Case Report and Review of Literature
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B2 Thymoma with Intracardiac Extension Presenting as Superior Vena Cava Syndrome: Case Report and Literature Review

Almedina Muhić[1], Šefika Umihanić[1], Hasan Osmić[1], Elma Mujaković[2], Faruk Šadić[3], Amila Kovčić Harčinović[1], Agan Muhić[4]

[1]Department of Oncology and Radiotherapy, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina, [2]Faculty of Medicine, University of Tuzla, Tuzla, Bosnia and Herzegovina, [3]Department of Radiology, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina, [4]Department of Orthopedics and Traumatology, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina

Correspondence: *almedina_terzic@hotmail.com*; Tel: + 387 35 303389

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Abstract

Objective. This article aims to emphasize the importance of considering invasive thymoma in the differential diagnosis of mediastinal masses and highlights the critical role of timely surgical and oncological management in improving patient outcomes. **Case Report.** We present the case of a 70-year-old woman who presented with signs of superior vena cava syndrome, including dyspnea, facial swelling, and fatigue. Advanced imaging and intraoperative findings revealed a large anterior mediastinal mass infiltrating the left brachiocephalic vein and superior vena cava, extending into both the right atrium and right ventricle. Surgical intervention was performed, and histopathological analysis confirmed B2 thymoma with a high Ki-67 proliferation index. Despite surgical intervention, the patient's condition deteriorated, and she ultimately succumbed to the disease. **Conclusion.** To the best of our knowledge, this is the first reported Bosnian case of B2 thymoma invading the brachiocephalic vein and superior vena cava and infiltrating both the right atrium and ventricle, causing superior vena cava syndrome. Despite their rarity, thymomas should always be considered in patients presenting with an enlarged mediastinum.

Key Words: B2 Thymoma ▪ Mediastinal Mass ▪ Superior Vena Cava Syndrome ▪ Echocardiography.

Introduction

Invasive thymoma extending into the brachiocephalic vein, superior vena cava (SVC), right atrium (RA), and right ventricle (RV) is a very rare but clinically significant condition. The incidence of thymoma is relatively low, with approximately 0.17 cases per 100,000 individuals globally (1). Invasive thymomas account for approximately 30% of all thymomas and can invade mediastinal organs, such as the pleura and pericardium (2). However, only a small number of thymomas are associated with SVC syndrome, often due to extrinsic compression rather than intravascular invasion. Intravascular invasion of the SVC extending into the RA and RV is an even rarer phenomenon, first reported by Suzuki et al. in 1976, with few cases documented since then (3-5). The main objective of this case report is to describe a rare manifestation of type B2 thymoma with simultaneous SVC and intracardiac invasion and to place its presentation within the current clinical and pathological context while highlighting its diagnostic and prognostic implications.

Case Reports

Clinical Presentation

A 70-year-old woman was admitted with a several-month history of difficulty in breathing, swelling of the face and neck, and excessive fatigue. She reported that her symptoms had gradually worsened, interfering with her daily activities, and had

become more acute in the past few days. Physical examination revealed facial plethora, distension of the jugular veins, and bilateral lower limb edema, consistent with impaired venous return. Auscultation revealed diminished breath sounds over the left hemithorax.

Imaging Findings and Surgical Approach

An initial chest X-ray, obtained during pulmonary evaluation, showed widening of the superior mediastinum, an enlarged cardiac silhouette, and a left-sided pleural effusion. As part of further cardiac work-up, transthoracic echocardiography was performed and revealed a large (8.5 cm × 4 cm) echogenic mass within the right atrium, prolapsing into the right ventricle during diastole, which was initially interpreted as a possible atrial myxoma. Owing to the urgent clinical presentation and high suspicion of an intracardiac tumor, the patient was referred directly for cardiovascular surgery without preoperative cross-sectional imaging. The patient underwent median sternotomy on December 14, 2023, with extirpation of the tumor formation involving the right atrium, right ventricle, and superior vena cava. Intraoperatively, a solid tumor formation was identified in the anterior mediastinum at the thymic site, infiltrating the wall of the left brachiocephalic vein and extending into the superior vena cava, right atrium, and right ventricle. Complete excision was not feasible because of vascular invasion, and the tumor was removed in fragments. The postoperative course was uneventful and without complications. Following surgery, contrast-enhanced computed tomography (CT) of the chest was performed for staging and postoperative assessment. It revealed a large residual anterior mediastinal mass, measuring approximately 10.3×5.2×8 cm with irregular margins and central necrosis (Figure 1).

The mass continued to infiltrate the wall of the left brachiocephalic vein and superior vena cava (SVC). In addition, multiple enlarged mediastinal lymph nodes were present, particularly in the pretracheal and aortopulmonary window regions, along with pathologic lymphadenopathy in the left axilla and upper abdomen (Figure 2). Free peritoneal fluid was noted around the liver, spleen, and within the pelvis, consistent with advanced systemic disease.

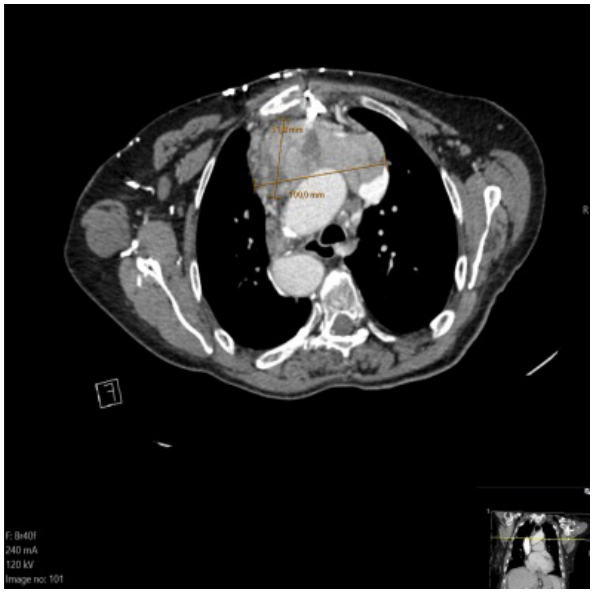

Figure 1. Contrast-enhanced CT of the chest showing a large anterior mediastinal mass infiltrating the left brachiocephalic vein.

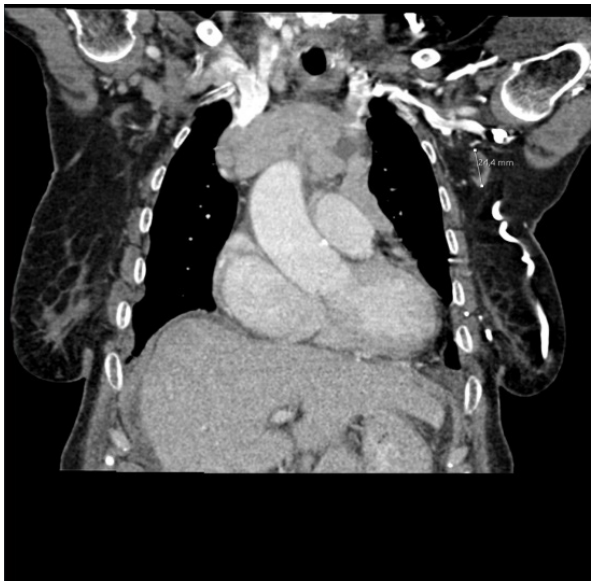

Figure 2. Contrast-enhanced CT demonstrating pathological lymphadenopathy: enlarged lymph node in the left axilla.

Pathohistological Findings

Pathohistological analysis of the resected tumor fragments revealed tumor tissue with a focally present connective capsule on the surface. The tumor is composed of smaller and larger nodules, as well as areas formed by tumor cells. Approximately 75% of these cells are characteristic of immature thymocytes, immunohistochemically positive for CD3, CD5, bcl2, TdT, CD1a (weakly positive), and CD99. Interspersed among these are another population of cells with epithelioid characteristics, displaying oval to polygonal, occasionally slightly elongated nuclei with finely dispersed chromatin and focally visible nucleoli. These cells are immunohistochemically positive for AE1/AE3 and p63. Acellular loose vascularized connective tissue can be seen between the tumor cells. Immunohistochemically, the tumor cells are negative for CD20, bcl6, synaptophysin, chromogranin, CD56, TTF1, and CD10. The Ki67 proliferative activity index is almost 100%. The morphological and immunohistochemical characteristics of the analyzed sections support a diagnosis of thymoma, B2 type. Because the specimen was received in fragments, the surgical margin status and pT stage could not be determined.

Outcome

Following surgery and further testing, the patient initially recovered without perioperative complications and was discharged in stable condition. She was subsequently evaluated by a multidisciplinary oncology board, which included specialists in medical oncology, radiation oncology, surgery, clinical pharmacology, and pathology. Given the extent of the disease and systemic involvement, she was deemed unsuitable for curative oncologic treatment and was referred for palliative management. Over the following weeks, her condition progressively deteriorated with worsening fatigue and edema. After several refusals, the patient was finally admitted to the Center for Palliative Care. Despite supportive therapy, her condition progressively deteriorated, and she passed away approximately three months after diagnosis.

Discussion

Thymomas are the most common primary tumors of the anterior mediastinum, although their overall incidence remains very low, estimated at 0.17 cases per 100,000 population annually (1). They are frequently detected incidentally or present with nonspecific thoracic symptoms, such as cough, chest pain, or dyspnea, caused by local compressive effects (2, 6, 7). In addition to local compressive symptoms, thymomas are notable for their frequent association with paraneoplastic syndromes, most prominently myasthenia gravis, which occurs in 30–50% of patients. Other autoimmune conditions, including pure red cell aplasia and hypogammaglobulinemia, may also be seen, although they are less common (8). Therefore, patients may present with a spectrum ranging from asymptomatic incidentalomas to chest symptoms and systemic autoimmune manifestations (9).

Radiologically, thymomas typically appear as well-circumscribed lobulated soft-tissue masses in the anterior mediastinum on chest CT or MRI (10). On CT, smaller tumors usually demonstrate homogeneous soft-tissue density, whereas larger lesions may contain areas of necrosis, cystic change, or calcification (11). MRI can further provide tumor composition, with thymomas often demonstrating signal intensity similar to or higher than muscle on T1-weighted sequences and higher than muscle on T2-weighted sequences, sometimes approaching that of fat, which may hinder distinction from surrounding mediastinal fat (12). Features suggesting invasive disease include irregular or ill-defined borders, loss of normal fat planes between the tumor and adjacent structures, vascular encasement, and pleural implants (13).

According to the 2021 WHO classification, thymomas are divided into types A, AB, B1, B2, and B3 based on the morphology of epithelial cells and the proportion of lymphocytes (14). Among the histological subtypes, type B2 thymoma accounts for approximately 20% of all thymomas. It is composed of scattered epithelial cells with vesicular nuclei and prominent nucleoli within a dense lymphocyte population (15). Type B2 thymomas

exhibit moderate aggressiveness, with reported five-year survival rates between 60% and 70% (12). Their prognosis is worse than that of types A, AB, and B1, but more favorable than that of type B3 or thymic carcinoma (16).

Invasive thymomas typically extend to adjacent mediastinal structures, such as the pleura, pericardium, or lungs (17). True intravascular or intracardiac growth is extremely rare. Since Suzuki et al. first described a thymoma with intravascular extension into the superior vena cava (SVC) and right atrium in 1976 (3), only a small number of such cases have been reported. In most instances, cardiac involvement is limited to pericardial invasion rather than direct intracaval or intracardiac extension (18). The presumed mechanism of intracaval spread involves the invasion of small thymic veins with subsequent downstream expansion into larger vessels and cardiac chambers (19).

Our case illustrates several unusual and clinically significant features. The patient presented with the classical signs of SVC syndrome, including dyspnea, facial swelling, and venous distension, resulting from tumor invasion of the left brachiocephalic vein and SVC. The tumor further extended into the right atrium and right ventricle, prolapsing through the tricuspid valve during the cardiac cycle, an exceptionally rare manifestation. Pathohistological analysis confirmed a type B2 thymoma with an almost 100% Ki-67 proliferation index, reflecting highly aggressive tumor behavior. Finally, the initial echocardiographic interpretation suggested a right atrial myxoma, highlighting the diagnostic challenges when invasive mediastinal tumors mimic primary intracardiac masses.

The therapeutic implications are equally important. Complete surgical resection remains the key treatment for thymoma and is associated with better long-term survival outcomes (20). In our case, however, en bloc resection was not feasible due to extensive vascular and intracardiac invasion, and the tumor was removed in fragments. Postoperative multidisciplinary evaluation concluded that the patient was not a candidate for further oncologic therapy, highlighting the limited treatment options available in such advanced stages. Reported outcomes of similar cases confirm the poor prognosis of thymomas with intracaval or intracardiac spread, despite surgical intervention (21).

Based on six well-documented cases with dual-chamber (RA + RV) invasion identified in the literature, together with the present case, a consistent clinicopathological pattern emerged, as shown in Table 1. All patients presented with manifestations of superior vena cava obstruction, such as dyspnea, facial and upper limb swelling, and distended neck veins (Table 1). Imaging confirmed a continuous tumor extension from the mediastinum into the right atrium and, in some cases, across the tricuspid valve into the right ventricle, indicating direct venous propagation rather than hematogenous spread. From a treatment perspective, complete surgical resection with cardiopulmonary bypass was associated with better outcomes compared to chemotherapy or radiotherapy alone (Table 1). The almost universal involvement of the right atrium and the rarity of right ventricular extension likely reflect the venous anatomy of the thymus. Thymic veins predominantly drain into the brachiocephalic veins and superior vena cava, providing a direct pathway for tumor spread into the right atrium (22). In contrast, right ventricular involvement requires additional progression across the tricuspid valve, which is quite unusual and an anatomically unfavorable pathway for tumor expansion. The histological spectrum of these cases ranged from type A to type B2 thymomas, including spindle-cell and mixed epithelial forms, without a clear correlation between subtype and invasive potential (Table 1). The present case of a B2 thymoma with a nearly 100% Ki-67 proliferation index illustrates how high proliferative activity may promote aggressive intraluminal growth beyond the right atrium.

Surgical resection under cardiopulmonary bypass (CPB) was performed in four of the seven patients, allowing partial or complete tumor removal. Three of these patients were alive at follow-up, whereas non-surgical management resulted in poor outcomes (Table 1). These findings emphasize that CPB-assisted resection with possible

Table 1. Reported Cases of Thymoma With Dual-Chamber (RA + RV) Intracardiac Invasion

Year	First author	Histology (WHO)	Intracardiac involvement	Extra-cardiac spread	Clinical presentation	Treatment	Outcome at reporting
1990	Airan (4)	Malignant thymoma (spindle-cell/ epithelial type)	RA + RV	SVC, right lung, aortic arch, tracheal bifurcation (compression)	SVC syndrome (neck and chest vein distension, hepatomegaly, ascites, weight loss)	Partial resection (CPB, RA + RV) + postoperative RT	Alive
2000	Hayashi (23)	Mixed (predominantly epithelial) thymoma	RA + RV	LBCV, SVC	SVC syndrome, dyspnea	Two resections under CPB + mediastinal RT	Alive
2003	Funakoshi (24)	Type A thymoma; Masaoka stage IVb	RA+RV	SVC, Bilateral BCV	Facial and left upper-limb swelling (SVC syndrome)	Urgent radical resection with SVC reconstruction + postoperative RT	Alive
2016	Chadha (25)	Invasive thymoma (histological type not specified)	RA + RV	SVC, Anterior mediastinum	Dyspnea, chest pain	Patient declined surgical treatment	Alive
2016	Senanayake (26)	Thymoma (histological type not specified)	RA + RV	SVC, Bilateral BCV	Exertional dyspnea, peripheral edema, bilateral pleural effusions (SVC syndrome)	Chemotherapy (cisplatin-based), palliative RT planned	Deceased before RT, 6 months after presentation
2022	Asami-Noyama (27)	Type A thymoma	RA + RV	Bilateral BCV, SVC, lung, lymph nodes	Facial and upper-limb edema, dyspnea, SVC syndrome	Carboplatin + paclitaxel chemotherapy; sudden obstructive shock; anticoagulation attempted	Deceased 5 days after start of chemo (massive intracardiac thrombosis)
2025	Present case	B2 thymoma	RA + RV	LBCV, SVC	SVC syndrome, facial swelling, dyspnea	Partial resection (fragmented)	Deceased

RA=Right atrium; RV=Right ventricle; SVC=Superior vena cava; CPB=Cardiopulmonary bypass; LBCV=Left brachiocephalic vein; RT=Radiotherapy; BCV=Brachiocephalic veins.

venous reconstruction provides the best chance of survival when anatomically feasible. Our patient exhibited the most extensive pattern of invasion, involving the left brachiocephalic vein, SVC, right atrium, and ventricle. Despite partial resection, the outcome was unfavorable, consistent with other reports of incomplete excision. Collectively, this focused analysis of seven RA + RV cases provides a clearer understanding of this rare disease behavior, underscoring the need for early imaging-based diagnosis and multidisciplinary planning. While histology alone does not predict invasion, the extent of venous involvement and the possibility of complete resection remain the main determinants of prognosis.

In summary, this case highlights the importance of including thymoma in the differential diagnosis of mediastinal masses presenting with SVC syndrome. It emphasizes the limitations of echocardiography in differentiating invasive mediastinal tumors from intracardiac lesions, and the need for comprehensive imaging and pathohistological confirmation. To our knowledge, this is the first Bosnian case of type B2 thymoma with simultaneous invasion of the brachiocephalic vein, SVC, right atrium, and right ventricle. Recognition of this rare growth pattern is critical for timely diagnosis and multidisciplinary management, although therapeutic options and outcomes remain limited in advanced disease.

Conclusion

Intracardiac extension of thymoma is rare but carries significant diagnostic and therapeutic challenges. This case adds to the limited global literature and represents, to the best of our knowledge, the first regional report of a B2 thymoma invading both the right atrium and ventricle. Recognition of this unusual growth pattern is important for timely diagnosis, surgical planning, and coordinated, multidisciplinary care.

What Is Already Known on This Topic:
Thymomas are the most prevalent primary mediastinal malignancy, but they are rare and often exhibit local invasion into surrounding structures. "Transvenous" cardiac metastases, or intracaval growth extending into the right atrium, rarely occur, and only a few examples of transcaval extension with intracardiac involvement have been documented.

What This Study Adds:
In contrast to previous studies, where the majority of thymomas with cardiac involvement were restricted to the right atrium, pericardium, or SVC, we present a case of thymoma metastasis to both right heart chambers with atypical clinical manifestations.

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

Informed Consent: Approval was obtained from the Ethical Committee of the University Clinical Center Tuzla for this case report. Date of approval: February 26, 2025. Approval number: 02-09/2-206-2/24.

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