Right atrial and ventricular angiosarcoma

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A 28-year-old man without previous medical history presented with a 2-week history of progressive exertional dyspnoea. Chest X-ray revealed an increased cardio-thoracic ratio with pleural effusion. A subsequent CT-scan showed a massive amount of pericardial effusion, yielding 3.5 L following pericardiocentesis. Echocardiography revealed a tumour in the right atrium with extension into the right ventricle (Panel A). Additional CT (Panel B) and MRI imaging (Panel C) confirmed an intramural mass in the right atrium extending through the tricuspid valve into the right ventricle. Furthermore, epicardial and pericardial involvement with pericardial effusion was seen. There were no signs of distant tumours or metastatic activity. Transjugularly obtained tissue biopsies of the tumour confirmed the suspected diagnosis of angiosarcoma (Panel D, CD31 staining). Cytology showed dissemination of angiosarcoma into the pericardial cavity.

Primary malignant cardiac masses are extremely rare. They are usually soft tissue sarcomas (angiosarcoma, fibrosarcoma, rhabdomyosarcomas, and mesotheliomas). Angiosarcomas are malignant endothelial vascular neoplasms that grow highly invasive and are often metastasized or display invasion of surrounding structures at presentation. The location is predominantly right sided, usually atrial, opposed to benign tumours, which are usually left sided. Response to chemotherapy is poor and surgical possibilities are limited.

Our patient had no surgical options due to the considerable involvement of right-sided cardiac structures. He refrained from chemotherapy. He needed pericardiocentesis twice more before dying just 2 months after first presentation.

A, atrial appendage; PE, pericardial effusion; PF, pleural fluid; LM, left main coronary artery; RA, right atrium; RCA, right coronary artery; TM, tumour mass; TT, tumour thrombus; TV, tricuspid valve.