 × 3 cm. The postoperative course was uneventful.

### Discussion

Cardiac haemangiomas, detected in 1-2% of all benign neoplasms of the heart, are extremely rare [1]. In a systematic review Pigato [2] found only 34 cases published in the literature until 1998. Most of these were identified while working up the patients' symptoms or clinical findings. The most common presentations were dyspnoea and arrhythmias. Signs of right heart failure were also frequently described. Differential diagnosis of mass lesions in the heart include thrombi, myxoma, lipoma, fibroma, cyst, and malignant tumour such as angiosarcoma [1]. Most of them are found to be intra-myocardial or intra-cardiac. Using the different MRI signal intensity characteristics, including rapid enhancement during first pass gadolinium contrast infusion, differential diagnosis may be specified to tumors of strong gadolinium enhancement such as pheochromocytoma, angiosarcoma, myxoma and rhabdomyosarcoma [3]. Upon appraisal of localisation, signal intensity, "flow void" effects, contrast enhancement and maybe additional examinations such as CT, the diagnosis can be ensured.

The outcome in patients with cardiac haemangioma depends on the surgical procedure. Patients with unresectable tumour may have a poor prognosis because of ven-

tricular tachycardia, sudden death, local progression or even systemic dissemination or stroke [1, 4]. Though an inter-atrial septum defect has been reported as co-existing cardiac lesion [4] the presented case is to the best of our knowledge the first description of a haemangioma in combination with a coronary fistula and tricuspid regurgitation.

MRI and CT investigations are able to differentiate myocardial masses. In the presented case, the diagnosis of a cardiac haemangioma was precise, but the localisation of the tumor was specified as intra-pericardial but extra-myocardial. However, intra-operatively the tumour was dissected from within the right ventricular wall and the intra-ventricular septum.

Both combined cardiac lesions and false anticipated tumor localisation should be considered in operative planning in the treatment of cardiac haemangioma.

### References

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