Undifferentiated Cardiac Sarcoma on the Mitral Valve Mimicking Myxoma

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A 53-year-old male presented with progressive fatigue and dyspnea for 2 months. On admission, echocardiography revealed a leaf-shaped mass anchored to the mitral valve (**Figure A**), which was causing moderate mitral stenosis with an effective orifice area of 1.5 cm² and forward blood flow acceleration with $V_{\max}=2.3$ m/s (**Figure B**). Cardiac magnetic resonance confirmed the mass and found that it moved with the opening and closing of the mitral valve (**Figure C**). The mass measured as 3.1×2.4 cm and showed intermediate T1 signal, high T2 signal, and heterogeneous late gadolinium enhancement (**Figure D**). Myxoma, a benign tumor, was initially suspected. However, single-photon emission computed tomography (PET-CT) scan demonstrated increased glucose metabolism of the mass and strongly suggested malignancy (**Figure E**). Therefore, the patient underwent concurrent mass resection and mitral valve replacement. Hematoxylin-eosin staining revealed obvious atypia and mitotic figures, a mixture of round, fat spindle, and pleomorphic tumor cells (**Figure F**). Immunohistochemistry staining was positive SMA (**Figure G**) and CDK (**Figure H**), whereas CD34, S-100, and MDM2 yielded negative results. This suggested the diagnosis of primary cardiac undifferentiated sarcoma. Chemotherapy was initiated and cranial radiotherapy was added 1 year later due to metastasis to the right temporal lobe (**Figure I**).

Undifferentiated sarcoma originating from the mitral valve is extremely rare.¹ As in the present report, a mass

without obvious signs of invasiveness or pericardial effusion may resemble a benign tumor. Thus, PET-CT scan and biopsy further help to identify the nature of masses on the mitral valve.

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Disclosures

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Ethics Committee

West China Hospital [2016(335)].

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Figure. Transthoracic echocardiogram showing a leaf-shaped mass anchored to the mitral valve (arrow, **A**). Color Doppler imaging shows moderate mitral stenosis with an effective orifice area of 1.5 cm² and forward blood flow acceleration with a $V_{\max}$ of 2.3 m/s (**B**). Cardiac magnetic resonance imaging shows the mass with heterogeneous late gadolinium enhancement (**C, D**, arrows) and PET-CT scan show increased glucose metabolism of the mass (**E**). Hematoxylin and eosin staining (×200) shows obvious atypia and mitotic figures and a mixture of round, fat spindle, and pleomorphic tumor cells (**F**). Immunohistochemical staining (×200) positive for SMA and CDK (**G, H**). Right temporal lobe metastasis detected 1 year after cardiac surgery (**I**).