A 20-year-old woman, whose father had a past medical history of Carney complex (cardiac myxoma and spotty pigmentation of the skin), was referred to our hospital with a necrotic fingertip. Vital signs and physical examination revealed no additional pathological findings. Blood count showed a slight anemia with a hemoglobin of 11.5 g/dL (reference range 12 to 18 g/dL), and the serum level of C-reactive protein was 57 mg/L (reference range 0 to 5 mg/L). 2D and 3D echocardiography revealed a giant left-ventricular tumor originating from the posterior papillary muscle and posterior wall, prolapsing into the left ventricular outflow tract during systole. A second tumor in the left atrium originating from the mitral valve with diastolic prolapse into the left ventricle could be diagnosed (Figure 1, see also Movies 1–3). The patient underwent immediate surgery. As a result of infiltrating tumor growth, a part of the papillary muscle and some chordae had to be resected. The mitral valve was reconstructed using artificial chordae and ring annuloplasty. Histological findings confirmed a diagnosis of cardiac myxoma (Figure 2). In a 1-year follow-up, the patient was free of symptoms, and echocardiography did not show any signs of recurrence. Two years later, however, the patient was readmitted with acute visual loss in the left eye, temporary right-sided hemiparesis, and aphasia. Magnet resonance imaging revealed subacute embolic left-sided middle cerebral artery infarction (Figure 3). Ophthalmologic examination revealed signs of a central retinal artery occlusion (Figure 4). Echocardiography could confirm a relapse of the myxoma in both left atrium and left ventricle (Figure 5, Movie 4). Six weeks later, after reconstitution of the blood-brain barrier...
permeability, the recurrent tumors were surgically resected without serious complications. Histological findings of the recurrent tumors were identical to the initial tumors (Figure 6).

The patient’s hemiparesis and aphasia were regressive in contrast to the persisting left-sided visual loss. The patient was screened for other characteristic features of Carney’s complex: there was neither spotty pigmentation of the skin nor mucosal lentiginosis. There were no clinical signs of Cushing’s syndrome. Thyroid gland sonography was without pathological findings. However, gynecological examination and sonography revealed a large, left-sided ovarian tumor. A left-sided ovariectomy was performed and histological findings revealed a monocystic mature teratoma (Figure 7). Five days after the operation, the patient was discharged home in good condition.

Myxomas can appear either as sporadic isolated condition or as Carney complex, a dominantly inherited disease and a unique multiple endocrine neoplasia syndrome. Patients with Carney complex can present with 1 or more cardiac myxomas at the same time in any intracardiac location, irrespective of sex and age. Individuals who undergo surgical resection of Carney-complex-related cardiac myxomas are at relatively high risk of recurrent myxomas at previously affected sites or other locations, even with adequate surgical margins. However, reports about relapsing tumors in both left atrium and left ventricle are very exceptional. Recurrent myxomas are particularly observed in young patients who have a familial history of tumor or multilocular myxomas. Several gene loci have been identified that might be related to Carney complex. A possible association of ovarian tumors with multiple endocrine neoplasias and their related syndromes, such as Carney complex, Peutz-Jeghers syndrome, von Hippel-Lindau disease, and Cowden’s disease has been observed.

Disclosures

None.

References

Figure 3. Cranial magnetic resonance tomography depicting subacute infarction of left-sided basal ganglia including parts of the frontal operculum. A, T2 weighted image with already marked left-sided middle cerebral artery stroke (hyperintense lesion indicated by white arrow). B, Diffusion weighted image indicating acuity of stroke (white arrow).

Figure 4. Color fundus photograph. A, Normal central retina, right eye. B, Central retinal artery occlusion with open cilioretinal artery and optic disc edema in the left eye. Note the whitening of the macula caused by intracellular edema and subsequent necrosis of the inner retina. The cilioretinal artery supplies a small area of normal appearing retina (dark orange) between optic disc and fovea. Arrows indicate some vessels with discontinuous blood flow.
Figure 5. Echocardiography of the recurrent myxoma. A, Transthoracic parasternal short axis view (white arrow indicates the tumor). B, Three-dimensional transesophageal echocardiography. View from the left atrium on the recurrent atrial myxoma and the reconstructed mitral valve. The tumor appears as lobate structure originating from the atrial wall. LA indicates left atrium; RA, right atrium; AV, aortic valve; MVA, mitral valve annulus; and M, myxoma.

Figure 6. Recurrent myxoma. A, Resectate (left ventricle) with associated endocardium (white arrow). The tissue defect was stabilized by two sutures with felt strips. The ventricular tumor's dimensions were $2.5 \times 1.2 \times 0.9$ cm, and the atrial tumor size was $1.2 \times 0.7 \times 0.3$ cm. B, Histological findings of the recurrent tumors were equivalent to the tumors two years before.

Figure 7. Mature biphasic cystic teratoma. A and B, Macroscopy. The sectioned surface of the large tumor (diameter 12 cm) shows a uniloculated cyst. C, Microscopy, hematoxylin and eosin stain. The tumor is composed of derivates of endoderm (mucinous tissue, intestine-type) and ectoderm (squamous epithelium; not shown).
Recurrence of a Familial Giant Multilocular Cardiac Myxoma in a Patient With Carney's Complex
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