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Case report

Intracardiac leiomyomatosis presenting as an intraoperative consultation

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ABSTRACT

Intravenous leiomyomatosis (IVL) is an extremely rare variant of leiomyoma in which nodular masses of tumor grow within venous channels. Rarely, the tumor can reach the vena cava and right heart. We present a case of a 45-year-old woman, admitted with rapidly evolving exertional dyspnea. Cardiac ultrasonography revealed a "mass in the right chambers". She was submitted to right atriotomy plus tumorectomy, with intraoperative consultation requested. Grossly, the tumor was polypoid, firm, with a smooth surface. The frozen section showed a lesion composed of tortuous vessels and some areas with a fibrillar eosinophil extracellular matrix and others with spindle cells, without significant atypia, mitosis or necrosis. The diagnosis was deferred for definitive paraffin sections. In the definitive H&E and immunohistochemical stains, the case was diagnosed as an IVL and confirmed in the hysterectomy specimen. This is the first case report describing an intraoperative consultation of an intracardiac leiomyomatosis. Clinical information and pathologist awareness to this entity are essential for the correct diagnosis in frozen section.

1. Introduction

Primary cardiac tumors are rare entities with an autopsy frequency of 0.001–0.030% [1]. Three-quarters of these tumors are histologically benign, mostly myxomas, followed by papillary fibroelastoma. Metastasis to the heart and pericardium, which are much more frequent than primary cardiac tumors, may reach the heart by direct or transvenous extension. The most common extracardiac tumors that reach the atria and other heart chambers through the vena cava inferior are renal cell carcinoma, hepatocellular carcinoma, leiomyoma of the uterus, nephroblastoma, pheochromocytoma and carcinoma of the adrenal cortex [2]. Cardiac tumors, when found incidentally during surgery and/or at unusual locations, may require intraoperative frozen section diagnosis to decide on further management [3].

We present an unusual case of a patient with rapidly evolving exertional dyspnea in whom a cardiac tumor was detected. The patient was submitted to cardiac surgery, and an intraoperative consultation was requested.

2. Case-report

A 45-year-old woman presented in the emergency room with a 3-week history of easy fatigue and rapidly evolving exertional dyspnea. A cardiac ultrasonography was performed, revealing a solid, hipoecogenic intracavitary mass, which was mobile, apparently friable, starting in the vena cava inferior and progressing to the right heart atrium, through the tricuspid valve to the entry chamber of the ventricle, with no other significant cardiac findings. The possibility of embolization was raised, and an urgent chest, abdominal and pelvic computed tomography CT scan was performed. This exam revealed a heart mass in the right atrium, having 4.5 × 3 cm, with extension to the iliac/femoral vein, and no evidence of pulmonary thromboembolism or lung infarction was

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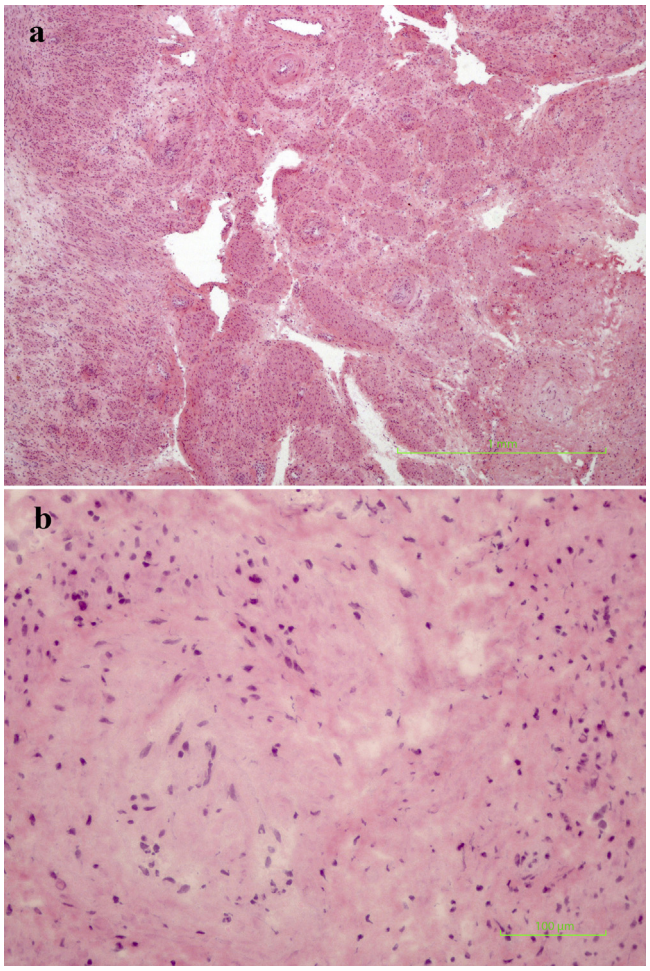

Fig. 1. Frozen section of the intracardiac leiomyoma. (Hematoxylin and eosin, (a)—x40; (b)—x200).

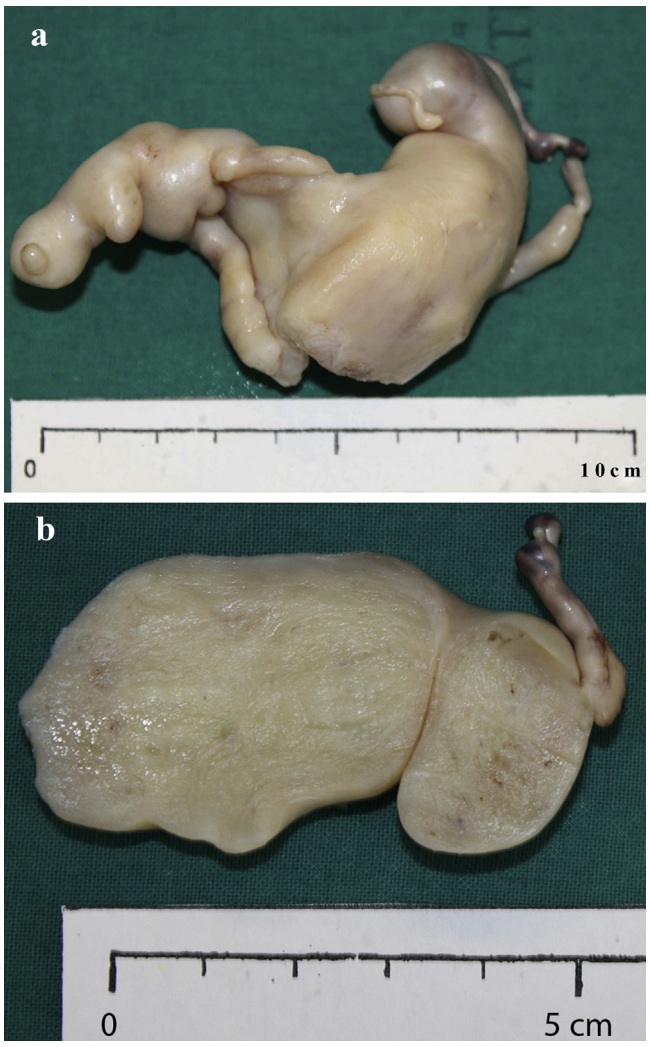

Fig. 2. Gross appearance of the intracardiac leiomyoma. (a) Polypoid with smooth contours and elastic consistency. (b) Fasciculate appearance in cross-section.

found. The patient was submitted to an emergent right atriotomy and resection of part of the tumor.

For intraoperative consultation, two fragments of the tumor were received in our laboratory, the larger one with 1 cm, with the only clinical information given to the pathologist of "right heart tumor". The frozen section examination revealed a cellular proliferation with medium to large caliber vessels and areas of variable cellularity with elongated cells with scant eosinophilic cytoplasm and poorly defined cellular membranes and other areas with fibrillar eosinophilic extracellular matrix. No atypia, mitosis or necrosis were observed (Fig. 1). The intraoperative consultation was signed-out as "tumor with histological features of benignity" with a definitive diagnosis being deferred to paraffin sections. Later on, the heart mass was received (Fig. 2), fixed in 10% buffered formalin and routinely processed. Immunohistochemistry was performed on representative sections with antibodies to smooth muscle actin (Dako, clone 1A4, diluted 1:800, no antigen retrieval), desmin (Leica, clone DERII, diluted 1:60, 15 min heat-induced epitope retrieval solution 1—Bond), estrogen (Leica, clone 6F11, diluted 1:80, 15 min heat-induced epitope retrieval solution 1—Bond) and progesterone receptors (Leica, clone LPGR312, diluted 1:300, 15 min heat-induced epitope retrieval solution 1—Bond). Macroscopically, it was a $10.5 \times 4 \times 2.5$ cm greyish-white elastic tumor with a polypoid, smooth surface and fasciculate appearance in section. Histologically, the tumor was formed by interlacing bundles of smooth muscle with areas of fibrosis and no significant atypia. There was no mitosis or necrosis. Immunohistochemically, the tumor cells stained for smooth muscle actin, desmin, estrogen and progesterone receptors. The diagnosis was of intravenous leiomyomatosis (IVL), given the appropriate clinical context. Later on, a post-operative CT scan was performed (Fig. 3) that showed a vascularized solid mass extending from the renal vein bifurcation of the inferior vena cava to the bifurcation of the common iliac veins and an uterus with multiple solid masses (from a few millimeters to 9 cm) with endovascular extension. Two months later, the patient was submitted to hysterectomy plus adnexectomy. In the uterine body, there were multiple leiomyomas, the largest one with 7 cm. Microscopically, in the external 1/3 of the myometrium, multiple leiomyomas with intravenous extensions were found. These had the same microscopic features as the intracardiac tumor. There were no other significant pathological findings (Fig. 4).

3. Discussion

Intraoperative consultation for heart tumors, despite being able to provide valuable information to the surgeon, is rarely performed [4]. Therefore, beside the limitations inherent to the frozen section technique, there is a lack of experience among pathologists in this particular area of intraoperative consultation [5–7]. As far as we know, this is the first case report describing an intraoperative consultation in a case of IVL, with extension to the heart.