

## Case Report

# Cardiac Myxoma Arising From the Superior Vena Cava: A Case Report

Rezvanieh Salehi<sup>1</sup>, MD; Maryam Chenaghlu<sup>1</sup>, MD; Asghar Mohamadi<sup>2\*</sup>, MSc;  
Elnaz Javanshir<sup>1</sup>, MD

### ABSTRACT

Myxomas, though uncommon in the general population, comprise the most common cardiac tumors. The majority of these tumors are located in the left atrium (75%), followed by the right atrium (20%), the right ventricle (8%), and the left ventricle (3%–4%). Less common forms involve all cardiac chambers, pulmonary vessels, and mitral and aortic valves. Cardiac myxomas arising from the superior vena cava (SVC) constitute an extremely rare presentation of these tumors. The involvement of the SVC in cardiac myxomas is generally secondary to expansion from the right atrium; nonetheless, an SVC origin with the secondary involvement of the right atrium is extremely rare. Herein, we describe a young man presenting with syncope. Further evaluations revealed a mass originating from the SVC with myxoma pathology. (*Iranian Heart Journal 2022; 23(3): 131-134*)

**KEYWORDS:** Myxoma, SVC, Cardiac tumor

<sup>1</sup>Cardiovascular Research Center, Tabriz University of Medical Sciences, Tabriz, IR Iran.

<sup>2</sup>Cardiovascular Research Center, Shahid Rahimi Hospital, Lorestan University of Medical Sciences, Khorramabad, IR Iran.

\* **Corresponding Author:** Asghar Mohamadi, MSc; Cardiovascular Research Center, Shahid Rahimi Hospital, Lorestan University of Medical Sciences, Khorramabad, IR Iran.

**Email:** asgharheart@gmail.com

**Tel:** +989106042707

**Received:** January 30, 2021

**Accepted:** March 10, 2021

Cardiac myxomas constitute one of the most frequent heart tumors, with an incidence rate of 0.5 per million population per year.<sup>1</sup> About 80% of cases involve the left atrium, 7% to 20% involve the right atrium, and the rest involve both atria.<sup>2-4</sup> Cardiac myxomas are considered benign tumors with an endocardial origin and slow proliferation.<sup>5</sup> The clinical manifestations of myxomas include palpitations, chest pain, coughs, syncope, myocardial infarction, heart failure, and cerebral embolism.<sup>6</sup> Transthoracic echocardiography is considered a diagnostic tool for cardiac myxomas, although transesophageal echocardiography can also

be used for diagnosis.<sup>7</sup> Surgical intervention is the treatment of choice for cardiac myxomas.<sup>8</sup> The extracardiac involvement of myxomas is very rare.<sup>9</sup>

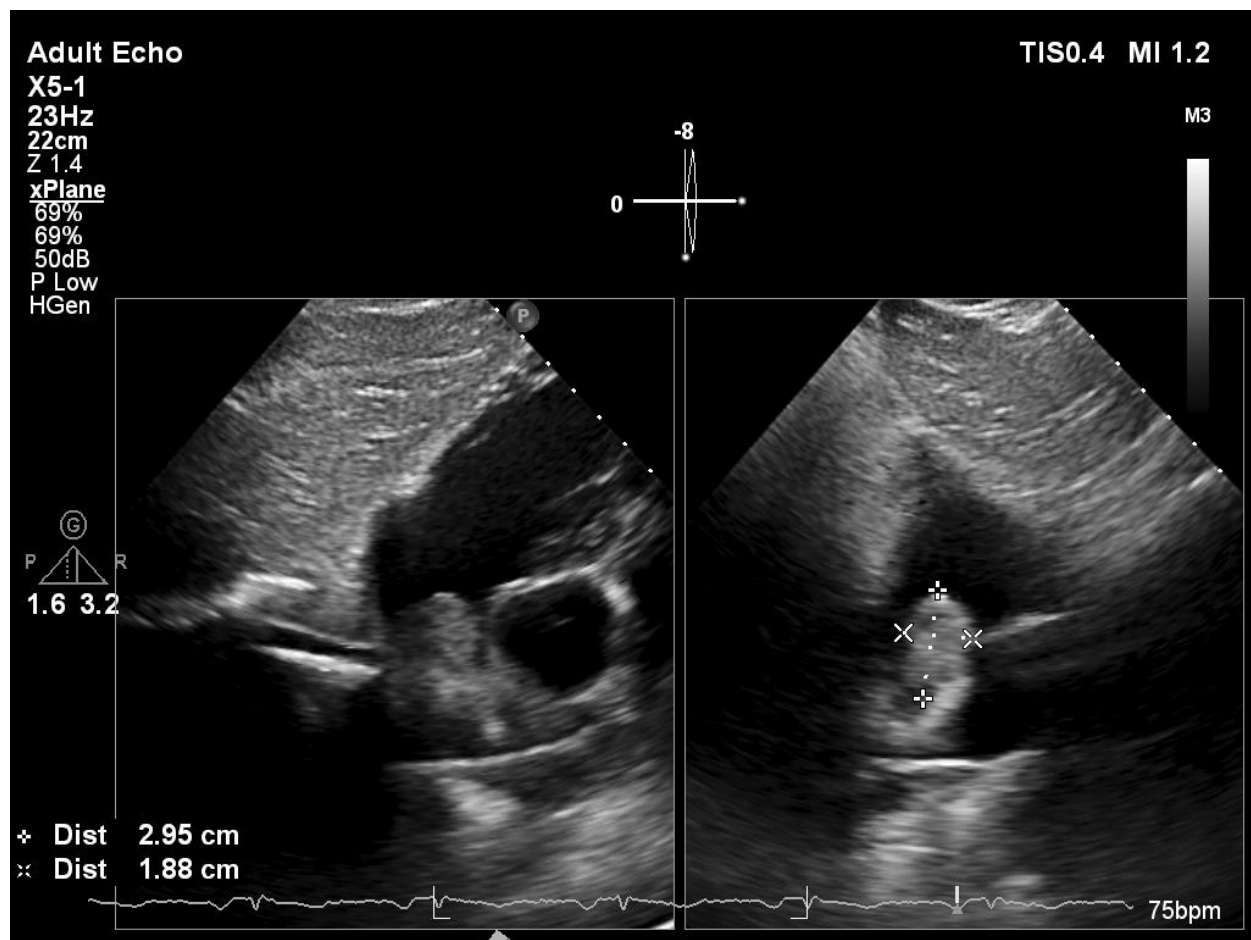
We herein report a very rare case of a cardiac myxoma arising from the superior vena cava (SVC).

### Case Report

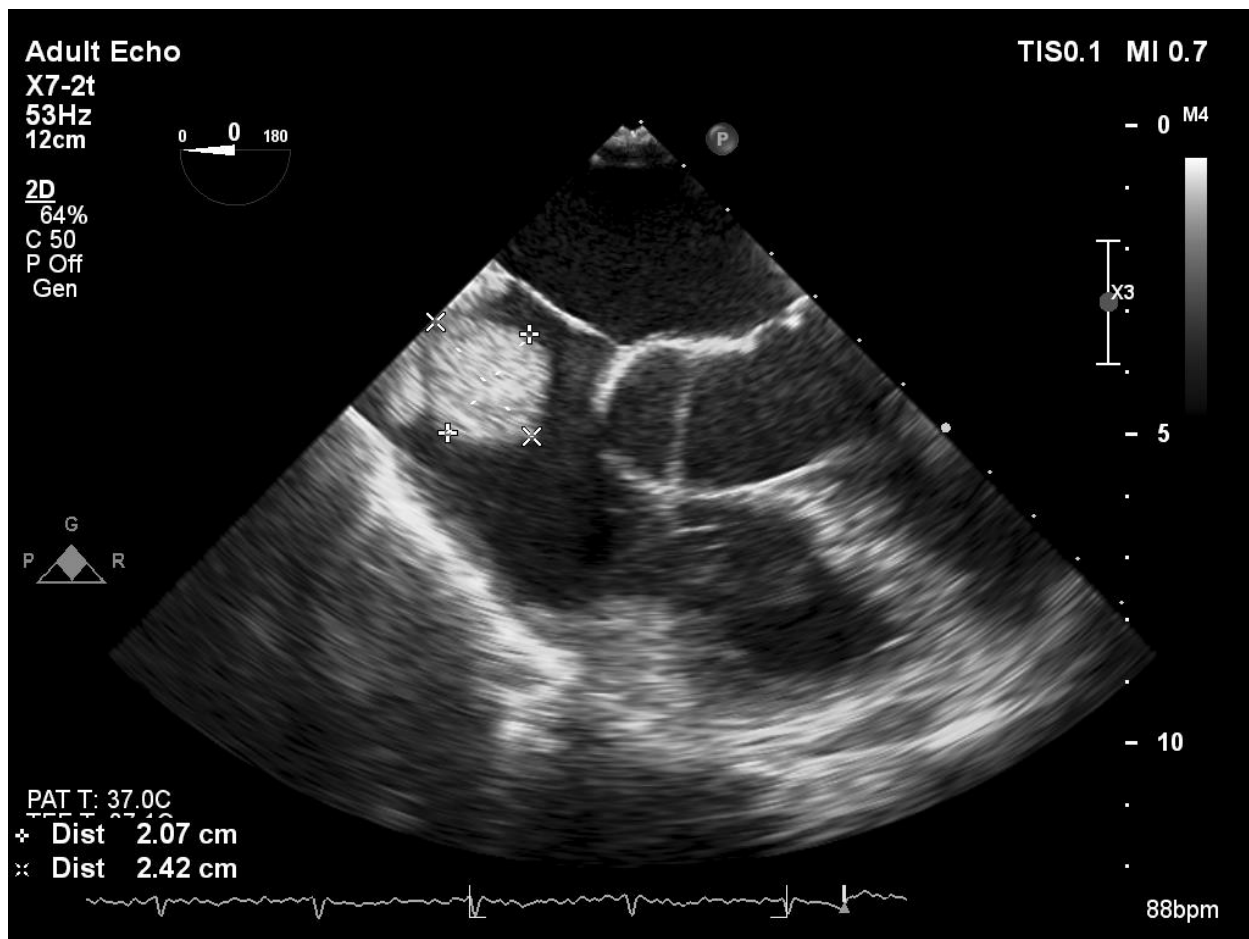
A 33-year-old man came to our emergency department following a fall in the playground. A physical examination revealed pallor, a blood pressure of 90/60 mm Hg, and a heart rate of 100 bpm with regular rhythms. The patient underwent transthoracic and transesophageal

echocardiographic examinations, which demonstrated a large (24×21 mm), well-defined, round, echodense, homogenous, mobile right atrial mass attached to the SVC via a tiny stalk (Fig. 1 & 2). Additionally, a patent foramen ovale with a left-to-right shunt and a right ventricular systolic pressure of about 30 mm Hg were reported. Based on the echocardiography and multislice spiral computed tomography findings, the patient underwent open-heart surgery and tumor excision. Due to the size

of the mass, we preferred to excise the myxoma during cardiac arrest with the aid of cardiopulmonary bypass. However, cannulating the SVC proved challenging because of the location of the mass. Under the guidance of transesophageal echocardiography, we managed to drain the SVC blood through the cannulation of the right internal jugular vein and then excise the myxoma completely. The pathologic findings of the tumor were compatible with a myxoma.



**Figure 1:** The deep transgastric views by transesophageal echocardiography show the superior vena cava origin of the mass.



**Figure 2:** The 4-chamber view of the right atrium in transesophageal echocardiography shows that the mass originates from the superior vena cava.

## DISCUSSION

Cardiac myxomas are benign tumors with multiple clinical cardiac and extracardiac manifestations.<sup>6</sup> The diagnosis is made by transthoracic echocardiography, although in some cases, transesophageal echocardiography and magnetic resonance imaging can be drawn upon as well.<sup>7,10</sup> Myxomas affect both atria, especially the left atrium. Nevertheless, Keeling et al<sup>10</sup> reported various manifestations in echocardiography findings, including prolapse into the left ventricle, elevated pulmonary artery pressure, reduced left ventricular function, mitral stenosis, left atrial dilation, mitral insufficiency, biventricular dilation, and prolapse into the

right ventricle. The interesting point about our case is that the cardiac myxoma arose from the SVC, which is a very rare location for the origination of this type of tumor according to previous investigations. Indeed, to our knowledge, only 3 cases have been reported to date.

Teixidó et al<sup>9</sup> described a 38-year-old man with a myxoma extending from the SVC to the right pulmonary artery. Sabzi et al<sup>11</sup> reported a rare case of a cardiac myxoma originating in the SVC and presenting as a right atrial mass in a 24-year-old man. The third case was reported by Xiao et al,<sup>12</sup> who described a 59-year-old Chinese woman with a large myxoma arising from the SVC, leading to tricuspid valve obstruction and

right heart failure. In addition, Bortolotti et al<sup>11</sup> reported a rare case of a right atrial myxoma originating from the inferior vena cava.

Cardiac myxomas are classified into 3 types: complex, familial, and sporadic. A characteristic of the familial and complex types is that not only do they usually involve patients at a younger age with atypical locations but also they have a high chance of recurrence.<sup>11</sup>

### CONCLUSIONS

Cardiac myxomas originating from the SVC are very rare and may have a high chance of recurrence. Accordingly, such tumors should be suspected in patients with a complaint of syncope.

### REFERENCES

1. MacGowan SW, Sidhu P, Aherne T, Luke D, Wood AE, Neligan MC, McGovern E. Atrial myxoma: national incidence, diagnosis and surgical management. *Irish journal of medical science*. 1993 Jun 1; 162(6):223-6.
2. Castells E, Ferran V, Octavio de Toledo MC, Calbet JM, Benito M, Fontanillas C, Granados J, Obi CL, Saura E. Cardiac myxomas: surgical treatment, long-term results and recurrence. *The Journal of cardiovascular surgery*. 1993 Feb 1; 34(1):49-53.
3. Nabil Chakfé MD, Kretz JG, Valentin P, Bernard Geny MD, Petit H, Popescu S, Edah-Tally S, Massard G. Clinical presentation and treatment options for mitral valve myxoma. *The Annals of thoracic surgery*. 1997 Sep 1; 64(3):872-7.
4. Bjessmo S, Torbjörn Ivert MD. Cardiac myxoma: 40 years' experience in 63 patients. *The Annals of thoracic surgery*. 1997 Mar 1; 63(3):697-700.
5. Johansson L. Histogenesis of cardiac myxomas. An immunohistochemical study of 19 cases, including one with glandular structures, and review of the literature. *Archives of pathology & laboratory medicine*. 1989 Jul 1; 113(7):735-41.
6. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma: a series of 112 consecutive cases. *Medicine*. 2001 May 1; 80(3):159-72.
7. de Isla LP, de Castro R, Zamorano JL, Almería C, Moreno R, Moreno M, Lima P, Fernández MA. Diagnosis and treatment of cardiac myxomas by transesophageal echocardiography. *American Journal of Cardiology*. 2002 Dec 15; 90(12):1419-21.
8. Keeling I, Oberwalder PJ, Rigler B. Transaortic access for excision of a left ventricular myxoma. *The Annals of thoracic surgery*. 1999 Dec 1; 68(6):2384-5.
9. Sabzi F, Nasiri B. Myxoma of the superior vena cava origin presented as a right atrial mass. *The Journal of Tehran University Heart Center*. 2013 Oct 28; 8(4):202.
10. Keeling IM, Oberwalder P, Anelli-Monti M, Schuchlenz H, Demel U, Tilz GP, Rehak P, Rigler B. Cardiac myxomas: 24 years of experience in 49 patients. *European journal of cardio-thoracic surgery*. 2002 Dec 1; 22(6):971-7.
11. Teixidó G, Galve E, Avegliano G, Majó J. Myxoma extending from the superior vena cava to the right pulmonary artery. *Revista Española de Cardiología (English Edition)*. 2007 Jun 1; 60(6):664-5.
12. Xiao ZH, Hu J, Zhu D, Shi YK, Zhang EY. Tricuspid valve obstruction and right heart failure due to a giant right atrial myxoma arising from the superior vena cava. *Journal of cardiothoracic surgery*. 2013 Dec; 8(1):1-4.
13. Bortolotti U, Faggian G, Mazzucco A, Milano A, Thiene G, Fasoli G, Gallucci V. Right atrial myxoma originating from the inferior vena cava. *The Annals of thoracic surgery*. 1990 Jun 1; 49(6):1000-2.