Case Report

A Rare Case of Malignant Thymoma with Superior Vena Cava Syndrome in a Young Man

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Abstract

AUJ was a 29-year-old male student who presented with a 4-month history of cough, vomiting, headache, distended neck veins, dysphagia to solid meals, and recurrent episodes of dyspnea. Physical examination revealed right supraclavicular soft-to-firm nontender mass with no palpable thrill. The superficial veins of the face, neck, and chest were dilated. Chest computed tomography (CT) scanogram showed widened mediastinum with a lobulated soft-tissue mass seen at that region, compressing the trachea, bronchi, and superior vena cava. The anterior mediastinal mass was lobulated and showed heterogeneous density with minimal contrast enhancement. It was complex mixed echogenic on ultrasound with no remarkable color signal changes on color Doppler interrogation. A diagnosis of anterior mediastinal mass, most likely thymoma with superior vena cava syndrome, was made. CT-guided biopsy confirmed the diagnosis of malignant thymoma. He was slated for debulking surgery, and this was to be followed by radiotherapy and/or chemotherapy. At the time of intubation during anesthetic procedure, however, he developed cardiac arrest and died on the 15th day of intensive care management.

Keywords: Computed tomography-guided biopsy, superior vena cava syndrome, thymoma

Introduction

Thymoma is a tumor originating from the epithelial cells of the thymus, a lymphoid organ located in the anterior mediastinum. It is the most common neoplasm of the anterior mediastinum and best known for its association with the neuromuscular disorder, myasthenia gravis, and, very rarely, a life-threatening superior vena cava syndrome (SVCS).[1,2]

The cause of thymoma is unknown, and the risk factors have not been identified.[3] Men and women are equally affected, most often in the fifth and sixth decades of life.[3] No clear histologic distinction between benign and malignant thymomas exists, hence the need for imaging. The propensity of a thymoma to be malignant is determined by its invasiveness.[1,3] The goal of imaging is to identify the tumor and stage it appropriately, with emphasis on local invasion and distant spread. The rarity of malignant thymoma in the third decade of life coupled with its rare association with SVCS prompted this case report.

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Case Report

AUJ was a 29-year-old male student from Southeastern Nigeria who presented to the Surgical Outpatient Department of Ahmadu Bello University Teaching Hospital with a 4-month history of cough, vomiting, headache, and recurrent episodes of dyspnea. Cough was of sudden onset with minimal whitish sputum production initially and later with streaks of blood. There was dysphagia to solid meals, but no odynophagia or hoarseness of voice was experienced. He neither smoked cigarette nor drank alcohol, and there was no family history of similar illness.

Physical examination revealed calm and well-built young man who was not in respiratory distress. Facial and neck veins were visibly engorged and dilated. The neck examination revealed right supraclavicular fullness with a palpable soft-to-firm

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mass devoid of thrill or bruit noted at the region. The right external jugular vein and facial and anterior chest wall veins were distended. A clinical assessment of mediastinal mass with SVCS to rule out thymoma, lymphoma, and ectopic thyroid was made.

Chest computed tomography (CT) scanogram [Figure 1] revealed widened mediastinum due to a lobulated opacity of soft-tissue density seen compressing the trachea and the bronchi. The demonstrated lung fields were clear. Chest ultrasound scans through a suprasternal approach revealed a fairly rounded mixed echogenic solid mass in the superior mediastinum, measuring 10.3 cm × 7.2 cm in dimensions [Figure 2]. The complementary neck ultrasound scan revealed normal lobes of the thyroid and the isthmus. The right internal jugular vein was significantly dilated [Figure 3], but the left internal jugular vein and both common carotid arteries were preserved. On chest CT scan, the mass was located in the anterior mediastinal region and appeared as heterogeneously dense, lobulated, minimally enhancing, and nearly occluding the trachea. It measured 11.9 cm × 7.1 cm (axial dimension) and 15.0 cm × 10.5 cm (sagittal reconstruction) with Hounsfield Unit (HU) value of 56 [Figures 4a-b and 5]. There were stippled hyperdense lesions of calcific density (HU = 268) seen within the mass. The hematological and biochemical parameters were essentially normal.

In view of the aforementioned radiological features, a diagnosis of anterior mediastinal mass to rule out invasive thymoma with SVCS was made. CT-guided biopsy was done, and it confirmed a diagnosis of malignant thymoma.

Following the radiological and pathological diagnosis of malignant thymoma, he was then worked up for debulking surgery, and this was to be followed by radiotherapy and/or chemotherapy. At the time of intubation during anesthetic procedure, the patient developed cardiac arrest. He was resuscitated, but there was no significant improvement on the level of consciousness. The planned surgery was stopped, and he was immediately transferred to the intensive care unit, where he had assisted respiration and other critical management, aimed at reviving him. He remained comatose for 15 days. Urgent brain and repeat chest CT scans were ordered. The chest findings were not different from the earlier findings, but the brain CT revealed loss of gray-white matter differentiation, sulcal effacement, and compression of the cerebral ventricles, all in keeping with cerebral edema, presumably due to hypoxic encephalopathy. There was no intracranial focus of infection or metastatic deposit seen. He died on the 15th day of intensive care management.

Discussion

Thymoma is the most common neoplasm of the anterior mediastinum, accounting for 20%–25% of all mediastinal tumors and 50% of anterior mediastinal masses.[4] It is generally a rare tumor with a largely indolent growth pattern. It does, however,

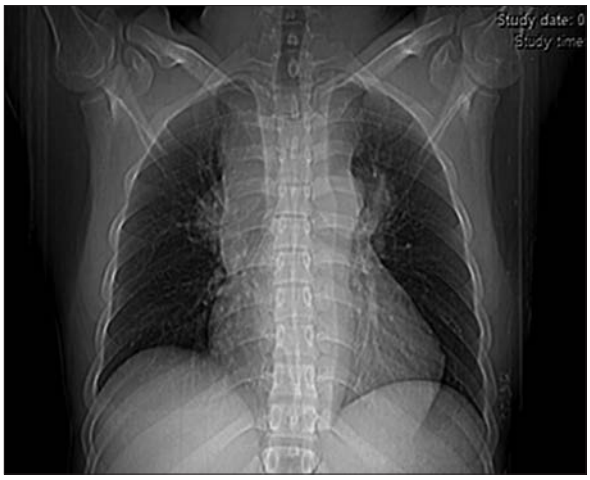

Figure 1: A scanogram of the chest in a computed tomography study showing superior mediastinal widening and a lobulated opacity of soft-tissue density in both paratracheal regions, more to the right and extending to the hila

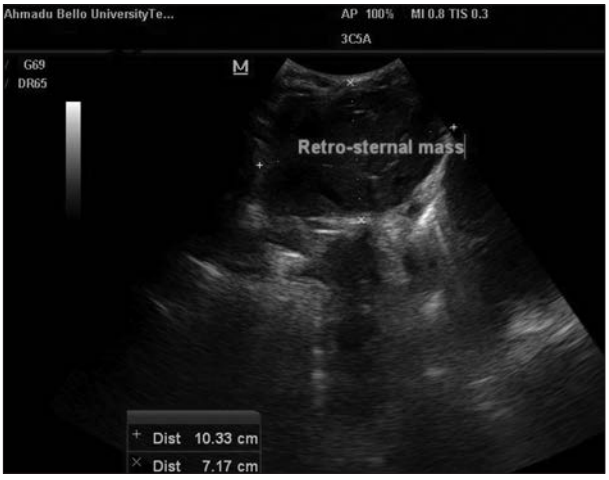

Figure 2: Ultrasonogram through the suprasternal approach, showing a lobulated mixed echogenic retrosternal mass, measuring 10.3 cm × 7.2 cm in dimension (original image)

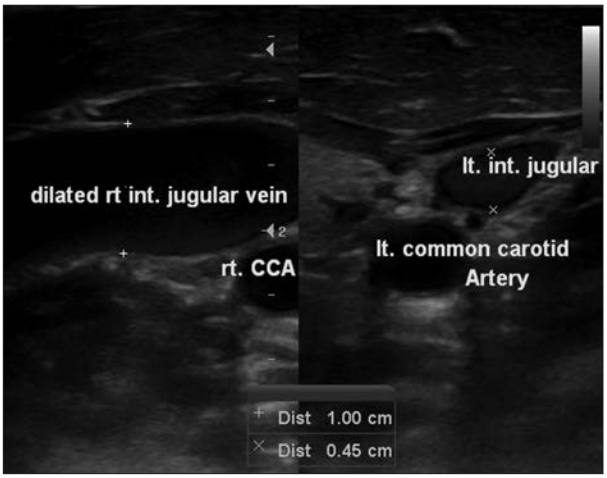

Figure 3: Doppler ultrasound of the neck of the same patient showing marked dilatation of the right internal jugular vein (original image)

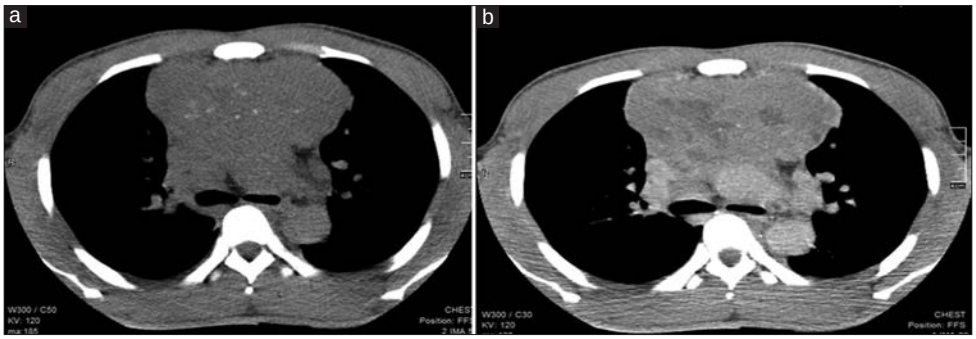

Figure 4: (a) Axial noncontrast computed tomography image at the level of the hilum showing lobulated isodense mass in the anterior mediastinum. Stippled hyperdense lesions of calcific density are also noted within the mass. The main bronchi are compressed (original image) (b) Contrast-enhanced computed tomography image at the level of the hilum as in Figure 4a showing heterogeneous enhancement following intravenous contrast administration (original image)

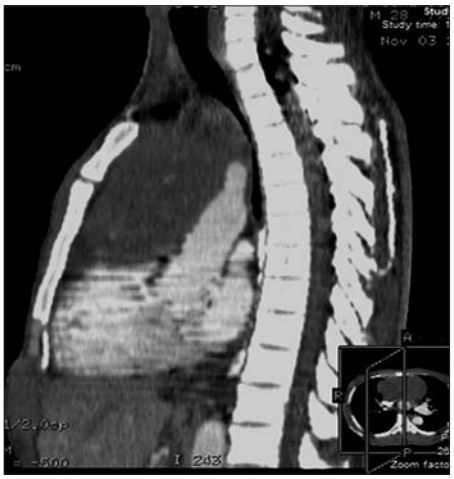

Figure 5: Sagittal reconstructed computed tomography image of the same patient as in Figures 4a and b above, showing significant compression of the trachea by the anterior mediastinal mass (original image)

have malignant potential as a result of its ability to invade locally and metastasize regionally.[5] It is often associated with a number of immune- and nonimmune-mediated paraneoplastic syndromes. Patient outcome is directly related to the stage of the disease and the ability to achieve a complete surgical resection.

Ninety percent of all thymomas occur in the anterior mediastinum, with the remainder occurring in the neck or other mediastinal areas.[6] Overall, however, it is considered a rare malignancy, with an incidence of 0.15 cases per 100,000.[7] There is a dearth of literature in Africa and particularly Nigeria on the prevalence of thymoma, although few cases have been reported in Enugu and Ibadan.[8,9]

Thymomas typically present in the fourth to sixth decades of life and do not exhibit gender predilection.[5] Tumor is uncommon in the young, making this case a rare presentation. Half of thymomas are asymptomatic and are detected incidentally on radiographic imaging, while half present with symptoms associated with paraneoplastic syndrome or with symptoms of local tumor growth such as cough, vague chest pain, dyspnea, and symptoms attributable to SVCS such as headache and distended neck veins. This patient presented with symptoms of local tumor growth as enumerated above.

Imaging is an essential part of the workup, and in conjunction with history and physical examination, it is often the only investigation required before treatment.[10] The goal of imaging is to identify the tumor and stage it appropriately, with emphasis on local invasion and distant spread, as invasion is a major prognosticating factor for survival.[10] The Masaoka staging is the most commonly used system to assess stage in thymoma.[11] Unlike tumor, node, and metastasis systems used for most cancer staging, the Masaoka staging system is based on the degree of invasiveness. Stage I tumors are completely encapsulated with no evidence of microscopic invasion. Stage II tumors have evidence of microscopic capsular invasion or macroscopic invasion into the surrounding fat or pleura. Stage III tumors invade locally into the surrounding structures, such as the lung, great vessels, and mediastinal structures, while Stage IV tumors present with more distant lymphatic or hematogenous metastases. 45%–80% of thymomas are visible by chest radiography[12] as seen in this patient where it appears as well-marginated and lobulated opacity of soft-tissue density projecting over the superior mediastinum.

CT is more definitive in the diagnosis of thymoma, with its increased sensitivity in identifying mediastinal masses, when compared with chest radiography. There has been one study published so far, comparing the CT appearance of 50 thymoma tumors to their Masaoka staging.[13] In that study, the authors attempted to separate Stage I disease from any capsular invasion. They found that larger tumors were more likely to be invasive, although no cutoff value was established. Furthermore, invasive tumors were more likely to be heterogeneous with low-attenuation areas within tumor, which was seen in 60% of invasive tumors, when compared with 22% of Stage I tumors[14,15]. Furthermore, CT findings of calcifications were more common in invasive tumors, seen in 54% versus only 6% of Stage I tumors. Lobulated contours were common in invasive tumors (59%) when compared with Stage I tumors (35%). In view of the

aforementioned research findings, this patient most likely had an invasive tumor as the CT appearances of tumor showed lobulation, heterogeneous density, stippled calcifications, and minimal enhancement following intravenous contrast administration [Figures 4a and b].

The mainstay of treatment for thymoma is surgery; the goal is to achieve a complete surgical resection which is the main determinant of survival.[16] This is quite true for encapsulated tumors. Adjuvant therapy, specifically postoperative radiation, is often recommended for invasive thymoma as in this patient, regardless of the resection status. In distant metastatic disease through hematogenous or lymphatic spread, debulking surgery coupled with postoperative radiotherapy and chemotherapy are the recommended treatment protocols.[16] Chemotherapeutic agents such as cisplatin, ifosfamide, doxorubicin, and cyclophosphamide in combination or single agent, together with corticosteroids, have been found useful. This index patient with invasive tumor was slated for surgery (debulking or total excision based on intraoperative findings) and postoperative radiotherapy and chemotherapy. He, however, had cardiac arrest at the time of intubation before the planned surgical excision.

On a general note, prognosis is worse for symptomatic thymomas as it was in this index case. The most important factor that determines prognosis is invasion of adjacent structures. Stage I disease usually has >90% 5-year survival rate, while Stage IV disease is usually associated with <25% 5-year survival rate.[16]

Conclusion

A rare case of malignant thymoma with SVCS in a young man has been presented. The usefulness of imaging in its staging and prognostication are further re-echoed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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