Endometrial stromal sarcoma – a rare tumor invading the heart

Clinical background
A 47-year-old female patient presented with progressive dyspnea on exertion, abdominal pain, loss of weight and tachycardia. The diagnosis of a retroperitoneal tumor with invasion of the vena cava inferior up to the right atrium was established by computed tomography. The patient was referred for echocardiographic assessment of cardiac involvement before surgery. It is worth noting that the patient had a history of hysterectomy under the histopathological diagnosis of leiomyoma ten years ago.

Findings, diagnosis and discussion
Transthoracic echocardiography revealed a dilated right atrium containing an intracavitary homogeneous dense mass, almost obliterating the right atrium, protruding through the tricuspid valve into the right ventricle. This tumor caused regurgitation and stenosis of the tricuspid valve by mainly diastolic bulging into the right ventricle thus compromising circulation and causing tachycardia. By transoesophageal echocardiography the tumor could be followed up to 45 mm into the vena cava inferior. The postoperative histopathological diagnosis was low-grade endometrial stromal sarcoma. Endometrial stromal sarcomas are rare and represent 0.2% of all uterine malignancies [1]. These tumors show intravascular growth in 54% [2]. Several cases have been reported on cardiovascular invasion of the vena cava inferior and the heart chambers [3]. Accordingly, this neoplasm is one differential diagnosis of intracavitary tumors invading the heart.

An additional finding was a thickened, myxomatous degenerated mitral valve, shown in the apical four chamber view (fig. 1). Both leaflets prolapsed, causing mild mitral regurgitation.

References

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