Accepted Manuscript

Submission Date: 2024-01-23
Accepted Date: 2024-05-01
Accepted Manuscript online: 2024-05-28

The Thoracic and Cardiovascular Surgeon Reports

Transcaval and intracardiac extension of type A tymoma and myxoma: a report of two rare cases

Zulfugar T Taghiyev, Lili-Marie Beier, Vadim Moustafine, Matthias Bechtel, Justus T Strauch, Andreas Böning.

Affiliations below.

DOI: 10.1055/a-2334-7158

Please cite this article as: Taghiyev Z T, Beier L-M, Moustafine V et al. Transcaval and intracardiac extension of type A tymoma and myxoma: a report of two rare cases. The Thoracic and Cardiovascular Surgeon Reports 2024. doi: 10.1055/a-2334-7158

Conflict of Interest: The authors declare that they have no conflict of interest.

Abstract:
We report two cases of rare invasive tumors presenting with transvenous and intracardiac extensions. In one instance, an unusual invasive thymoma type A penetrated into the heart chamber; the other case was an extension of a myxoma into the right atrium that was associated with superior vena cava syndrome. Our interest was stimulated by the rarity of these clinicopathological observations and the unusual clinical features of diagnostic and therapeutic methods presented by these cases.

Corresponding Author:
Dr. Zulfugar T Taghiyev, Berufsgenossenschaftliches Universitatsklinikum Bergmannsheil, Bochum, Germany, taghiyev@mail.de, taghiyev@gmx.de

Contributors' Statement: Z. T. Taghiyev drafting the manuscript
L-M. Beier, V.Moustafine, M. Bechtel, J. Strauch, A. Boening critical revision of the manuscript

Affiliations:
Zulfugar T Taghiyev, Berufsgenossenschaftliches Universitatsklinikum Bergmannsheil, Bochum, Germany
Zulfugar T Taghiyev, UKGM, Department of Adult and Pediatric Cardiovascular Surgery, Giessen, Germany
Lili-Marie Beier, UKGM, Department of Adult and Pediatric Cardiovascular Surgery, Giessen, Germany
[...]
Andreas Böning, UKGM, Department of Adult and Pediatric Cardiovascular Surgery, Giessen, Germany

Transcaval and intracardiac extension of type A tymoma and myxoma: a report of two rare cases

Abstract

We report two cases of rare invasive tumors presenting with transvenous and intracardiac extensions. In one instance, an unusual invasive thymoma type A penetrated into the heart chamber; the other case was an extension of a myxoma into the right atrium that was associated with superior vena cava syndrome. Our interest was stimulated by the rarity of these clinicopathological observations and the unusual clinical features of diagnostic and therapeutic methods presented by these cases.

Keywords

invasive thymoma

myxoma

intracardiac tumor

Introduction

Cardiac tumors represent 0.3% of all human tumors [1]. These tumors are divided etiologically into primary or secondary, the latter also known as metastatic. Secondary or

metastatic cardiac tumors are up to 100 times more frequent than primary tumors. Primary intracardiac tumors are very uncommon, and they may be either benign or malignant. Resections of primary tumors account for 0.3% of all open-heart operations [2]. Around 75% are benign, and about 50% are myxomas, with a prevalence of 0.0017% in the general population [3]. Cardiac tumors may occur topographically in any position of the heart. The growth characteristics comprise two different types, solid or ovoid, whose consistency may be soft or papillary [4].

Accounts of cardiac invasion by malignant thymoma are very rare in the literature [5]. Superior vena cava (SVC) syndrome related to the presence of a thymoma is unusual (4%), and extrinsic expansion with compression of the SVC is most common [6]. A thymoma with this growth pattern is extremely rare, and only a handful of cases have been reported.

Multiple intracavitary masses of the heart are mostly an incidental finding in echocardiography but can also present with arrhythmias and signs of intracardiac obstruction and SVC syndrome [6]. An obstructing growth in the cavity of the heart may also inhibit blood filling with accompanying hemodynamic consequences, leading to symptoms of congestive heart failure, pulmonary hypertension, paroxysmal dyspnea (especially in the horizontal position), arrhythmias, syncope, and sudden death [7].

We report here two cases of cardiac invasion by tumors, one of a ‘benign’ thymoma type A with malignant potential extending into the SVC and RA and one of a myxoma located in the RA that spread to the SVC. Both were successfully treated by surgery and reconstruction.

Case Reports

Case 1:

A 68-year-old Caucasian woman was admitted to the hospital with paralysis of the facial nerve. There were notably no symptoms of SVC syndrome such as edema of the face and extremities. Transesophageal echocardiography (TEE) examination demonstrated a hyperechogenic, mobile mass with soft tissue density in the RA (**Fig. 1a**).

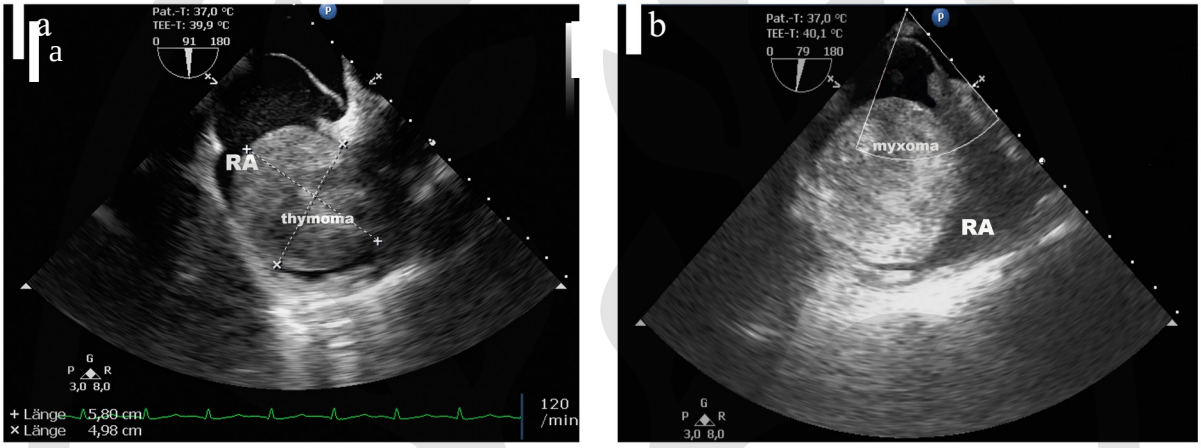

Figure 1 Preoperative TEE showing massive right atrial obstruction and occlusion of the tricuspid valve due to a thymoma (**a, case 1**) and a myxoma (**b, case 2**).

Computed tomography (CT) of the chest exposed a mediastinal tumor extending transvenously into the right brachiocephalic vein and intracardially to the right atrium. A triple-rule-out CT (TRO-CT) scan showed occlusion of the right axillary, brachiocephalic, and subclavian veins and the SVC (**Fig. 2a-c**).

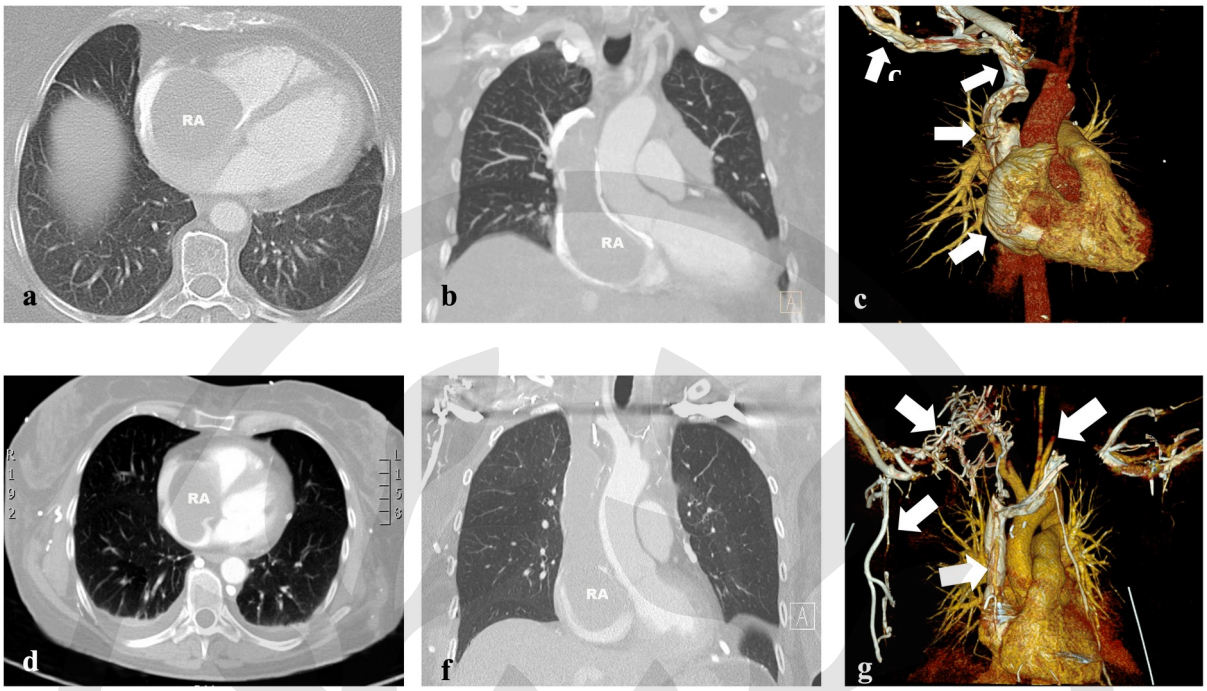

Figure 2. CT scans in horizontal (**a** and **d**) and frontal (**b** and **f**) planes show a massive structure extending transvenously into the SVC and intracardially into the RA. Three-dimensional TRO-CT scans show occlusion of the right axillary, brachiocephalic, and subclavian veins and the superior vena cava and RA through expansion of a thymoma type A **(c, Case 1)** and a myxoma **(g, Case 2)** (arrows). Case 1, a-c; case 2, d-f.

Material extracted by CT-guided core needle biopsy in the anterior mediastinum via a left parasternal approach was highly cellular and consisted of polygonal or rounded epithelial cells with an admixture of lymphocytes. Immunohistochemistry was performed according to standard methods. A histopathological diagnosis of thymic tumor, favoring type A, was made. Pathological examination verified a thymoma type A (**Fig. 3a-b**), similar to the diagnosis based on the biopsy material. The mass in the SVC and RA showed the same histology as the primary tumor, and immunohistochemistry showed the same results as the primary mediastinal tumor.

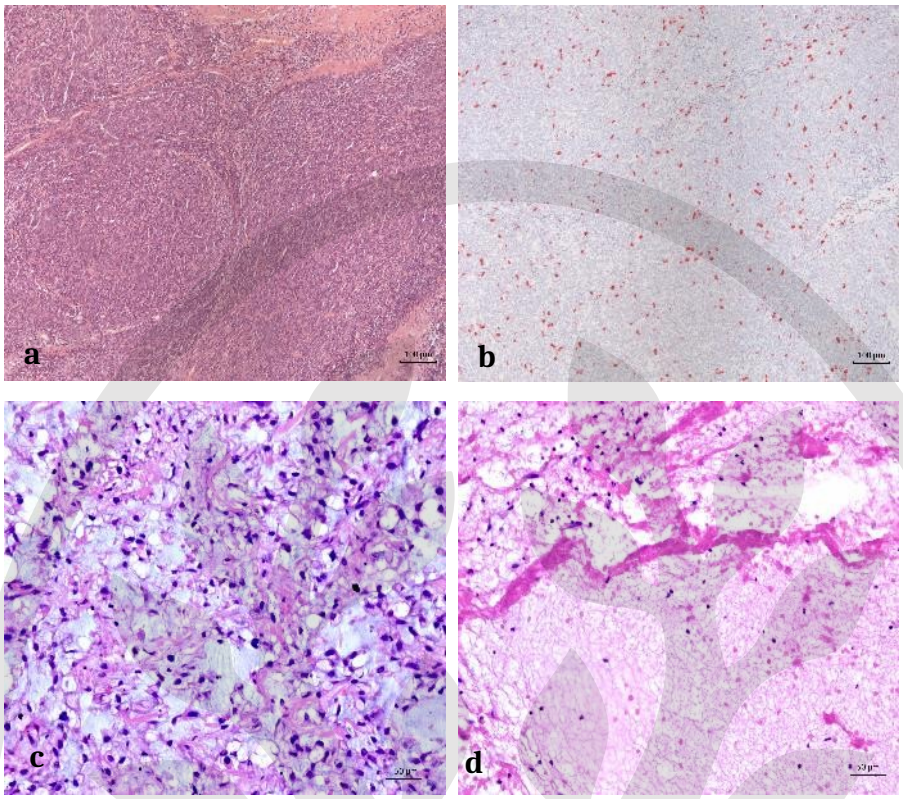

Figure 3. a) Hematoxylin and eosin (H&E) stain of thymoma type A; **b**) Ki67 TR-FRET positive cores (mAb Mib1); **c**) H&E stain of the myxoma with hyperchromatic multinucleated cells in the profuse myxoid matrix and **d**) of the retrieved thrombus. Scale bars 100 µm (a, b) and 50 µm (c, d). Case 1, a & b; case 2, c & d.

Case 2:

A 47-year-old also Caucasian woman was admitted to our hospital after presenting with edema of the face and upper extremities six days previously. A palpable mass was present in her neck, right shoulder, and axillary areas. As in the first case, a TEE examination demonstrated a hyperechogenic, mobile mass in the RA (**Fig. 1b**). Despite these findings, we could not differentiate between diagnosis of myxoma, thymoma, or giant thrombotic mass.

CT and three-dimensional images from TRO-CT were consistent with SVC syndrome and

involvement of both brachiocephalic veins with dilated paravertebral veins and posterior external venous plexus (**Fig. 2d-g**).

Right atrial mass pathology with hematoxylin and eosin staining of the 9-cm mass showed spindle, stellate, and oval-shaped tumor cells on a myxoid matrix that was indicative of a myxoma. Interestingly, the mass found in the SVC and the azygos vein was a thrombus (**Fig. 3c, d**).

Surgical treatment

Taking into account the relatively young age of both patients and the wide extension of the tumor, the decision was made to treat surgically via median sternotomy. After cannulation of either the inferior vena cava (IVC) (case 1) or the right femoral vein (case 2), and the ascending aorta, cardiopulmonary bypass in moderate hypothermic circulatory (28°C) arrest was accomplished. In case 1, we made three incisions on the brachiocephalic vein, SVC, and RA to resect the tumor mass. An 8x5-cm anterior mediastinal thymoma extended through the brachiocephalic veins (5 cm) and SVC (6 cm) into the RA, constituting a globular form, 9x4.5x3 cm^3 in volume (**Fig. 4a**), without the need for conduit anastomoses. A similar surgical approach was taken in case 2; interestingly, the tumor volume was identical to that of case 1. The myxoma was located at the top of the RA and adhered to the lateral atrial wall and SVC (**Fig. 4b**). After resection in toto, the atrium and SVC were reconstructed with patch plastic using the bovine pericardium and sutured continuously via 5-0 prolene. Operation time and cardiopulmonary bypass time were similar in the two cases: 200 min and 100 min in the first case and 240 min and 111 min in the second case, respectively. Both of the patients survived the procedure without any complications.

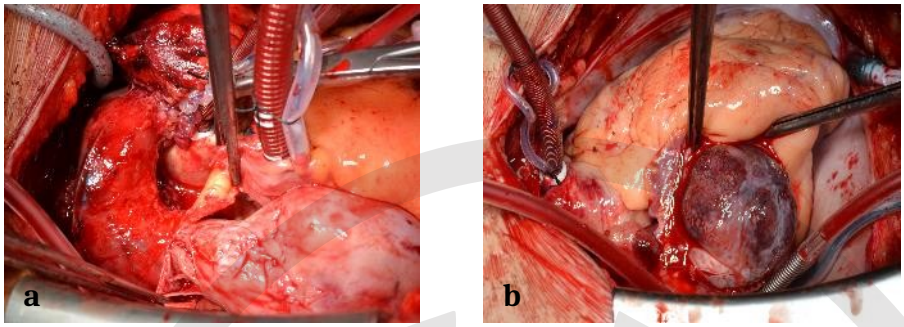

Figure 4. Intraoperative presentation of thymoma (**a**) and myxoma (**b**).

Discussion

Intracardiac tumors presenting as giant masses are uncommon. There are several reports of cardiac invasion by malignant thymoma type A in the literature [1,5,8]. Thymomas that grow from the thymus gland are unusual. The WHO has established a scheme for thymoma classification that distinguishes three types: A (medullary, spindle cell), AB (mixed), and B (cortical), with subtypes of B1, B2, and B3, where an atypical thymoma type A was specified to the WHO thymoma classification [9]. Type A thymoma may exhibit behavior closer to benign tumors compared to other thymoma types with an excellent prognosis; it is associated with a 100% survival rate at 5 and 10 years [9]. It is also recognized that all thymomas have malignant potential, so these tumors should not be regarded as strictly benign. These morphologic observations are complimented by new genetic data that provide support for the existing classification and potential for deeper insights into the molecular characteristics of these tumors [10].

Intracaval and transvenous cardiac metastasis of thymoma type A with extension into the RA is extremely rare. However, an aggressive potential of benign types A and AB with the ability to penetration into adjacent organs has also been reported [11-13]. In advanced stages, it is

generally associated with SVC syndrome. Multiple tumor emboli of the pulmonary arteries or tumor strangulation at the tricuspid orifice has been identified as the reason for death. In case 1, there was neither a sign of the SVC syndrome nor the formation of venous collateral pathways, and the thymoma was an incidental finding.

The most prevalent primary intracardiac mass in adults is an atrial myxoma. Intracardiac myxomas account for less than 5% of all cases [14]. Most myxomas occur in the left atrium, while some are found in ventricles, SVC, or pulmonary veins [15]. The remaining 15-20% of myxomas emerge from the RA [16]. The morphology of cardiac myxoma is commonly polypoid with a peduncle and typically circular or oval [17]. The myxomas usually commence in the fossa ovalis or at the base of the interatrial septum [18].

Our case was a rare example of myxoma that was located in the RA and spread to the SVC, with collateral venous pathways through the right lateral thoracic and left internal thoracic routes. This was accompanied by abnormalities of the vertebral venous route, with dilated paravertebral veins and posterior external venous plexus due to thrombosis of the superficial and deep veins.

A TRO-CT scan showed acquired abnormalities of collateralization of the veins of the upper extremity and thoracic inlet due to thrombosis of the superficial and deep veins. These included the right jugular, brachiocephalic, subclavian, and axillary veins. The dilated internal thoracic vein is fed by enlarged intercostal veins and drains via diaphragmatic veins. Meier et al. (2013) observed the azygos route in 39 of 56 patients (70%), and it was the only collateral pathway in one (2%). These findings confirm that the azygos venous system connects the IVC with the SVC and provides an essential cavocaval anastomosis. Pathophysiologically, an obstruction after the influx of the azygos vein into the SVC results in a stream inversion, and blood may flow in a retrograde manner via this pathway and return to the RA via the IVC [19,

20].

Consent

Written informed consent was obtained from the patients for the publication of these case reports and any accompanying images. Copy of the written consent forms are available for review.

Abbreviations

CT: computed tomography

H&E: Hematoxylin and eosin

IVC: inferior vena cava

mAb: Monoclonal antibody

RA: right atrium

SVC: superior vena cava

TEE: transesophageal echocardiography

TRO-CT: triple-rule-out computed tomography

TR-FRET: Time-resolved fluorescence resonance energy transfer

WHO: World Health Organisation

References

[1] Song Y., Hu R., Yao Q. (2003). Pathological analysis of 268 cases of tumors in the heart and the pericardium. *J SUN Yat-sen-Univ (Med Sci)*., 24:197– 201

[2] Roberts, W. C. (1997). Primary and secondary neoplasms of the heart. *American Journal of Cardiology*, *80*(5), 671-682.

[3] Nina, V. J., Silva, N. A., Gaspar, S. F., Rapôso, T. L., Ferreira, E. C., Nina, R. V., ... & Filho, N. S. (2012). Atypical size and location of a right atrial myxoma: a case report. *Journal of medical case reports*. *6*, 1-5.

[4] Yu, K., Liu, Y., Wang, H., Hu, S., & Long, C. (2007). Epidemiological and pathological characteristics of cardiac tumors: a clinical study of 242 cases. *Interactive cardiovascular and thoracic surgery*, *6*(5), 636-639.

[5] Gleeson RE, Price R, Nicholson AG, Pastorino U. Invasive thymoma extending to the right atrium: excision without cardiopulmonary bypass. *J Cardiovasc Surg (Torino)*. 1997 Aug;38(4):429-31

[6] Chen JC, Bongard F, Klein SR. A contemporary perspective on superior vena cava syndrome. *Am J Surg.* 1990 Aug;160(2):207-11.

[7] Pinede, L., Duhaut, P., & Loire, R. (2001). Clinical presentation of left atrial cardiac myxoma: a series of 112 consecutive cases. *Medicine*, *80*(3), 159-172.

[8] Peachell, J. L., Mullen, J. C., Bentley, M. J., & Taylor, D. A. (1998). Biatrial myxoma: a rare cardiac tumor. *The Annals of thoracic surgery*, *65*(6), 1768-1769.

[9] Travis, W. D., Brambilla, E., Burke, A. P., Marx, A., & Nicholson, A. G. (2015). Introduction to the 2015 World Health Organization classification of tumors of the lung, pleura, thymus, and heart. *Journal of Thoracic Oncology*, *10*(9), 1240-1242.

[10] Marx, A., Chan, J. K., Coindre, J. M., Detterbeck, F., Girard, N., Harris, N. L., Ströbel, P. (2015). The 2015 World Health Organization classification of tumors of the thymus: continuity and changes. *Journal of Thoracic Oncology*, *10*(10), 1383-1395.

[11] Jain, R. K., Mehta, R. J., Henley, J. D., Kesler, K. A., Loehrer, P. J., & Badve, S. (2010). WHO types A and AB thymomas: not always benign. *Modern Pathology*, *23*(12), 1641-1649.

[12] Vladislav, T., Jain, R. K., Alvarez, R., Mehta, R. J., Gökmen-Polar, Y., Kesler, K. A., ... & Badve, S. (2012). Extrathoracic metastases of thymic origin: a review of 35 cases. *Modern Pathology*, *25*(3), 370-377.

[13] Moran, C. A., Kalhor, N., & Suster, S. (2010). Invasive spindle cell thymomas (WHO type a) a Clinicopathologic correlation of 41 cases. *American journal of clinical pathology*, *134*(5), 793-798.

[14] Peachell, J. L., Mullen, J. C., Bentley, M. J., & Taylor, D. A. (1998). Biatrial myxoma: a rare cardiac tumor. *The Annals of thoracic surgery*, *65*(6), 1768-1769.

[15] Lamparter, S., Moosdorf, R., & Maisch, B. (2004). Giant left atrial mass in an asymptomatic patient. *Heart*, *90*(5), e24-e24.

[16] Diaz, A., Di Salvo, C., Lawrence, D., & Hayward, M. (2011). Left atrial and right ventricular myxoma: an uncommon presentation of a rare tumour. *Interactive cardiovascular and thoracic surgery*, *12*(4), 622-625.

[17] Reynen, K., Köckeritz, U., & Strasser, R. H. (2004). Metastases to the heart. *Annals of oncology*, *15*(3), 375-381.

[18] Stolf, N. A., Benício, A., Moreira, L. F. P., & Rossi, E. (2000). Mixoma de átrio direito com origem na veia cava inferior: uma localização rara com implicações diagnósticas e terapêuticas. *Brazilian Journal of Cardiovascular Surgery*, *15*, 255-258.

[19] Piciucchi, S., Barone, D., Sanna, S., Dubini, A., Goodman, L. R., Oboldi, D., Poletti, V. (2014). The azygos vein pathway: an overview from anatomical variations to pathological changes. *Insights into imaging*, *5*, 619-628.

[20] Kapur, S., Paik, E., Rezaei, A., & Vu, D. N. (2010). Where there is blood, there is a way: unusual collateral vessels in superior and inferior vena cava obstruction. *Radiographics*, *30*(1), 67-78.