Right ventricular obstruction by metastatic malignant mixed Müllerian tumour

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A 56-year-old woman was referred to our hospital because of new onset weakness. Surface ECG showed sinus rhythm, right axis deviation, and long QT interval (Panel A). Transthoracic echocardiography revealed the presence of a huge mass (50 × 30 mm) in the right ventricle (RV), attached to the interventricular septum (IVS) and almost completely occluding the inflow and outflow tracts. An RV enlargement and a remarkable IVS displacement towards the left ventricle were observed, both suggesting an RV overload (Panel B). Results from a colour-Doppler examination showed a turbulent flow at the RV inflow tract (Panel C). Peak gradients across tricuspid and pulmonary valves, assessed by continuous-wave Doppler, indicated a severe obstruction to the RV inflow and outflow tracts.

Total body computed tomography with multiplane reconstruction proved that the mass adhered to the IVS and extended above the tricuspid valve, while engaging a great portion of the RV outflow tract (Panels D and E; star indicates the pulmonary artery). Lung metastases and two lower abdomen oval masses infiltrating into the uterus were also detected: the first one (*) was 16 × 11 cm, filled with fatty matter and showing fluid level; the second one (+) was 7.5 × 8 cm, filled with heterogeneous matter and presenting a calcified tooth-like coin (white arrow) (Panels F and G).

Pathological examination of uterus biopsy showed many large epithelioid tumour cells with rhabdoid features, including large eccentric vesicular nuclei, with irregular nuclear contours and prominent nucleoli. Immunohistochemical stains had diffuse strong cytoplasmic reactivity with Vimentin (Panels H and I). Biopsy appearances were consistent with a metastatic, poorly differentiated, malignant mixed Müllerian tumour (MMMT) presenting rhabdoid features. The patient was given cytostatic drugs (cisplatin-based chemotherapy regimen), but she died from an acute heart failure episode a month later.

Myocardial involvement by tumours is a rare condition. However, heart metastases are far more common than primary cardiac tumours. Malignant mixed Müllerian tumour is characterized by an aggressive clinical course, resistance to treatment, and a rapidly fatal outcome. The poor prognosis of this neoplasm makes its histopathological recognition of paramount importance. To the present day, there are no guidelines for the optimal management of patients with ventricular metastases of MMMT.