CASE

Atrial fusocellular sarcoma: an uncommon presentation

--Manuscript Draft--

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Bogotá, July 20th, 2022

Vincent L. Sorrell, MD, FACP, FACC, FASE, FSCCT, FSCMR
Editor in chief
CASE – Cardiovascular Imaging Case Reports

Dear Dr. Sorrell

We are pleased to submit the enclosed manuscript entitled “Atrial fusocellular sarcoma: an uncommon presentation” for possible publication in CASE - CASE – Cardiovascular Imaging Case Reports

We submit a case report of a 71-year-old who presented an uncommon type of undifferentiated sarcoma within the right atrium as its primary focus. The authors believe the case presented in this paper is an important contribution to the field.

We declare this manuscript is original, has not been published before and is not currently being considered for publication elsewhere.

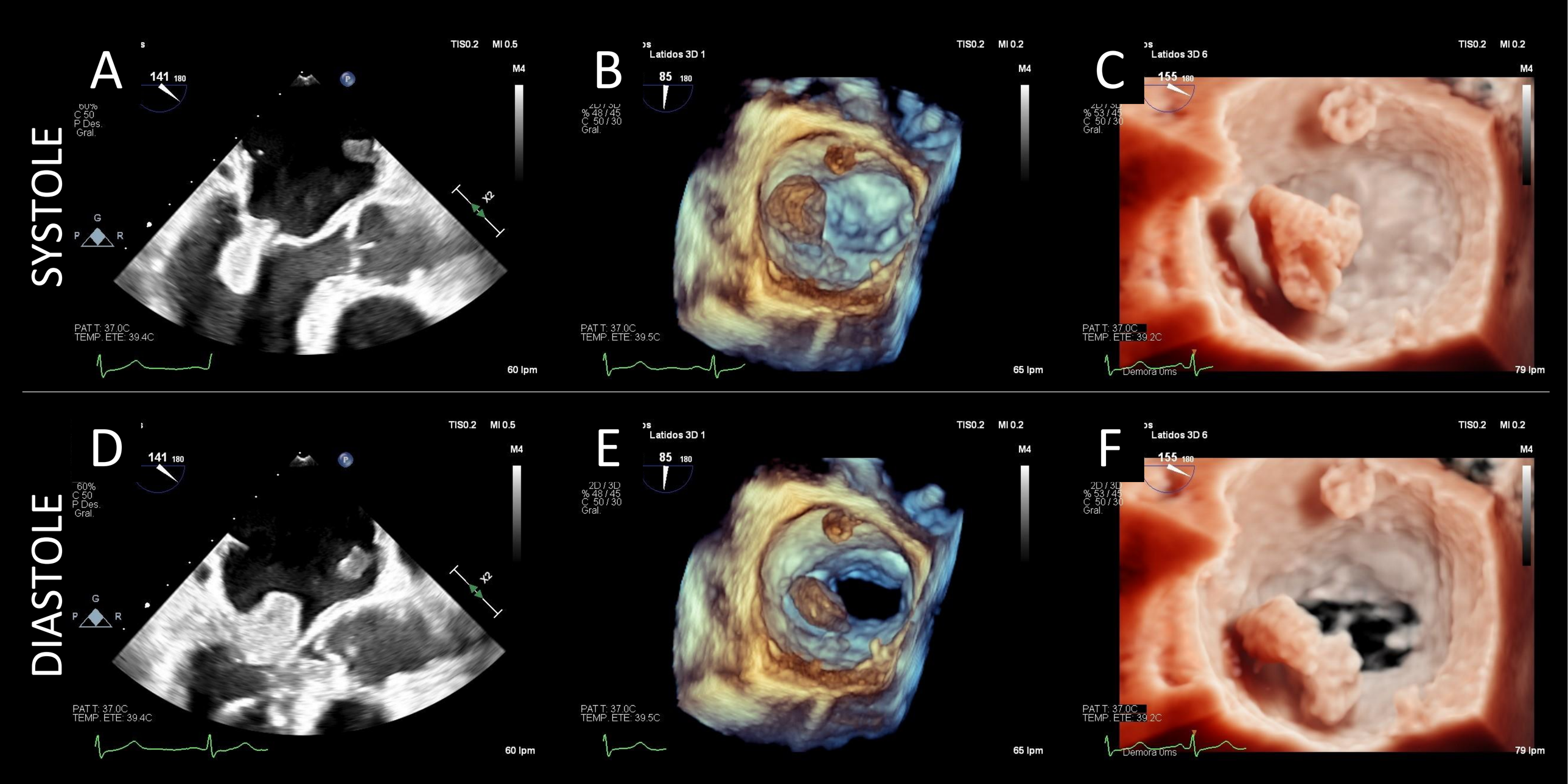
All authors listed have contributed sufficiently to the project to be included as authors. To the best of our knowledge, we do not have any conflict of interest. As the corresponding author, I confirm that the manuscript has been read and approved for submission by all the authors. We take responsibility for the data presented in this manuscript.

Sincerely,

Juan Sebastian Montoya-Beltran

Highlights

- Cardiac tumors are rare but not uncommon.
- There is no definitive treatment available for spindle cell sarcoma
- TTE and TEE are usually the initial imaging modality of this tumors.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Title of article:

Atrial fusocellular sarcoma: an uncommon presentation

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Introduction

Cardiac tumors are an entity with low incidence. Classically they can be divided into non-neoplastic and neoplastic, with the latter further classified as primary and secondary (1,2). Patients with these masses may be asymptomatic or have symptoms related to tumor size and location. Often, these tumors are found incidentally during routine imaging with transthoracic (TTE), transesophageal echocardiography (TEE) or cardiovascular magnetic resonance (CMR)(3,4).

Case presentation

We present the case of a 71-year woman with a previous history of a cardiac mass, first characterized as a classical Hodgkin lymphoma treated with surgical resection and chemotherapy with ABVD protocol (Doxorubicin, Bleomycin, Vinblastine, Dacarbazine), after which they achieved clinical remission. The first pathology reported an angiectasic hyaline pleomorphic tumor CD34 positive, S100 and EMA (epithelial membrane antigen) negative. Five years later, the patient sought medical attention due to fast onset worsening dyspnea and unintentional weight loss. She was found with normal vital signs, and physical examination revealed peripheral edema and normal cardiac auscultation without further abnormal findings. Follow-up TTE revealed a pedunculated mass in the interauricular septum. Further examination with TEE confirmed the recurrence of multiple masses in the left atrium. The primary mass was pedunculated, large (37 X 12 mm) and homogenous with irregular borders. It was attached to the posterior mitral annulus and prolapsed into the left ventricle (LV) in diastole (Figure 1) (Video 1). A smaller, secondary mass was found attached to the interauricular septum (15 x 8 mm) (Figure 2). Mild mitral regurgitation was also observed. At surgery, intraoperative echocardiography confirmed the pre-operative echocardiographic findings (Figure 3). After surgical resection of a round lesion located at the upper portion of the interauricular septum (10 mm in diameter), a second one was found (25 mm in diameter) attached to the mitral valve on its auricular portion at the P2-P3 junction (Figure 4). Histopathology

analysis reported a fusocellular sarcoma (undifferentiated) with MEK1 and CDK4 mutations without microsatellite instability. Lytic lesions on the thoracic vertebrae and focal hepatic masses were also found. Appreciating the fast progression and extent of the disease, palliative care was established, and the patient died one year later.

For this case, a Philips EPIQ CVx® with a probe X5-1 was used for the initial characterization of TTE, and detailed examination through TEE required an X8-2t both on the diagnostic stage and intraoperatively. In addition to previous clinical history, the 2D characteristics of the masses being pedunculated, hyperechogenic and with density disparities with the surrounding tissue suggested malignancy (Video 2). Real-time 3D images obtained in sinus rhythm, apnea and four to six-beat reconstruction allowed a frame rate of up to 70 Hz without stitching phenomenon; these provided helpful information regarding accurate localization of the masses and its relation to the mitral valve (Video 3). TrueVue gave additional characteristics about the auricular tissue surrounding the lesions suggesting local infiltration complementing the information gathered via 2D imaging (Video 4).

Discussion

Cardiac masses are rare but not uncommon. Autopsy findings put their incidence in about 0.001 to 0.003% of the general population; about 25% of these findings are malignant. The malignancy findings are mainly dominated by angiosarcomas, representing 34% of the total malignancies, followed by lymphomas (27%), undifferentiated sarcomas (24%) and mesotheliomas (8%) (5,6). Cardiac fusocellular tumors are rare and aggressive. They have a poor prognosis because of their rapid progression, with a five-year survival rate not higher than 17%, with surgical resection being the only definitive treatment (1,7).

Sarcomas are the most prevalent malignant cardiac tumor in the adult population, primary angiosarcomas being the most frequent within this subgroup. Our case was

abnormal in presentation; an undifferentiated cardiac sarcoma was diagnosed as the primary malignancy. This kind of tumor is grouped within the fibrosarcoma presentation derived from mesenchymal cells and is highly infiltrative. It can appear at any location within the heart chambers.

Sarcoma-type cardiac tumors present clinically in various ways; however, their most common form is pulmonary congestion. As our patient did, they may present with sudden onset, intermittent or positional symptoms of chest discomfort, orthopnea, dyspnea, syncope, or cough. Likewise, it can present as heart failure, arrhythmias, valvular abnormalities, pericardial effusion with tamponade or obstructive symptoms depending on the affected heart valve. On the other hand, thrombotic events can occur that precede systemic embolisms, with the brain and pulmonary system being the most common targets and a characteristic of the clinical presentation (2,5,8,9).

Initially, the diagnosis is made with non-invasive modalities such as TTE or TEE. Echocardiography provides valuable information regarding localization, shape, size, and dynamics of masses that may change clinical and surgical course. CMR allows a better tissue characterization of the mass (10). There is a relationship between the type of tumor and the localization within the heart chambers, with sarcomas being more prevalent in the ventricles and attached to the valvular tissues. The findings of contrast enhancement are considered suggestive of malignancy and highly vascularized tumors.

For this particular case, optimal acquisition of images was paramount; thus, early diagnosis through non-invasive tests allowed surgical planning before further symptoms appeared and were corroborated intraoperatively.

A definitive diagnosis of cardiac sarcomas is made with a biopsy of the affected tissue. The prognosis depends on the specific grade of the tumor, which is based on mitotic activity, the degree of cell differentiation, and necrosis. Poor differentiation and extensive tumor necrosis have a worse prognosis (11). Surgical management

with cytoreduction is the mainstay of treatment (12,13); It works as palliative management to improve symptoms. In some cases, neoadjuvant chemotherapy or radiotherapy may be used to reduce tumor size prior to surgical excision. Finally, despite the handling, the prognosis remains poor, with tumor recurrence being the most common cause of death (10,14).

Conclusions

Currently, there is no definitive treatment for spindle cell sarcoma, a sporadic cardiac tumor. Primary cardiac sarcoma has a poor prognosis and is usually asymptomatic until advanced stages, and due to its aggressive course, the therapeutic approach is limited and case-specific. Echocardiography is usually the initial imaging modality since it is non-invasive and widely available.

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Captions

- **Figure 1:** Mid-Esophageal long axis view with mitral zoom (A) Systole (B) Diastole. *M1: Larger mass, M2: Smaller mass, LA: Left atrium, LV: Left*

ventricle, LVOT: Left ventricle outflow tract, Ao: Ascending Aorta.

- **Figure 2:** Live 3D reconstruction in surgeon's view with two masses (A) Systole (B) Diastole. *M1: Larger mass, M2: Smaller mass*
- **Figure 3:** Live 3D TrueVue Reconstruction in surgeon's view (A) Systole (B) Diastole. *M1: Larger mass, M2: Smaller mass*
- **Figure 4:** Intraoperative findings of the surgically removed pieces related to the echocardiographic discoveries.
- **Video 1:** Mid-Esophageal 4 chambers view with mitral zoom; pedunculated mass on the lateral aspect of the atrial wall and mitral annulus protruding towards the left ventricle.
- **Video 2:** Mid-Esophageal long axis view with mitral zoom; pedunculated mass related to the posterior leaflet of the mitral valve and protruding towards the left ventricle.
- **Video 3:** Live 3D reconstruction in surgeon's view with two masses, the main one on the atrial aspect of the P2 annulus protruding towards the left ventricle; the second pedunculated mass in the anterior wall of the left atrium.
- **Video 4:** Live 3D TrueVue Reconstruction in surgeon's view with the previously described masses.

Figure 1

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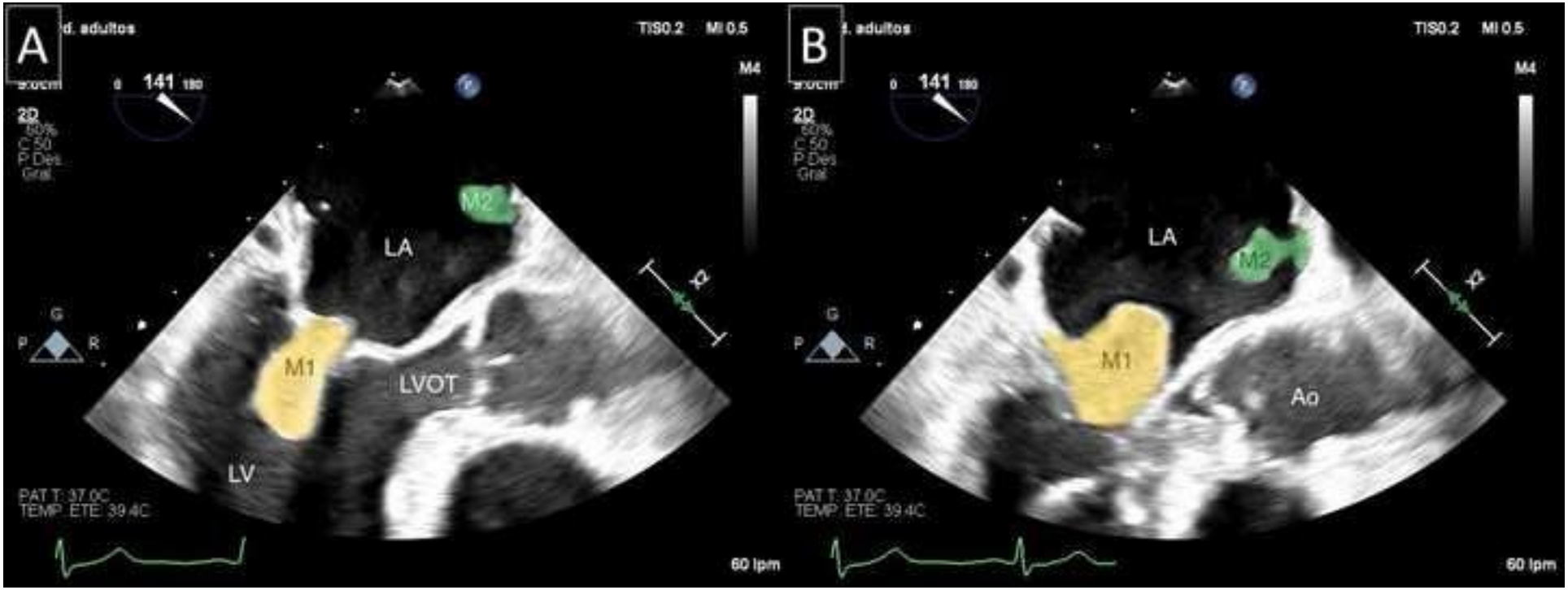

Figure 2

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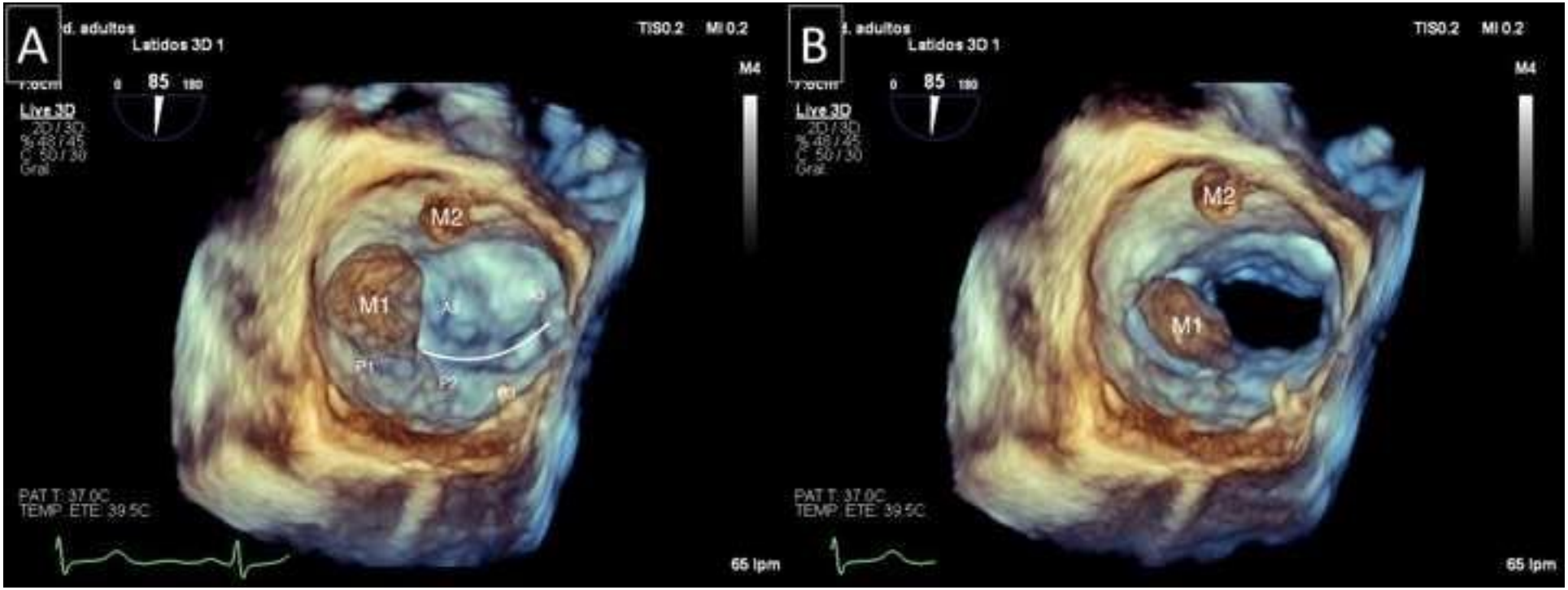

Figure 3

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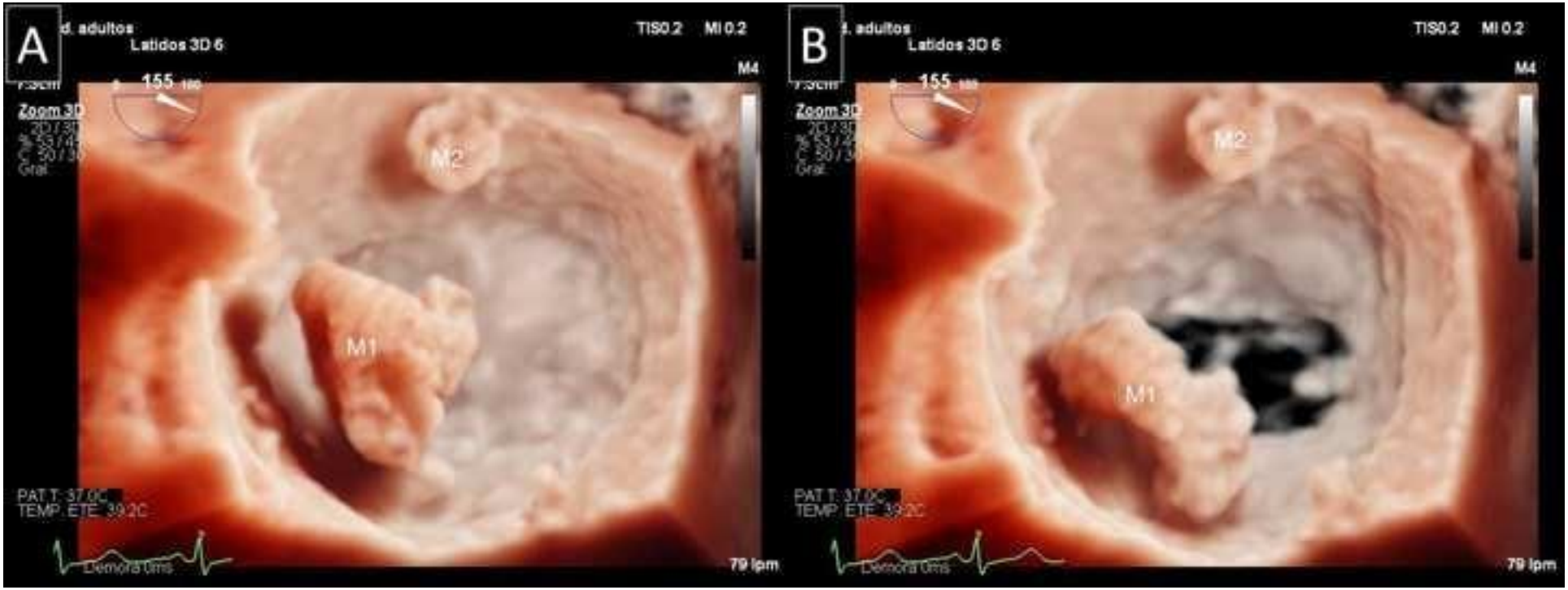

Figure 4

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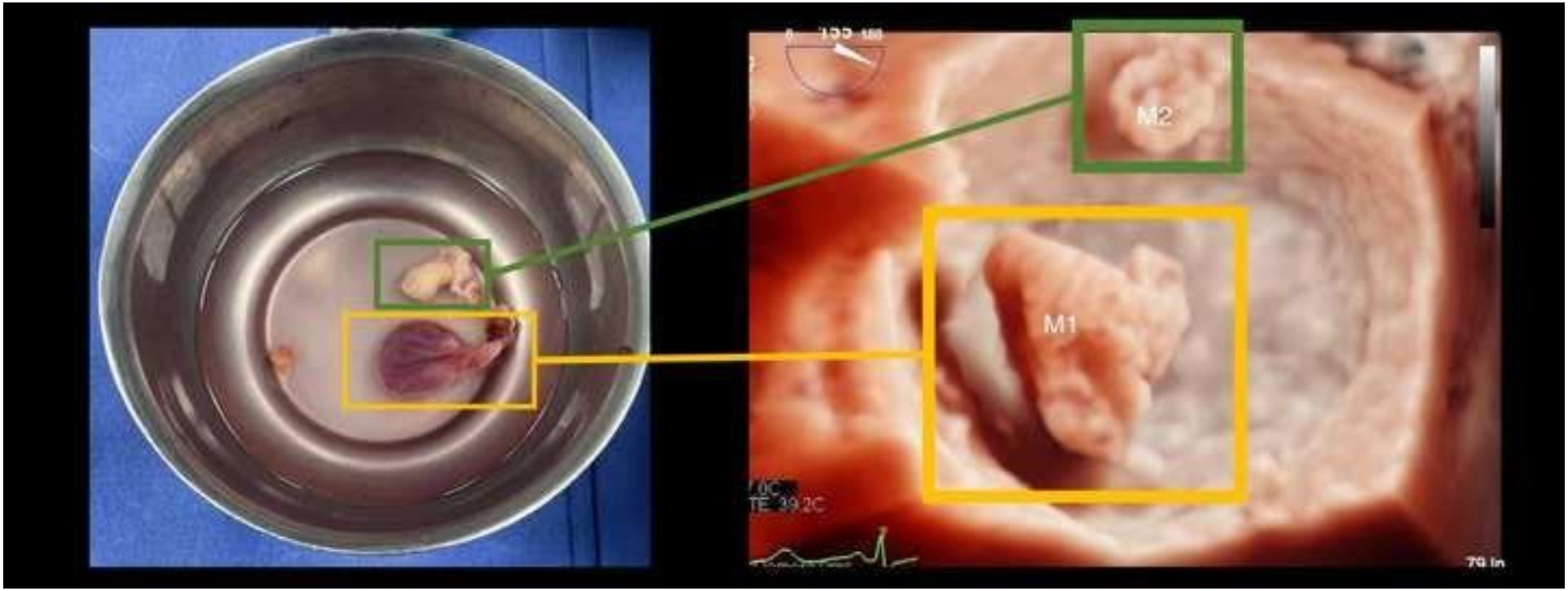

Bogotá, July 20th, 2022

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We declare this manuscript is original, has not been published before and is not currently being considered for publication elsewhere.

All authors listed have contributed sufficiently to the project to be included as authors. To the best of our knowledge, we do not have any conflict of interest.

Sincerely,

Juan Sebastian Montoya-Beltran

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