Primary Cardiac Leiomyoma Causing Right Ventricular Obstruction and Tricuspid Regurgitation

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We report the unique case of a primary cardiac leiomyoma originating from the right ventricle and involving the tricuspid valve in a 43-year-old woman. Echocardiography showed a giant mass causing severe pulmonary stenosis and tricuspid valve regurgitation. The patient underwent surgical excision and histologic examination revealed a primary cardiac leiomyoma. To the best of our knowledge only three cases of primary cardiac leiomyoma have so far been reported, and this is the first case of primary cardiac leiomyoma involving the tricuspid valve apparatus.

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Primary cardiac tumors are extremely rare, and epidemiologic data result mainly from autopsy or surgical series. According to the 2015 World Health Organization (WHO) classification of heart tumors, the estimated prevalence in the general population ranges from 0.05% and 0.2% [1]. In one of the largest studies on 323 surgically treated cardiac tumors, ElBardissi and colleagues [2] found most to be of benign neoformation (94%) rather than malignant ones (6%).

In a recent report from our institution on primary cardiac tumors, we described 107 benign neoplasias, 94 myxomas (88%) and 13 nonmyxoma (12%). Among the second group, we diagnosed one leiomyoma [3] that is described in detail in this case report.

Primary cardiac leiomyoma has been reported only in three previous cases. Clinical presentation is related to the mass size, ranging from total asymptomaticity to ventricular outflow obstruction in case of large formations. Primary cardiac leiomyoma is not included in the latest WHO cardiac tumor classification and is not mentioned in cardiovascular pathology specialized textbooks.

A 43-year-old woman presented to a local hospital for congestive heart with dyspnea, palpitation, and syncope; she was promptly transferred to our institution for further diagnostic-therapeutic investigations.

Echocardiography revealed the presence of a neoformation apparently arising from the right ventricle wall producing a severe right ventricular outflow tract obstruction (maximum gradient 90 mm Hg, right ventricle pressure 110 mm Hg) and tricuspid valve regurgitation.

Surgical treatment was mandatory because of the patient's symptoms and mass localization.

Operation was performed through median sternotomy with standard bicaval cardiopulmonary bypass. A multilobulated mass was found arising through a short pedunculus from the right ventricle outflow track and protruding into the ventricular cavity in tight proximity to the anterior tricuspid leaflet, adhering to its ventricular face and to the chords. A second isolated smaller papillary neoformation was intraoperatively detected on a chord. The masses were well capsulated without signs of local spread in the surrounding myocardial tissue. A careful resection of both neoplasm was performed, and they were completely enucleated from the tricuspid valve and the right ventricle myocardium.

The operation was completed with tricuspid valve repair according to De-Vega technique and with chordae reimplantation.

The patient was successfully weaned from cardiopulmonary bypass, and her postoperative course was uneventful.

Echocardiography at discharge showed trivial tricuspid regurgitation without signs of residual masses.

At gross examination, the main mass measured 5.5 × 3.2 × 2.3 cm and presented a smooth and shiny capsule. The cut surface showed a whitish, whorled, and firm tissue with a stiff elastic consistence. The second mass

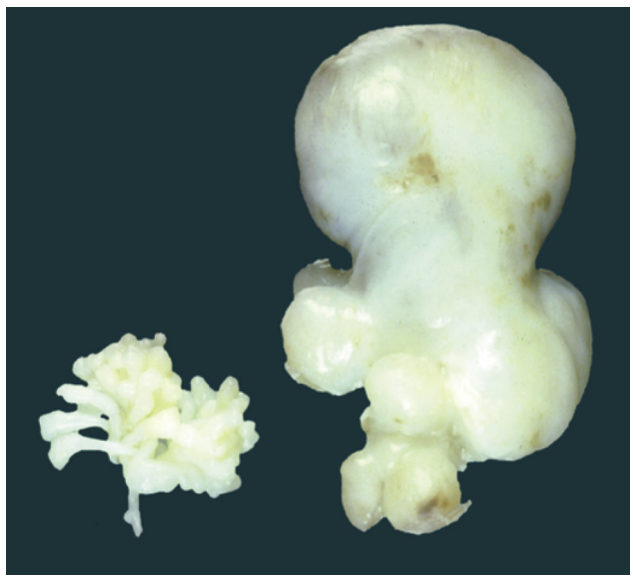

Fig 1. Macroscopic aspect of both cardiac masses. The main mass (right side) shows a smooth and shiny capsule; the smaller one (left side) presents a papillary structure and is inserted on a tendinous chord.

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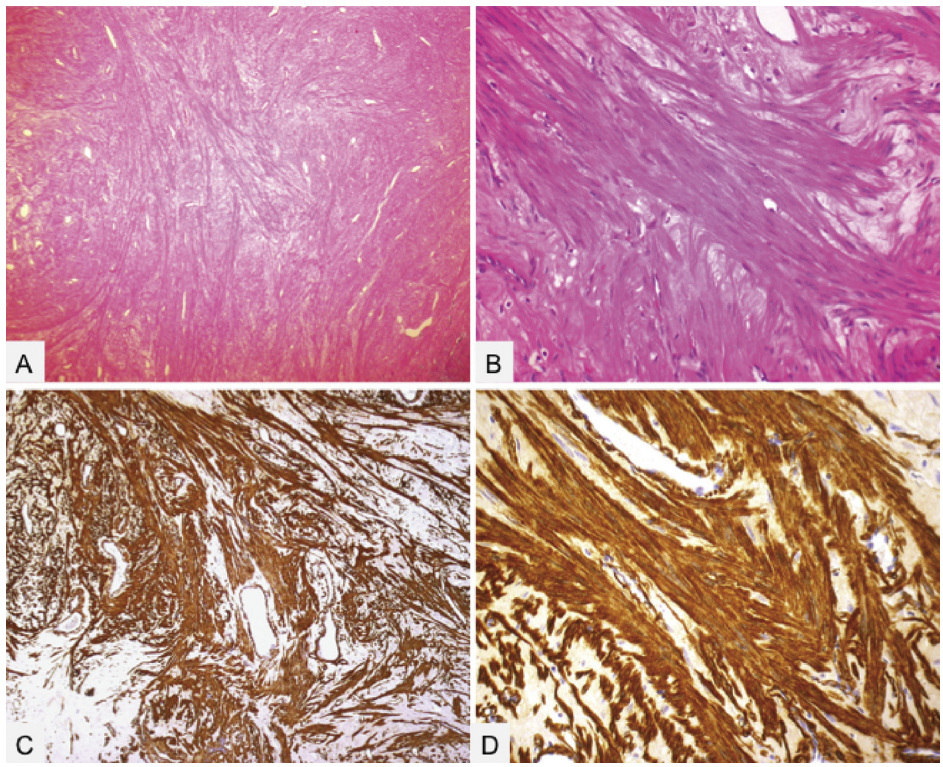

Fig 2. Proliferation of smooth muscle spindle cells with (A) interlacing bundle organization and (B) richly vascularized stroma. (C, D) Immunostaining for smooth muscle actin confirms the smooth muscle nature of the proliferation. (Hematoxylin and eosin; A, ×25; B, ×200; C, ×100; D, ×200.)

measured 1.5 × 2.5 × 0.7 cm and was constituted by the same tissue (Fig 1).

Histologic examination revealed a proliferation of spindle cells with eosinophilic cytoplasm and central nuclei, resembling smooth muscle cells, - organized in interlacing bundles immersed in a loose mesenchymal abundant stroma, richly vascularized. No significant cell atypia or mitoses were found (Figs 2A, 2B). Cell growth showed a proliferative aspect in the absence of an organoid architecture. Vascular component was diffusely distributed and was constituted by both thin and thick wall vessels. The smaller papillary mass, despite the different macroscopic structure, presented the same histologic features.

Immunohistochemistry was positive for smooth muscle actin and desmin (Figs 2C, 2D); the cellular negativity for vimentin ruled out the myofibroblatic nature of the neoplasia.

From both the histologic and immunohistochemistry examination the definite diagnosis was cardiac leiomyoma.

At the 14-year follow-up, the patient was asymptomatic and in good health. Cardiac ultrasound scanning showed a trivial mitral and tricuspid regurgitation and good ejection fraction (71%).

Comment

Cardiac leiomyoma is often secondary to a uterine primary localization. In fact, this benign neoplasia may directly invade the uterine veins, intravenous leiomyomatosis, and extend into right cardiac chambers through the inferior vena cava [4]. However, a few patients with no uterine involvement were identified, reinforcing the hypothesis of intravascular leiomyomatosis resulting from vascular proliferation [5].

Note, our patient had neither a previous history of uterine leiomyomas nor evidence of intravenous leiomyomatosis at the time of the operation.

Primary cardiac leiomyoma has been described in three cases. In these cases the neoplasia originated from the right ventricle wall, creating a right ventricular outflow obstruction [6, 7], or extensively involving the ventricular septum [8]. In our patient, symptoms (dyspnea, palpitation, and syncope) were related to severe right outflow obstruction and tricuspid regurgitation caused by the mass.

Operation was mandatory; subsequently, in the operative room we could appreciate the presence of two masses. In our report, both tumors presented with a smooth and shiny capsule without signs of spread in the surrounding myocardial tissue or other signs of malignancy; for that reason, we decided to not proceed with frozen section. In the presence of undefined capsule or signs of myocardial spread, a frozen section would be immediately performed to exclude malignancy.

Histologic examination subsequently confirmed that both masses were primary cardiac leiomyomas of the right ventricle. Leiomyoma is a neoplasia of smooth muscle tissue; therefore, cardiac leiomyoma originates

most likely from vascular smooth muscle cells present in the tunica media of blood vessels. Interestingly, our case showed involvement of the tricuspid valve apparatus, in particular the smaller mass was located exclusively on a tendinous chord. Although tendinous chords are mainly composed of collagen fibers, some may contain a small central muscle core (chordae muscularis) or sporadic blood vessels. In our opinion, the smaller neoplasia might have originated from these structures evolving to unusual cardiac leiomyoma.

Although a few cases of primary cardiac leiomyoma have so far been reported, its possible occurrence should be acknowledged for the differential diagnosis in cases of right ventricular outflow obstruction and occasionally for the tricuspid valve apparatus involvement. In such cases, surgical option remains the most feasible and appropriate solution for the treatment of heart neoplasms. Primary cardiac leiomyoma is not currently included in the WHO cardiac tumor classification. For the above-mentioned reasons, we believe that this type of tumor should be included in the WHO cardiac tumors classification.

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