




Transcaval and Intracardiac Extension of Type A Thymoma and Myxoma: A Report of Two Rare Cases

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Thorac Cardiovasc Surg Rep 2024;13:e25–e28.

Abstract

Keywords

- invasive thymoma
- myxoma
- intracardiac tumor

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.

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 approximately 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 the 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 incidental findings 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 right atrium (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

received

January 23, 2024

accepted after revision

May 1, 2024

accepted manuscript online

May 28, 2024

DOI <https://doi.org/10.1055/a-2334-7158>

10.1055/a-2334-7158.

ISSN 2194-7635.

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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).

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).

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.

Case 2

A 47-year-old also Caucasian woman was admitted to our hospital after presenting with edema of the face and upper extremities 6 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 were 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 8 × 5-cm anterior mediastinal thymoma extended through the brachiocephalic veins (5 cm) and SVC (6 cm) into the RA, constituting a globular form, $9 \times 4.5 \times 3 \text{ cm}^3$ 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 minutes and 100 minutes in the first case and 240 minutes and 111 minutes in the second case, respectively. Both of the patients survived the procedure without any complications.

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 World Health Organization (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

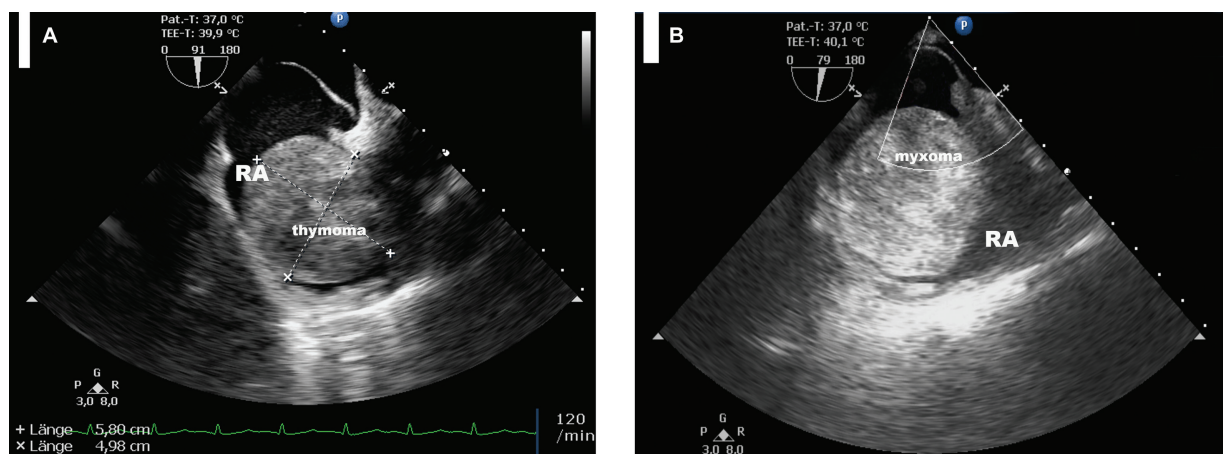


Fig. 1 Preoperative transesophageal echocardiography showing massive right atrial obstruction and occlusion of the tricuspid valve due to a thymoma (A, case 1) and a myxoma (B, case 2).

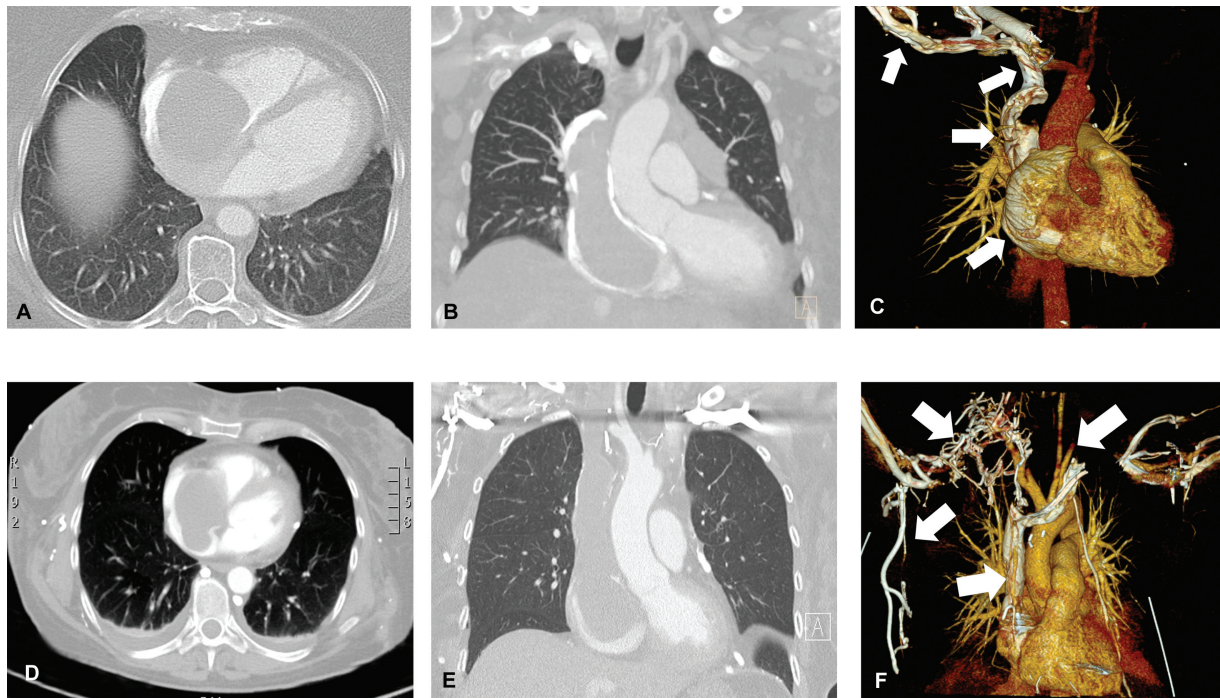


Fig. 2 Computed tomography (CT) scans in horizontal (A and D) and frontal (B and F) planes show a massive structure extending transvenously into the superior vena cava and intracardially into the right atrium (RA). Three-dimensional triple-rule-out-CT scans show the occlusion of the right axillary, brachiocephalic, 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.

specified to the WHO thymoma classification.⁹ Type A thymoma may exhibit behavior closer to benign tumors compared with 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 com-

plemented 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 penetrate 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.⁸ Most myxomas occur in the left atrium, while some are found in ventricles, SVC, or pulmonary veins.¹⁴ The remaining 15 to 20% of myxomas emerge from the RA.¹⁵ The morphology of cardiac myxoma is commonly polypoid with a peduncle and typically circular or oval.¹⁶ The myxomas usually commence in the fossa ovalis or at the base of the interatrial septum.¹⁷

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

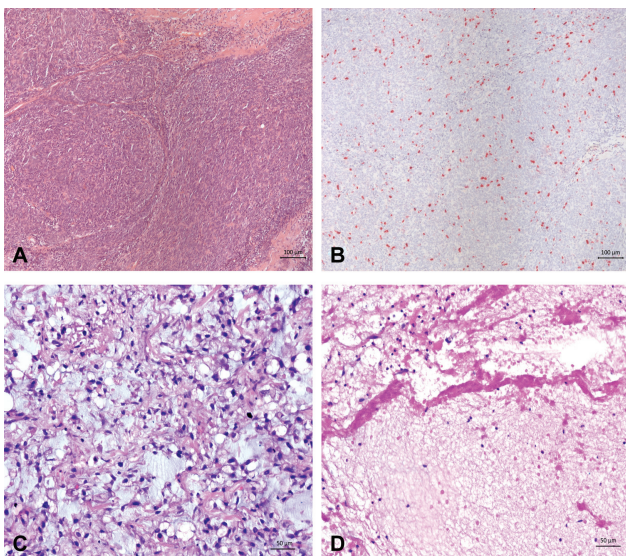


Fig. 3 (A) Hematoxylin and eosin (H&E) stain of thymoma type A; (B) Ki67 time-resolved fluorescence resonance energy transfer positive cores (mAb Mib1); (C) H&E stain of the myxoma with hyperchromatic multinucleated cells in the profuse myxoid matrix; and (D) of the retrieved thrombus. Scale bars 100 µm (A and B) and 50 µm (C and D). Case 1, A and B; case 2, C and D.

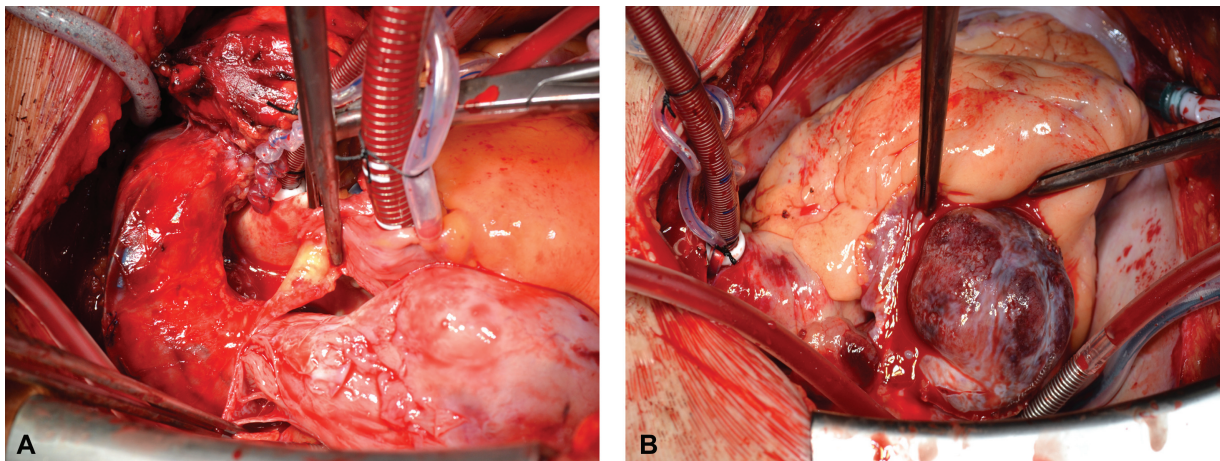


Fig. 4 Intraoperative presentation of thymoma (A) and myxoma (B).

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.^{18,19}

Patients' Consent

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

Conflict of Interest

None declared.

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