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CASE REPORT

Surgically treated aggressive mediastinal sarcoma: a tumor of possible thymic origin

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Abstract True sarcoma of thymus has no epithelial component. This tumor behaves aggressively with invasion of adjacent structures. Only a few anecdotal case reports of this tumor are found in the English-language literature, and they describe a poor outcome. This case report describes a rapidly growing thymic sarcoma diagnosed at an advanced stage with a compressive effect on the heart. The tumor was resected en bloc with adjacent invaded structures.

Key words Thymus · Thymic sarcoma · Thymoma

Introduction

Thymic sarcomas are rare tumors composed of sarcomatoid cells on the background of thymic parenchyma. The World Health Organization (WHO) classification describes them as thymic carcinoma in which part of or the entire tumor resembles soft tissue sarcoma.[1] True sarcoma of the thymus, however, has no epithelial component, is rare, and is not well documented in the literature.

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Case report

A 69-year-old man presented with fever, dry cough, and progressive dyspnea for the past 4 months. Clinically, air entry was reduced in the left hemithorax with elevated jugular venous pressure. He had no myasthenia gravis. Chest radiography revealed a large mediastinal mass. Contrast-enhanced computed tomography (CECT) of the chest showed an 18×18 cm enhancing mass in the anterior mediastinum with the bulk of the tumor lying on the left side (Fig. 1). A preoperative image-guided Tru-Cut biopsy of the tumor revealed it to be a spindle cell tumor.

The tumor was approached through an upper median sternotomy extending through an anterolateral thoracotomy at left fifth intercostal space. The tumor had a compressive effect on the heart. The patient had cardiac arrest while the tumor was being assessed for resectability. The arrest was probably due to cardiac tamponade because of the combined effect of pericardial effusion, weight of the tumor, and pressure due to retraction of the chest. The superficial part of the tumor was delivered out of the chest, and pericardiotomy was done to release the effusion and perform internal cardiac massage. Pericardiotomy and internal cardiac massage led to revival of the heart and return of blood pressure to normal. We decided to continue the procedure as the patient was stable after restoration of the cardiac rhythm and blood pressure.

There was a $20 \times 18 \times 7$ cm tumor in the thymic region adherent to the pericardium and anterior segment of the

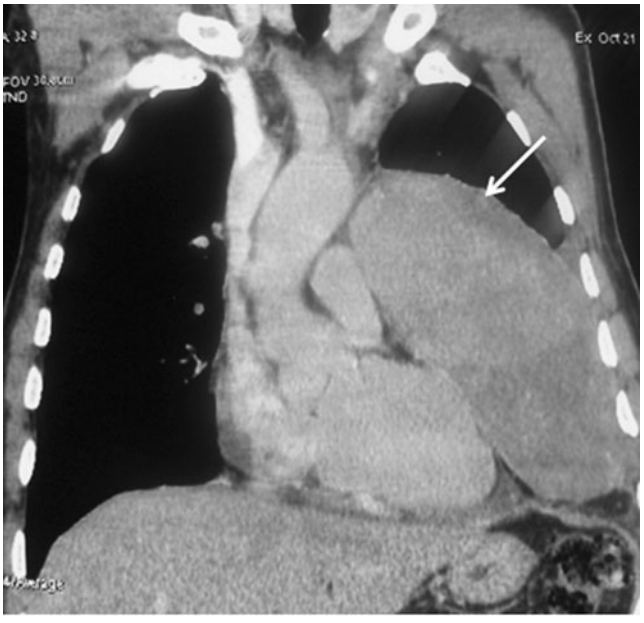

Fig. 1 Contrast-enhanced computed tomography chest coronal section shows a huge enhancing anterior mediastinal mass (*arrow*) closely abutting the anterior chest wall, the main pulmonary artery, and the pericardium

left upper lobe lung engulfing the left phrenic nerve (Fig. 2a,b). The trachea and bronchus were not involved by the tumor. En bloc tumor excision was performed that included the involved pericardium, left phrenic nerve, and a wedge of the left upper lobe of the lung. He was electively ventilated and extubated 12 h after surgery.

At histopathology (Fig. 2c,d), it was diagnosed as a pleomorphic sarcoma of the thymus with a small part of normal thymic parenchyma measuring 4×3 cm in the upper margin of the tumor capsule. Immunohistochemistry of tumor tissue was positive for S-100, smooth muscle antigen, vimentin, CD 34, and α-actin. The lymph nodes excised were free of tumor, and the patient had an uncomplicated recovery. Postoperatively, the patient was treated with radiotherapy to the tumor bed and combination chemotherapy. At the 18-month follow-up, he was doing well with normal CECT of the chest.

Discussion

Thymic tumors with both epithelial and spindle cell components have been described by authors using different nomenclatures. Thymoma with sarcomatoid differentiation is a known entity and has been described as thymic sarcomatoid carcinoma or thymoma with pseudosarcomatous stroma. Most of these tumors were biphasic tumors with a sarcomatoid component associated with thymoma.[1,2] In the English-language literature, however, there were only anecdotal case reports of purely sarcomatoid thymic tumors.[3,4]

It was apparent from both intraoperative observation and gross examination of the excised specimen by the pathologist that the tumor had arisen from the left lobe of the thymus gland. Histological examination of sections from the tissue present over the capsule of the tumor was confirmatory of thymic tissue. The standard protocol followed by the pathologist for sectioning tumors is one section per centimeter in the largest dimension of the tumor. In the index case with a 20-cm tumor, 20 sections were obtained for adequate sampling of the tumor. All the sections were analyzed histopathologically and immunohistochemically by the pathologist. The tumor was reported as pure thymic sarcoma due to the absence of any epithelial component.

It is not uncommon to encounter hemodynamic abnormalities due to compressive effects by a huge mediastinal mass. We routinely have cardiopulmonary bypass (CPB) ready for all patients with invasive thymomas where there is evidence of infiltration of major mediastinal vessels based on preoperative CECT of the chest. There was no preoperative radiological evidence of involvement of major mediastinal vessels in the index case, and so the need for CPB was not expected in this patient.

Thymomas with spindle cell differentiation that exhibit cellular atypia and mitotic activity are reported to have aggressive clinical behavior and a poor outcome.[5] All of the reported cases developed recurrence of the tumor despite radical resection and adjuvant combination chemotherapy and radiotherapy.[3,4] Our patient had a stage III tumor as described in clinical postsurgical staging for thymoma.[6] There is no literature that describes adjuvant therapy for this rare neoplasm. Because of the reported high recurrence rate and uniform fatality with this tumor, our patient was treated with adjuvant combination chemotherapy and external radiotherapy to the tumor bed to prevent tumor recurrence. The chemotherapy regimen consisted of five cycles of combination chemotherapy using cyclophosphamide, adriamycin, and carboplatin. He also received total field radiation of 40 Gy (2-Gy fractions, five fractions per week over 4 weeks). This was followed up with tumor boost radiation of 20 Gy (2-Gy fractions, five fractions per week over 2 weeks).

Conclusion

A scant literature comprising anecdotal case reports makes it difficult to establish a preferred treatment for thymomas. We believe that radical thymectomy with

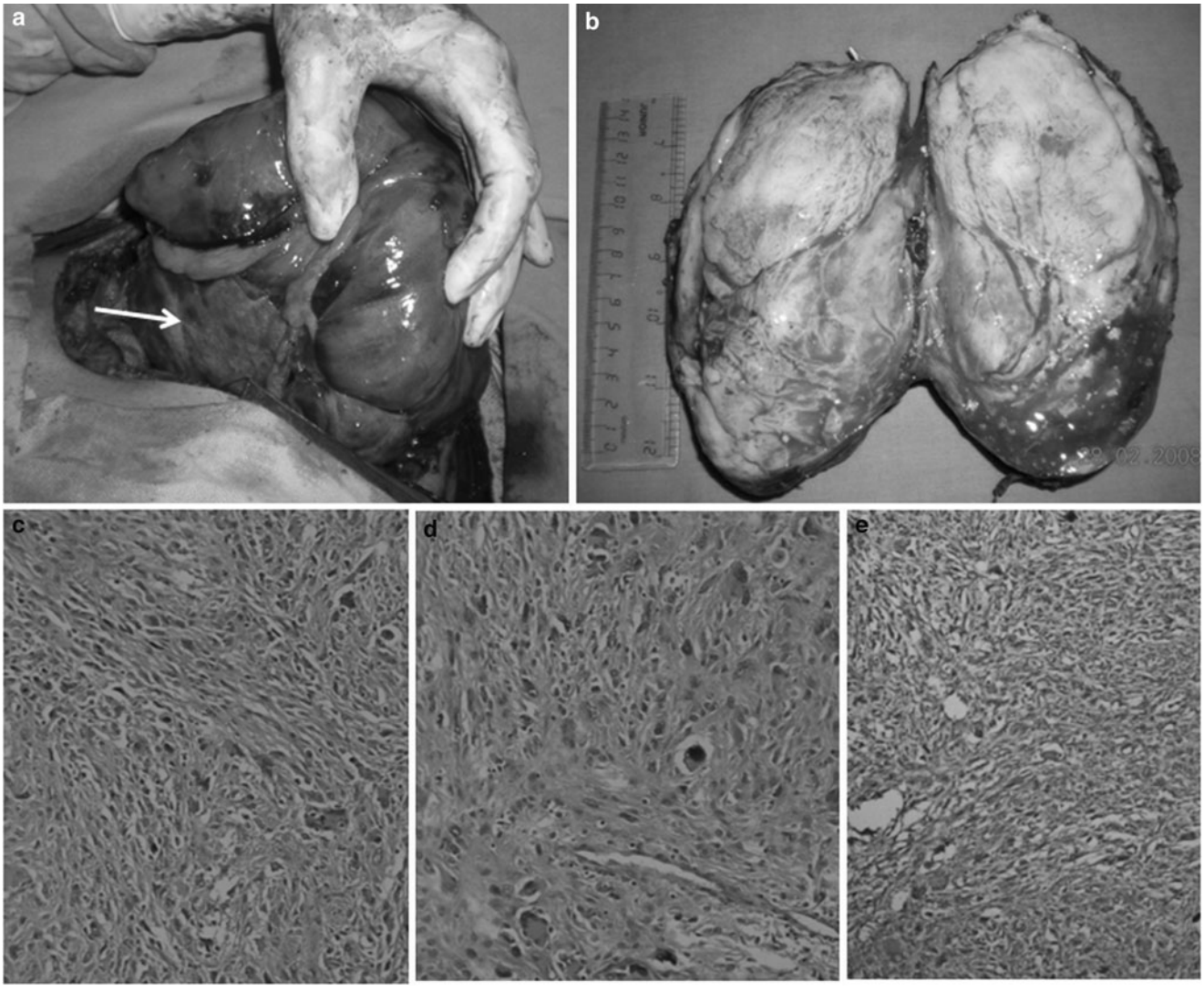

Fig. 2 Operative photographs show (**a**) the tumor with invaded pericardium (*arrow*) and (**b**) the cut surface of the tumor, which indicates a solid tumor with a lobulated appearance. **c** Photomicrograph shows a malignant spindle cell lesion with marked pleomorphism and tumor giant cells (H&E, ×200). Other photomicrographs show (**d**) tumor giant cells and atypical mitosis (H&E, ×200) and (**e**) immunohistochemical positivity for vimentin (immunoperoxidase, ×200)

postoperative radiotherapy to the tumor bed and aggressive systemic chemotherapy should be offered to these patients.

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