Similar to primary localized tumor, we strived for a potential cure and long-term survival with adjuvant mechanical and oncologic therapies. Multidisciplinary platforms were crucial for our ability to move forward with treatment and expedite institutional support. Also critical to this case was the patient's autonomy in the decision-making process.

References

Primary Leiomyma: A Rare Space Occupying Lesion in the Right Ventricle
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A case of primary cardiac leiomyoma arising from the infundibulum of the right ventricle of a 24-year-old woman is presented. The mass protruded into the cavity of the right ventricle and caused severe right ventricular outflow tract obstruction. We resected the tumor under cardiopulmonary bypass successfully, and confirmed it histologically to be a benign leiomyoma. To the authors’ knowledge, this is the first case report of a primary cardiac leiomyoma in an adult woman.


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Primary cardiac tumors are typically rare with an autopsy frequency of 0.0017% to 0.003%, of which 75% are benign, mostly myxoma, followed by lipoma, papillary fibroelastoma, and rhabdomyoma [1]. Location of the benign tumor rather than its histologic type often determined the clinical manifestations [2], which varied from being asymptomatic to severe events such as ventricular outflow tract obstruction, arrhythmia, tumor emboli, valvular or myocardial dysfunction, or even sudden death. Primary tumors should be included in the differential diagnosis of intracardiac space occupying lesions.

A 24-year-old asymptomatic woman presented to our department with a cardiac space occupying mass detected in an echocardiogram at a routine checkup. Physical examination revealed a systolic ejection murmur grade 4/6 at the third intercostal space of the left sternal border but was otherwise unremarkable. She had a prolonged menstrual cycle but denied hypermenorrhea or past surgical history; no genitourinary or pelvic mass was found in ultrasound scan tests.

A transthoracic echocardiogram exhibited a 24 to 28 mm circumscribed mass protruded into the right ventricular outflow tract (Fig 1A). Computed tomography (CT) confirmed the lesion originated from the infundibulum of the right ventricle in close proximity to the right coronary sinus at the aortic root and extended into the right ventricular chamber causing significant right ventricular outflow tract obstruction (Figs 1B, 1C).

Through median sternotomy, right atriotomy and longitudinal infundibulotomy allowed complete visualization of the mass and a patent foramen ovale. There was a boundary between the intact tumor capsule and the infundibulum of the right ventricular myocardium. We generously separated the tumor attachment from the surrounding myocardium until its pedicle was dissected completely without damaging the right coronary sinus wall. The incisions were re-approximated, and the patent foramen ovale was closed. Palpation indicated there was good reduction of the thickness of the protrusion.

The mass was solitary, with intact borders located in the right ventricular outflow tract of the gross examination (Fig 2A). On section its whorl-like surface appeared firm and grayish white in the absence of hemorrhage or focal necrosis (Fig 2B). Immunohistochemical examination showed elongated cells with spindle-shaped nuclei arranged loosely in bundles, and no mitotic activity or atypical cells were found (Fig 2C). Positive staining for smooth muscle actin (Fig 2D) and negative staining for myoglobin strongly suggested that it was an intracardiac leiomyoma.

The patient had an uneventful postoperative recovery. The CT scan and echocardiograms showed no recurrent tumor and normal ventricular function at 4-month follow-up assessment.

Comment
Leiomyomas are the most common type of noncancerous neoplasm in the uterine myometrium in women of reproductive age, but occasionally occur with unusual growth
patterns or in extrauterine sites that make their identification more challenging. In such a case, synchronous uterine leiomyoma or a previous hysterectomy may be indicative of the diagnosis [3]. We concluded that its cardiac involvement, which was not even listed in the extrauterine leiomyomatous classification [3], can be divided into 3 categories.

Intracardiac leiomyomatosis is the direct extension of leiomyoma originating from the uterus into the right cardiac cavities through the inferior vena cava. By the year 2009, fewer than 100 cases of intracardiac leiomyomatosis had described in the literature [4].

Benign metastasizing leiomyoma represent an unusual growth pattern characterized by histologically benign uterine leiomyoma “metastasizing” to the right ventricular cavity from the original uterine leiomyoma, which has been reported in only 2 cases. Galvin and colleagues [5] speculated the etiology was secondary tumor embolism through venous channels at the time of a previous uterine operation.

Primary cardiac leiomyoma seldom, if ever, occurs due to myocardium tissue dominance. Because the secondary intracardiac leiomyomatosis and benign metastasizing leiomyoma exclusively occurred in a situation where a concurrent uterine leiomyoma is presented, the next obvious question is whether primary cardiac leiomyoma will be possible in adult females. Our presenting case seemed to address this question in some ways. The asymptomatic 24-year-old woman, who denied history of solitary genital leiomyoma or reproductive system operation, presented with an intracardiac space occupying lesion. Preoperative imaging and intraoperative gross inspection indicated a benign mesenchymal tumor. Surgery was to resect the entire tumor with achievable sufficient margins and relieve the right ventricular outflow tract obstruction. The tumor specimen had the typical feature of smooth muscle cell fibers but showed no evidence of cellular dysplasia or...
rhabdomyoblastic differentiation. We thus claimed the tumor histologically to be a primary cardiac leiomyoma arising in the layer of vascular endothelial cells, which was consistent with the previous report [6]. Given that the pedicle of the tumor was attached to the infundibulum of the right ventricle we believe that the tumor originated from intramyocardial feeding vessels, representing independent foci of smooth muscle proliferation. We hypothesized that the tumor cells grew and migrated toward the right ventricular outflow tract, where the infundibulum was pushed to form a protrusion with the tumor together. Serial echocardiogram follow-up is mandatory in order to monitor the recurrence of a cardiac tumor and the long-term prognosis.

This is the first case of primary cardiac leiomyoma originating from the infundibulum of the right ventricle, protruding as an intracardiac space occupying lesion of an adult woman, without evidence of intracardiac leiomyomatosis or benign metastasizing leiomyoma. Related reports describe primary cardiac leiomyoma located either in the lateral free wall of the right ventricle or in the ventricular septum of asymptomatic adolescent boys [6, 7]. This presenting case has widened the spectrum of recognizing the intracardiac involvement of extraterine leiomyoma in adult women.

References


We report the case of a young patient who developed paraplegia after CABG without CPB. It is important to point out that this patient’s aorta was not manipulated in any way because the only graft performed was a left internal mammary artery to left anterior descending artery.

Comment

Neurologic lesions are severe postoperative complications of coronary artery bypass graft (CABG) surgery. Among them, paraplegia is one of the rarest, with only a few cases reported. It is almost always related to the use of cardiopulmonary bypass (CPB). Those reports associate medullar ischemia with arterial hypotension, atherosclerotic microembolization caused by aortic clamping, or counterpulsation therapy.

The patient is a 31-year-old female with systemic arterial hypertension, insulin-dependent diabetes, chronic renal failure with hemodialysis for 2 years, and other comorbidities like hypothyroidism, dyslipidemia, mesenteric angina, and diabetic retinopathy, who underwent a PTCA [percutaneous transluminal coronary angioplasty] with stenting to the left anterior descending artery and right coronary artery. Her clinical presentation consisted of a history of recurrent and severe angina 1 year post-PTCA. Coronary angiography showed 70% restenosis of both stents and lesions of little concern in secondary coronary branches, requiring surgical treatment.

Surgery consisted of a single graft of the left internal thoracic artery to the left anterior descending artery without CPB. The right coronary artery was very thin and had distal vascular bed lesions, unsuited for CABG. The aorta was not manipulated in any way. There were no episodes of significant arterial hypotension intraoperatively. The patient was extubated while still in the surgical room with the use of low-dose noradrenaline. The first postoperative day was marked by a scenario of motor and sensitivity deficit in the lower limbs (muscle strength grade zero), with significant hyporeflexia and unaltered cerebellar and vestibular examinations. The T2-weighted magnetic resonance imaging showed hypersignal foci in the anterolateral areas of the spinal cord, ranging from the T3 to T6 segments, suggesting ischemic etiology (Fig 1). Imaging examinations of the aorta did not show any signs of dissection or other aortic alterations.

During outpatient follow-up the patient presented a partial recovery of motor function, being able to stand up with help after 6 months and to walk slowly after a year. Presently, the patient’s neurologic condition is stable.

Paraplegia After Off-Pump Coronary Artery Bypass Grafting

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