Giant primary right ventricular synovial sarcoma

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A 61-year-old man without prior medical history presented with progressive dyspnoea on exertion and right hypochondrial discomfort since 2 months. Clinical examination showed markedly dilated jugular and upper extremity veins without other signs of congestion, congruent with a vena cava superior syndrome. A transoesophageal echocardiogram (Panels A1 and 2) revealed a giant inhomogeneous intracavitary mass with broad insertion to the interventricular septum, obliterating the dilated right ventricle and extending into the right atrium and pulmonary trunk. Magnetic resonance imaging (MRI) depicted the large (10 × 4.5 cm) lobulated tumour with early and late inhomogeneous gadolinium contrast enhancement (Panels B1 and 2). An endovascular biopsy was performed and morphological analysis was consistent with a diagnosis of a poorly differentiated synovial sarcoma (Panel C). This diagnosis was confirmed with a fluorescence in situ hybridization test, which showed a translocation involving the SYT gene on chromosome 18. Positron emission tomography (PET) depicted the cardiac tumour, but no metastasis (Panel D). Because complete tumour resection was not feasible, palliative chemotherapy with doxorubicin and ifosfamide was started with clear regression of the tumour on MRI (Panels E1 and 2) and disappearance of PET avidity (Panel F) as a result. However, after chemotherapy discontinuation, the sarcoma progressed again.

Primary cardiac synovial sarcoma is an extremely rare malignancy, with a very poor prognosis. Complete macroscopic resection is usually not possible, and maintains a high local recurrence and metastasis rate.

There is no relationship with industry.

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