

CASE REPORT

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Large ventricular myxoma causing inflow and outflow obstruction of the right ventricle; A Case Report

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Abstract

Background Myxomas are the most common primary benign heart tumors, typically found in the left atrium, with only 2–4% occurring in the right ventricle. Clinical presentations vary widely, including congestive heart failure and systemic embolic phenomena. This case report describes a rare right ventricular myxoma causing both inflow and outflow obstruction, presenting as progressive exertional dyspnea.

Case Presentation A 23-year-old male presented with two weeks of worsening exertional dyspnea. He was stable but tachypneic with a systolic murmur over the tricuspid area. Elevated erythrocyte sedimentation rate (ESR) and C-Reactive protein (CRP) were noted, while other lab tests were normal. Imaging, including echocardiography and chest tomography scan (CT) revealed a 4 × 3.8 × 4.6 cm mass in the right ventricle extending to the pulmonary trunk. Surgical resection via right ventriculotomy was performed, and histopathology confirmed myxoma. The patient recovered uneventfully.

Conclusion Right ventricular myxomas, though rare, can cause significant obstruction and present with diverse symptoms. Timely diagnosis using imaging techniques like echocardiography is crucial. Surgical resection remains the definitive treatment, offering excellent outcomes and low recurrence rates. Early intervention is vital to prevent serious complications and ensure favorable patient prognosis.

Keywords Myxoma, Cardiothoracic, Palestine, Surgery

Introduction

Myxomas are the most common primary benign cardiac tumors. Most commonly, they arise from the left atrium. In 20% of the cases, they arise from the right atrium. However, rarely, in 2–4% of cases, they occur in the right ventricle. The presentation of these cases is usually non-specific and can vary widely. Symptoms range from congestive heart failure and atrial arrhythmias to systemic findings such as experiencing embolic phenomena in either the pulmonary or systemic circulation [1, 2]. However, Patients may remain asymptomatic or suffer from mild shortness of breath. The majority of patients

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are young, typically between 30 and 60 years old, with a slight female predominance [2].

This case report describes a patient presenting with progressive exertional dyspnea and was found to have a cardiac myxoma in the right ventricle obstructing both the inflow and outflow tract and extending to the base of pulmonary trunk.

Case presentation

A 23-year-old male presented with a two-week history of worsening exertional dyspnea. His past medical history was unremarkable except for left calf pain and tenderness one year prior to presentation.

On examination, the patient was stable but slightly tachypneic with a respiratory rate of 20 breaths per minute. His blood pressure was 130/80 mmHg, and he had a regular heart rate. A systolic murmur was heard along the left sternal border over the tricuspid area, with normal S1 and S2 heart sounds. The rest of the physical examination was unremarkable.

Laboratory tests revealed a slightly elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels. Thrombophilia studies (including protein C, protein S, anti-thrombin, factor V Leiden, and anti-cardiolipin) and tumor markers (including alpha-fetoprotein, carcinoembryonic antigen, cancer antigen 19–9, cancer antigen 125, and cancer antigen 15–3) were normal.

An Electrocardiogram ECG and chest radiography were normal. Doppler ultrasound of both extremities revealed an old resolved deep vein thrombosis (DVT). The patient was treated with low molecular weight heparin. Several days later during treatment, he experienced shortness of breath and easy fatigability, associated with low oxygen saturation (SaO₂ 87%). Consequently, the patient was admitted to the Coronary Care Unit (CCU) for further evaluation.

Transthoracic echocardiography (TTE) revealed an oval soft tissue mass measuring 3.2×2.8 cm attached to

the right ventricular wall via a wide short pedicle. The right ventricle was mildly dilated and associated with mild tricuspid valve regurgitation. The ejection fraction was 55% (Fig. 1).

A whole-body CT scan with Intravenous (IV) contrast confirmed the presence of a large, well-defined, oval-shaped hypodense heterogeneously enhancing mass in the right ventricle, measuring approximately 4×3.8×4.6 cm. The mass extended to the base of the pulmonary trunk, with no evidence of pulmonary embolization. These findings were suggestive of an intraventricular myxoma (Fig. 2A, B,C).

Given the previous findings, the cardiothoracic surgeons decided that surgery is necessary. To gain access to the heart, a median sternotomy incision was made, and a total cardiopulmonary bypass was instituted. Three cannulas were put in place, one in the ascending aorta, another in the superior vena cava, and the third in the inferior vena cava. The cross clamp time was 53 min, total pump time was 84 min, and a systemic temperature of 32 degrees Celsius was maintained using warm fluids and adjusting room temperature, cold cardioplegia was used. The mass was approached through a right atriotomy, and the tricuspid valve leaflets were found to be normal. Due to difficulty in excising the mass, a right ventriculotomy was performed. The mass was occupying the right ventricular chamber and was attached to the septal chordae tendineae of the tricuspid valve and the posterolateral wall of the right ventricle through a wide (2 cm) fibrotic base (Fig. 3). The tendinous chordae which were excised, were secondary (attached to the ventricular surface of the septal leaflet). Only one primary tendinous chordae (which was attached to the margin of the septal leaflet) was cut.

Dissection of the mass from the right ventricular wall and chordae tendineae was performed, and complete excision of the mass was achieved. The pulmonic valve was clear, and the tricuspid valve was tested and found to be normal. The patient was weaned off bypass smoothly,

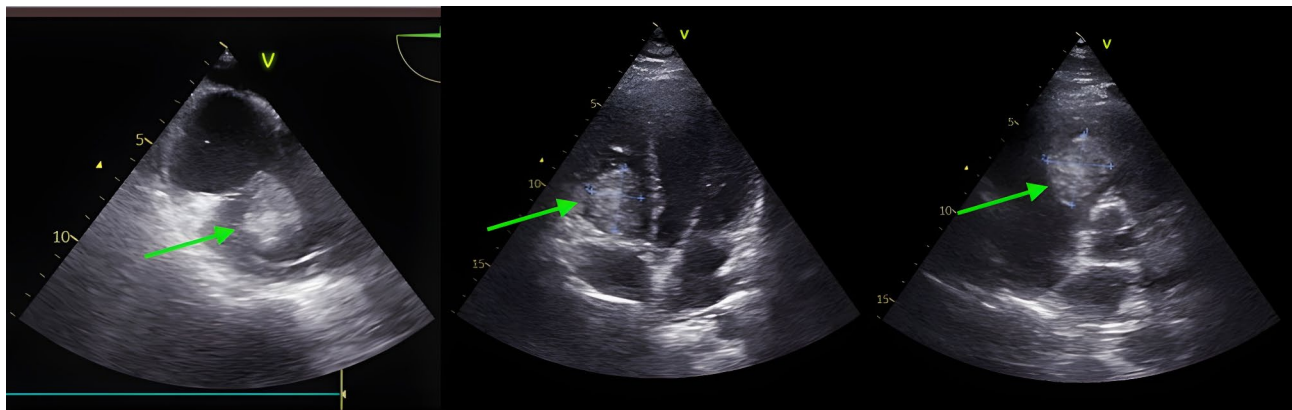


Fig. 1 TTE showing an oval soft tissue measuring 3.2×2.8 cm attached to the right ventricular wall via a short pedicle (Green arrow)

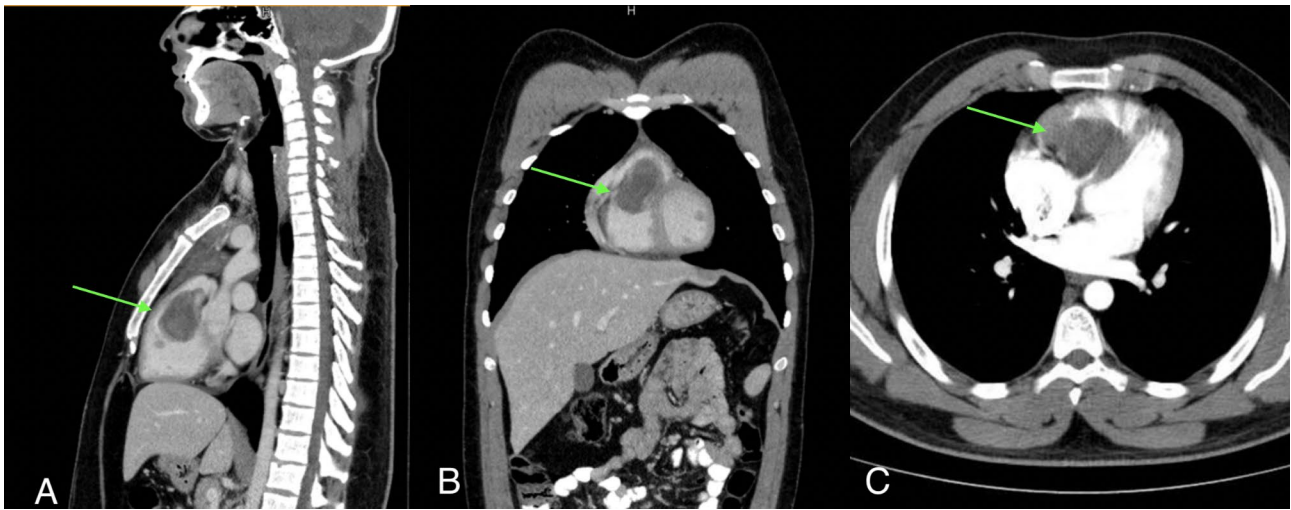


Fig. 2 CT scan with IV contrast showing the presence of a large, well-defined, oval-shaped hypodense heterogeneously enhancing mass in the right ventricle, measuring approximately $4 \times 3.8 \times 4.6$ cm (Green Arrow) (A) Sagittal Plane (B) Coronal Plane (C) Axial Plan

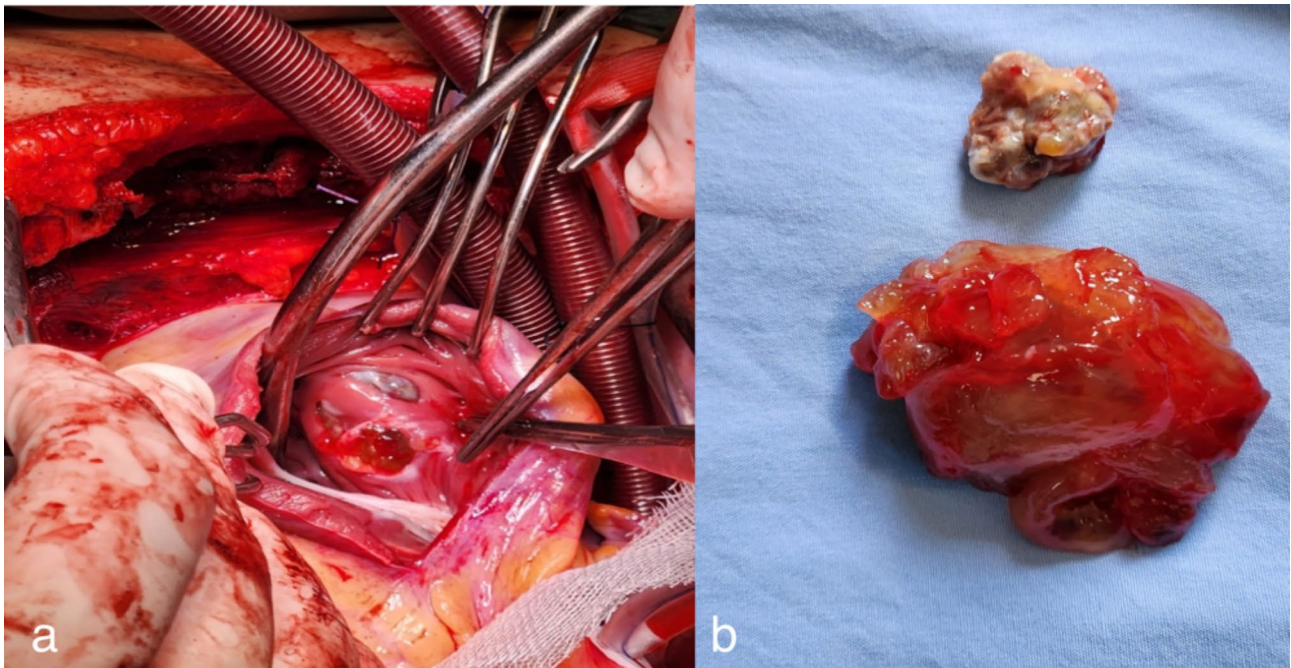


Fig. 3 (a) Mass occupying the right ventricle. (b) excised mass along with chordae tendinae

and the postoperative course was uneventful. The diagnosis of myxoma was confirmed by histopathology.

At the most recent follow-up, 10 months after the initial treatment, the patient is in good health with no exertional shortness of breath, pain, or other complaints. A follow-up echocardiogram (Fig. 4) showed no residual masses, with an ejection fraction of 55%. All heart valves, including the pulmonic and tricuspid valve, showed no signs of insufficiency, regurgitation, or stenosis.

Discussion

Myxomas are the most common type of primary cardiac tumor among adults, though they are rare. They typically occur in the left atrium (75% of cases), while 20% arise in the right atrium, and only 2–4% occur in the right ventricle. Most cases are sporadic, with only 10% being familial with autosomal dominant transmission.

Myxomas are mesenchymal tumors with a gelatinous appearance, consisting of myxoma cells embedded in a stroma rich in glycosaminoglycan. Most myxomas have a smooth surface, though one-third have a friable or

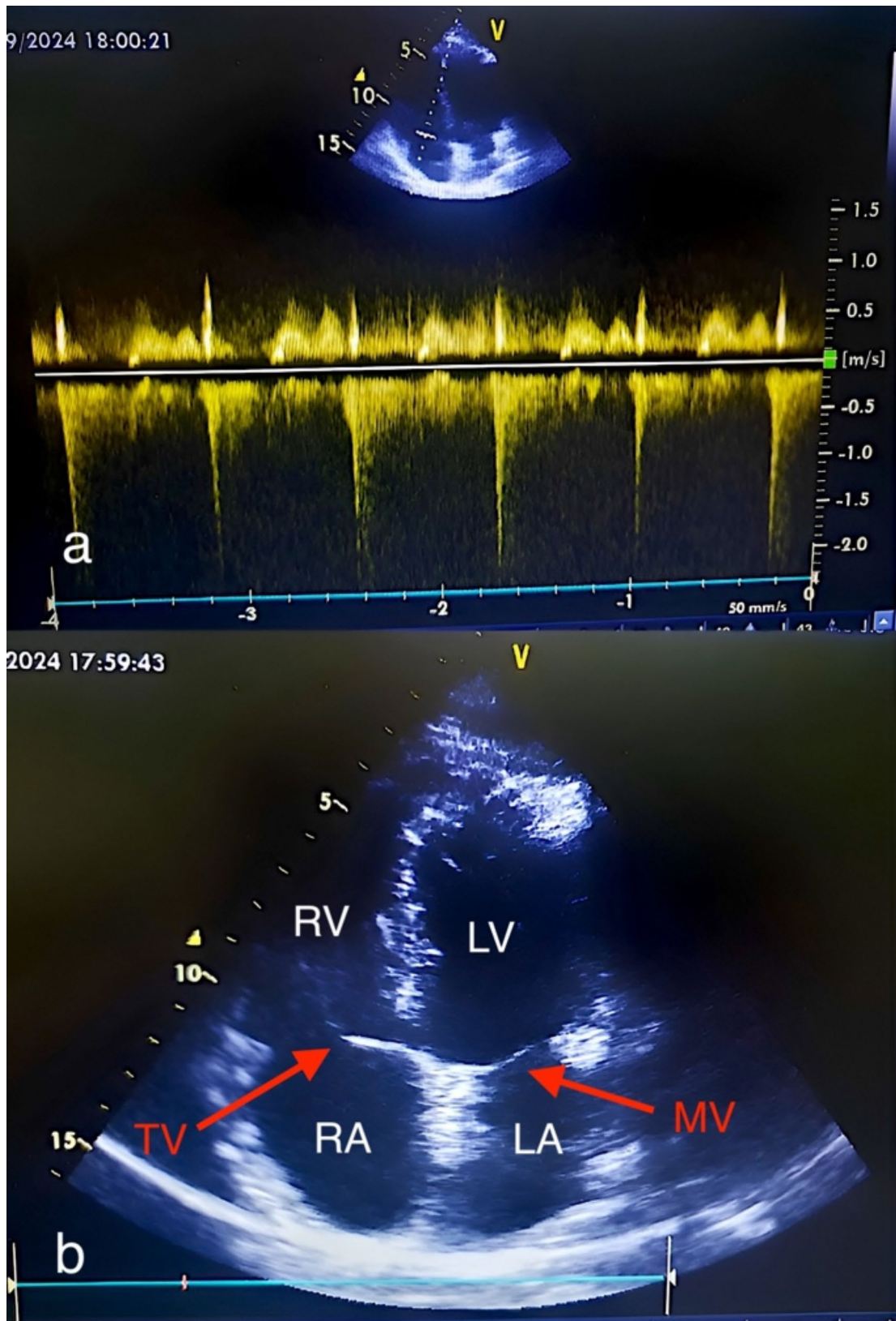


Fig. 4 (a) normal tricuspid valve flow velocity showing no signs of valvular insufficiency or stenosis. (b) TTE showing no residual masses villous surface, which carries a high risk of embolization [2]. They vary in size, ranging from small (less than 1 cm

in diameter) to massive (up to 10 cm across). Their morphology also differs: two-thirds of cases are pedunculated masses that may swing through the mitral or tricuspid valve during systole, causing obstruction. The remaining cases are sessile, with a higher risk of embolization due to their fragile nature [3].

The presentation of myxomas varies widely, depending on the location and the size of the tumor. They can be asymptomatic, or present with constitutional, obstructive, or embolic symptoms. The obstructive symptoms include dyspnea, syncope, and other systemic circulation ischemia related symptoms in left-sided myxoma, whereas in right-sided myxomas, the symptoms are shortness of breath as the case of our patient, syncope, fatigue, or related to right heart failure and systemic congestion as peripheral edema, ascites, or superior vena cava syndrome [4, 5]. The embolic symptoms depend on the site of myxoma, in left-sided myxomas they cause peripheral emboli, such as stroke, while in right-sided ones, they can cause pulmonary embolism.

Transthoracic and transeophageal echocardiogram represent the first best non-invasive imaging modality that help in the diagnosis of myxoma, it allows for assessment of the tumor size, the site of attachment, and potential invasion of valves [2].

Surgical resection is the definitive treatment for myxoma [2]. Once the diagnosis is suspected on imaging, it should not be delayed, as there is a risk of sudden death or developing other severe complications such as stroke [4]. Pathological diagnosis is necessary to definitively diagnose myxomas and differentiate them from malignant neoplasms, as imaging modalities alone cannot rule out malignancy [4]. The recurrence rate after surgical resection is 2–5% in sporadic cases and 12–22% in familial cases [2].

Conclusion

Myxomas are the most common benign primary cardiac tumors, typically arising in the left atrium but rarely can occur in the right ventricle. We reported in this case a rare location of cardiac myxoma in the right ventricle obstructing both the inflow and outflow tract and extending to the base of pulmonary trunk. clinical presentations for myxomas vary widely, emphasizing the need for timely diagnosis and surgical intervention when indicated. Imaging, especially echocardiography, is pivotal for initial assessment, guiding treatment decisions by evaluating tumor characteristics. Surgical resection remains the cornerstone of treatment, offering a curative approach with low recurrence rates, supported by histopathological confirmation to exclude malignancy. Early intervention is crucial to mitigate risks such as stroke

and ensure favorable outcomes in managing this rare, yet clinically significant, condition.

Abbreviations

ESR	Erythrocyte sedimentation rate
CRP	C Reactive Protein
CT	Chest Tomography
ECG	Echocardiogram
DVT	Deep vein thrombosis
SaO ₂	Oxygen Saturation
CCU	Coronary Care Unit
TTE	Transthoracic Echocardiography

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None.

Author contributions

Concept and design was done by A.D and Y.Y. Acquisition, and interpretation of data were done by Y.Y and I.R. Drafting of the manuscript was done by F.A, R.S, A.H, D.Y and M.H. Critical review of the manuscript for important intellectual content was done by M.H and Y.Y. Revision and formatting of the manuscript was done by M.H. Supervision was done by A.D and Y.Y.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Consent for publication

Written informed consents were obtained from the patient for publication of this case report and accompanying images.

Competing interests

The authors declare no competing interests.

Ethical approval and consent to participate

This study was conducted in accordance with the Declaration of Helsinki. Informed written consents were acquired from the patient prior to their enrollment in this study.

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References

1. Katiyar G, Vernekar JA, Lawande A, Caculo V. Cardiac MRI in right ventricular outflow tract myxoma: Case report with review of literature. *J Cardiol Cases* [Internet]. 2020;22(3):128–31. <https://linkinghub.elsevier.com/retrieve/pii/S187854092030044X>
2. D'Anna C, Villani A, Ammirati A, Francalanci P, Ragni L, Cecconi G et al. New Onset Cardiac Murmur and Exertional Dyspnea in an Apparently Healthy Child: A Rare Localization of Obstructive Myxoma in the Right Ventricular Outflow Tract without Pulmonary Embolization-A Case Report and Literature Review. *Int J Environ Res Public Health* [Internet]. 2022;19(19). <http://www.ncbi.nlm.nih.gov/pubmed/36232202>
3. Islam AKMM. Cardiac myxomas: A narrative review. *World J Cardiol* [Internet]. 2022;14(4):206–19. <http://www.ncbi.nlm.nih.gov/pubmed/35582466>
4. Lu C, Yang P, Hu J. Giant right ventricular myxoma presenting as right heart failure with systemic congestion: a rare case report. *BMC Surg* [Internet]. 2021;21(1):64. <https://bmc Surg.biomedcentral.com/articles/https://doi.org/10.1186/s12893-020-00977-4>

5. Gajjar TP, Shah GB, Desai NB. Mixoma ventricular direito gigante obstruindo via de saída do ventrículo direito. *REVISTA BRASILEIRA DE CIRURGIA CARDIOVASCULAR* [Internet]. 2011;26(4):663–6. <http://www.gnresearch.org/doi/https://doi.org/10.5935/1678-9741.20110060>

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