Case Report

Middle Mediastinal Thymoma of Unusual Pathologic Type

Jarosław Kużdżał, MD, PhD,[1] Marcin Zieliński, MD, PhD,[1]
Bolesław Papla, MD, PhD,[2] and Artur Szlubowski, MD[1]

Among the many sites of ectopic thymoma development, the middle mediastinum is the rarest — in the English language literature we found only two case reports. We present a case of 69-year-old woman with slow-growing, ectopic middle mediastinum thymoma of an very unusual histological type, neither classified in the Müller-Hermelink nor the World Health Organization (WHO) classification. (Ann Thorac Cardiovasc Surg 2006; 12: 200–2)

Key words: mediastinum, thymoma

Introduction

Although a vast majority of thymomas are located in the anterior mediastinum, there have been many sites of ectopic localization of thymomas described. Ectopic thymomas were found in the superior and the posterior mediastium, in the neck, but also in rare locations: at the base of the skull,[1)] intrapericardially.[2)] in the lung parenchyma,[3)] and in the pleural cavity.[4)] In the English literature we found only two case reports of a thymoma located in the middle mediastinum.[5,6)] We present a case of a slow-growing ectopic thymoma located in the middle mediastinum, and of a very rare histologic type, not classified in the Müller-Hermelink nor the World Health Organization (WHO) classification.

Case Report

A 69-year-old woman was referred to our department because of a great mediastinal mass, diagnosed 15 years earlier. The patient initially refused an invasive diagnostic work-up and an operation so she was only followed up radiologically. She remained asymptomatic and control chest X-rays showed slow, but systematical enlargement of the tumor (Fig. 1), and finally she decided to undergo an operation.

After admission to our department the CT scan showed a mass adherent to the anterior chest wall, the right atrium, the superior vena cava and the ascending aorta (Fig. 2). The borders of the tumor were sharp, and there were no signs of lymph node enlargement as well as no signs of metastatic disease on the ultrasound and CT imaging of the abdomen. The diagnosis of the benign mediastinal tumor was made and the patient was scheduled for surgery.

The patient was operated on through the anterolateral thoracotomy in the 5th right intercostal space. We found a 10×10×7 cm large tumor with a pedicle arising from between the ascending aorta and the superior vena cava, covered by parietal pleura and protruding to the pleural cavity. Neither had signs of infiltration of the lung nor any structures of the mediastinum. The only connection of the tumor with the mediastinum was its pedicle, so it should be considered to be of a primary middle mediastinal origin. The vascular pedicle of the tumor was sutureligated and divided, then the tumor was removed from the chest.

The postoperative course was uneventful. The pathologic study of the specimen showed an extremely rare type of thymoma with pseudosarcomatous stroma, neither classified in the Müller-Hermelink nor the WHO classification (Fig. 3). The tumor contained two distinct components, seen throughout the specimen and being clearly

From [1]Department of Thoracic Surgery, Pulmonary Hospital Zakopane, Zakopane, and [2]Chair and Department of Clinical and Experimental Pathology, Jagiellonian University, Cracow, Poland

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Address reprint requests to Jarosław Kużdżał, MD, PhD: Department of Thoracic Surgery, Pulmonary Hospital Zakopane, ul. Gładkie 1, 34-500 Zakopane, Poland.

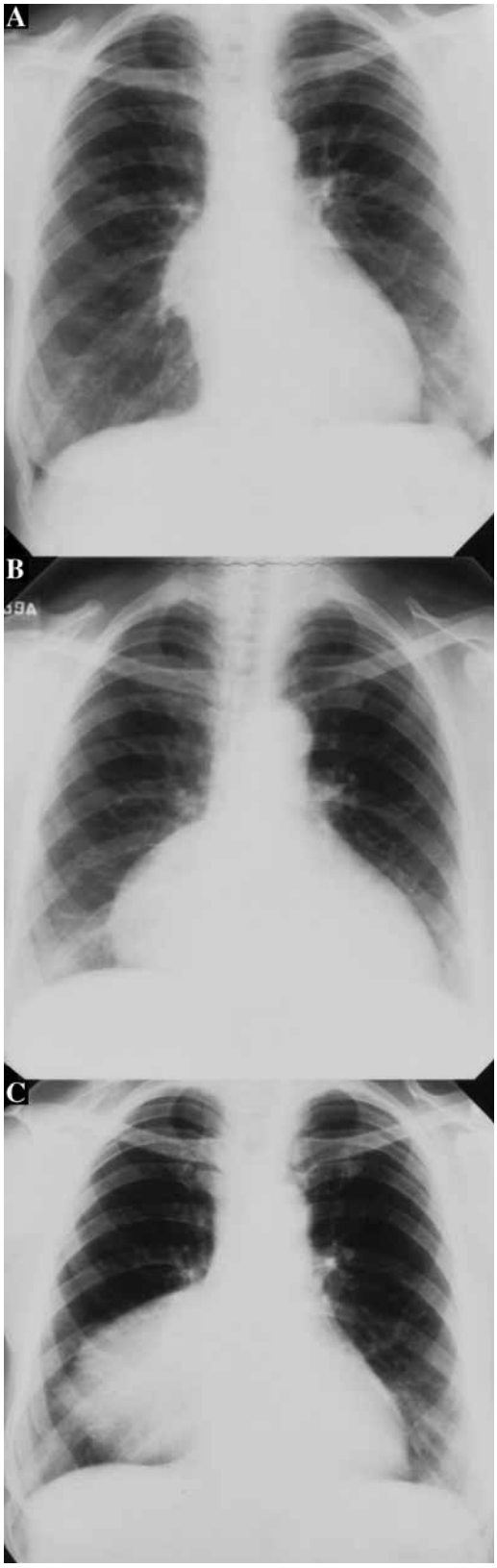

Fig. 1. Follow-up chest X-rays in 1988 (**A**), 1995 (**B**) and 2003 (**C**) showing slow, but systematic enlargement of the tumor.

separated. One component was composed of tightly packed medium-sized epithelial cells with round nuclei and scant cytoplasm, whilst the other, mesenchymal, consisted of spindle cells with elongated nuclei. Neither of these components showed atypia nor mitotic figures. Such tumor was recently referred to as "metaplastic thymoma".[7] The histological features of this very rare tumor are described in detail elsewhere.[8]

Comment

The thymus gland develops from the third and fourth branchial pouches and — in the course of the embryological development — descends into the anterior mediastinum. As a result of disturbances of this process, foci of the thymic epithelial cells may persist in different areas of the mediastinum. In fact, the presence of ectopic thymic tissue in the mediastinum has been shown by Masaoka,[9] and then in some other studies. In our own series of 100 patients with nonthymomatous myasthenia we found ectopic thymic tissue in the cervical (10%), the perithymic (37%) and the left and right pericardiophrenic area fat pads (both 7%), as well as in the aorto-pulmonary window (33%) and the aorto-caval groove (4%).[10] It may be hypothesized, that ectopic thymomas arise from these ectopic foci of the thymic tissue. Only a small percentage of thymomas were found out of the anterior mediastinum; in these cases the location was, in decreasing prevalence in: the superior mediastinum, the neck and the posterior mediastinum.

In our patient the whole tumor was located in the middle mediastinum, and had a blood supply exclusively from the pedicle arising from between the ascending aorta and the superior vena cava. It may be suspected, that the source of thymoma in our patient was the ectopic focus of the thymic tissue located in the middle mediastinum.

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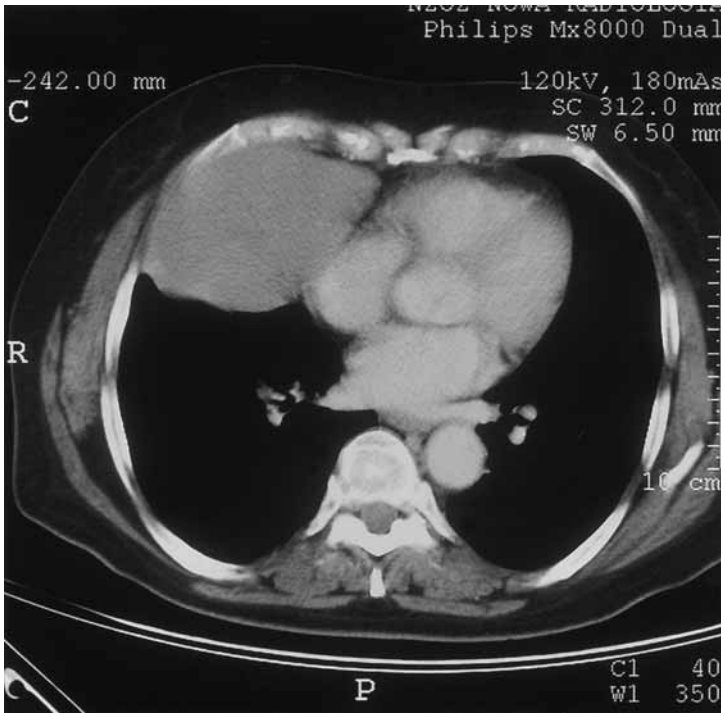

Fig. 2. CT scan showing a 10 cm mass adherent to the anterior chest wall, the right atrium, the superior vena cava and the ascending aorta.

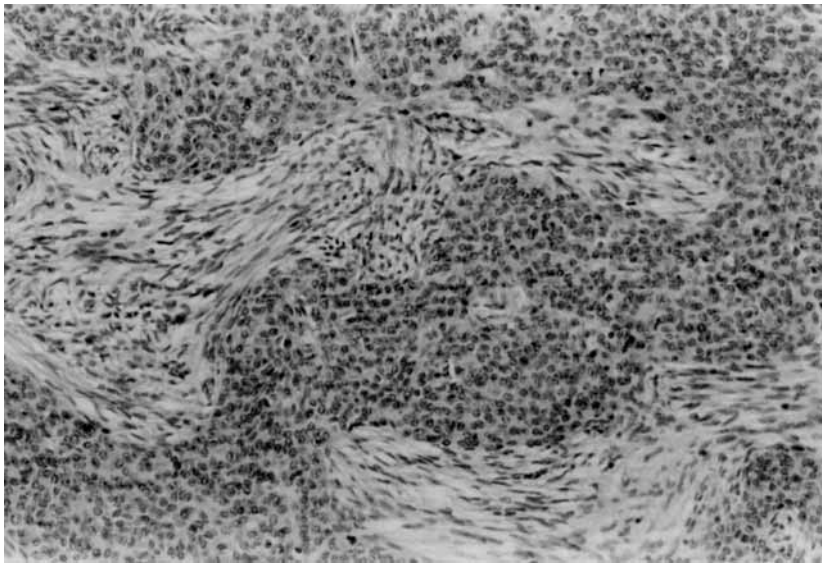

Fig. 3. Tumor with mixed histology, epithelial and pseudosarcomatous. (HE stain: ×140)

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