A 33-year-old woman was referred for cardiovascular magnetic resonance imaging to further assess a left ventricular mass found after an echocardiography. Her past medical history was remarkable for multiple basal cell carcinomas and surgical resection of odontogenic cysts, and her family history was negative for cardiac tumors. The patient was in her usual state of health until 6 months before presentation, when she noted increasing fatigue, atypical chest pain, and exertional dyspnea. She also complained of intermittent palpitations without syncope. A transthoracic echocardiography demonstrated a 6×3-cm mass in the left ventricular lateral wall, with deformation of the left ventricular cavity. Left ventricular systolic global function was normal, and there was a trace amount of mitral regurgitation.

The patient underwent cardiovascular magnetic resonance examination with a 1.5 T scanner (Signa CV/i, General Electric, Milwaukee, Wis) with an 8-channel cardiac phased-array coil. Bright blood cine images by steady-state free-precession techniques revealed a 5.5×3.8×4.2-cm mass that was hypointense compared with normal myocardium arising within the lateral wall of the left ventricle (Figure 1). T1-weighted, double-inversion recovery fast-spin echo (black-blood technique) images showed the discrete mass was isointense to slightly hypointense relative to the surrounding normal myocardium (Figure 2). First-pass perfusion images by fast gradient recalled-echo echo-planar technique immediately after a bolus injection of 0.075 mmol/kg gadolinium-diethylenetriamine pentaacetic acid (Magnevist, Berlex Pharmaceuticals, Wayne, NJ) demonstrated a lack of first-pass enhancement (Figure 3), suggesting low tumor vascularity. Delayed enhancement images (Figure 4) were obtained using an inversion-recovery-segmented gradient echo sequence (to null normal myocardium) 10 minutes after gadolinium administration (cumulative dose, 0.15 mmol/kg). There was intense late gadolinium enhancement reflecting an increased extracellular volume of distribution in the left ventricular myocardium. Collectively, the tumor appearance and tissue characteristics were consistent with a fibroma. A diagnosis of cardiac fibroma in association with Gorlin syndrome was made.

The patient subsequently underwent surgical resection of the myocardial mass, which was confirmed to be a cardiac fibroma on pathological examination (Figures 5 to 7). Post-operatively, she developed significant mitral regurgitation necessitating mechanical prosthetic (St Jude Medical, St Paul, Minn) mitral valve replacement without further complications.

Gorlin syndrome is a rare autosomal dominant disorder with complete penetrance and variable expressivity. Its estimated prevalence is 1 in 57,000. It is caused by mutations in the *Patched* gene, which acts as a cell-cycle regulator. The hallmark of this syndrome is the presence of multiple basal cell carcinomas, which may appear early in infancy. Other associated features may include craniofacial, central nervous system, musculoskeletal, and genitourinary anomalies. Approximately 3% of cases are associated with cardiac fibromas, which may present later during adulthood rather than the typical infancy or childhood period.

**Disclosures**

None.

**Reference**

Figure 1. Steady-state free-precession (bright-blood) short-axis and 4-chamber images showing a slightly hypointense mass in the lateral wall of the left ventricle.

Figure 2. Double-inversion recovery fast spin-echo image in the short-axis plane.

Figure 3. First-pass perfusion image in the short-axis plane showing hypoperfusion of the mass surrounded by the normally perfused myocardium, which is suggestive of low tumor vascularity.
Figure 4. Delayed myocardial enhancement images demonstrating high signal intensity of the mass compared with the nullled normal myocardium.

Figure 5. Gross pathology of the resected left ventricular mass. The cut section showed an off-white whorled surface with foci of calcification. The lesion extended to the inked surgical margin. There was no hemorrhage or necrosis.
Figure 6. Hematoxylin & eosin-stained section demonstrating proliferation of bland spindle cells, hyalinized collagen, and focal calcifications. No cytological atypia, necrosis, or mitotic activity was identified. Magnification ×4 (top) and ×10. (bottom).

Figure 7. Desmin immunohistochemical stain. Magnification ×2. At the periphery of the resection, the lesion intermingled with the surrounding myocardium (immunopositive staining with desmin). The tumor cells showed negative staining with desmin, smooth muscle actin, S100, CD34, and C-Kit.
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