Cystic thymoma coexisting with vascular dysplasia

T. PAPADAS¹, P.A. DIMOPOULOS², F. SAMPSONAS³, N. MASTRONIKOLIS¹, K. SPIROPOULOS³, P.D. GOUMAS

¹Department of Otolaryngology, ²Department of Radiology, ³Department of Pneumology, University Hospital, Rion, Patras (Greece)

Abstract. — We report the case of a 23-year-old female patient, who was admitted to our hospital because of aggravating pain in the right shoulder, right chest and ear accompanied by cough and dyspnea. Imaging revealed a soft tissue mass in the mediastinum in close relation with an additional large mass extending in the right upper thorax, right axilla and neck. The tissue obtained during surgery, showed the former mass to be a cystic thymoma, being in contact with an extensive vascular dysplasia. The characteristic clinical presentation, the contribution of imaging, the results of operation, and the follow-up are rather unique. The review of the literature did not reveal similar cases.

Key Words: Cystic thymoma, Mediastinum, Vascular dysplasia, CT imaging.

Introduction

Thymomas, epithelial neoplasms of the thymus, represent the most common primary neoplasms of the anterosuperior mediastinum. They usually appear in adults older than 40 years of age with 70% of the cases occurring in the 5th to 7th decade². They affect males and females equally. Symptomatic patients present either with mass effects on adjacent organs or with systemic effects attributed to one of the paraneoplastic syndromes associated with thymomas. About 30%-50% of the thymomas is associated with myasthenia gravis, whereas hypogammaglobulinemia and red cell aplasia have also been described³.

Cystic thymomas represent 40% of all thymomas. Their main histological features include multiple cystic cavities, focally lined by squamous, cuboidal or columnar epithelium, and nests of non-neoplastic thymus within the cyst wall. They are often associated with acute and chronic inflammation, fibro-vascular proliferation, necrosis, haemorrhage, and cholesterol granuloma formation³.

Soft tissue lesions of the chest wall in adults include infection, hemangioma, plasmacytoma or extension of a primary bone tumor, and masses such as lipoma, liposarcoma, neurogenic or desmoid, and elastofibroma dorsi. Although hemangioma is a relatively common lesion in young children, a vascular dysplasia of that size is rare in adults.

Furthermore, coexistence of mediastinal cystic thymoma and large vascular dysplasia has not been described till now, therefore we present this case.

Case Report

A 23 year old female was admitted to our Hospital with a history of progressive pain in the right hemithorax, shoulder, and ear, accompanied by cough and dyspnea.

Clinical examination revealed a palpable, soft and non-tender mass on the right supraclavicular fossa.

Posteroanterior chest radiograph showed several round calcifications in the right supraclavicular and axillary regions, the larger of which had a diameter of 2 cm. Their appearance was compatible with phleboliths. The right upper lung was less extended than the left one but no difference in transradiancy was noted (Figure 1A). On the lateral chest radiograph a faint opacity was seen in the upper retrosternal space (Figure 1B).

Computed tomography (CT) scan confirmed the presence of a soft tissue mass in the right upper mediastinum, which extended upwards, sur-
rounded and restricted the right upper lung, and occupied the right subclavian fossa, the right side of the neck and the axillary region. The chest wall component of the mass had as posterolateral limit the subscapular muscle, and posteromedial the superior and inferior serratus muscles, and showed a mixed attenuation. After contrast administration, CT demonstrated some dilated vascular channels transversing the lesion and containing several phleboliths. However the underlying osseous structures, including the right thoracic ribs had a normal appearance. The subclavian vein was dilated and had a tortuous course (Figure 2).

During open biopsy a soft and vascular mass was found in the upper right hemithorax, surrounding the right lung apex, and extending in the mediastinum, the supraclavicular region and the neck. Samples obtained from multiple sites revealed reactive non-specific inflammatory tissue, some small inflammatory lymph nodes, and calcifications adherent to vessel walls.

On thoracotomy, the mediastinal component of the mass was adjacent to the pericardium and in close contact with the large vessels.

Although one could not discern any difference between the mediastinal mass and the one surrounding the lung, biopsy revealed that the former was a thymoma. Excision of the mediastinal lesion and of a large part of the upper chest and subclavian mass followed (12 × 8 × 5 cm). Histology revealed cystic thymoma in conjunction with vascular dysplasia.

The patient had an uneventful postoperative course and was discharged in good health. During the last three years, she has been examined successively and found to be in good condition, whereas CT showed no alteration of the remaining dysplastic tissue.

**Discussion**

Coexistence of cystic thymoma with a vascular dysplasia in a young patient has not been found in the literature.

Most of the cystic thymomas are of developmental origin from a remnant of the third branchial pouch-derived thymopharyngeal duct\(^4\,^5\), whereas some of them are of a degenerative or reactive nature, arising from cystic dilatation of Hassall’s corpuscles. It should also be reminded that Hodgkin’s disease and, less frequently, non-Hodgkin’s lymphoma can be accompanied by prominent secondary cystic changes of the thymic parenchyma. In a recent clinicopathological study in patients with cystic thymomas, 9 out of 25 patients were asymptomatic and their mediastinal tumor was discovered on routine chest radiograph, while the remaining sixteen patients had presented symptoms of chest pain and cough\(^6\), as in our case.

The radiographic manifestations of thymoma range from a subtle mediastinal contour abnormality, a frequent finding similar to that shown
on the lateral chest radiograph of our patient, to a large, anterior mediastinal mass. Larger thymomas are commonly unilateral and tend to protrude into one hemithorax, a feature encountered also in our case. As it has been reported, CT proved to be a very accurate imaging modality providing information on the precise anatomical location of the lesion, its relationship to surrounding structures, and tissue attenuation. Even in cases, in which clinical symptoms arise suspicions of thymoma undetectable with both chest radiographs, CT or Magnetic Resonance Imaging (MRI) can reveal the mass and define the degree of invasion in the adjacent organs.

Surgical diagnosis and therapy of thymoma are rendered simultaneously through excisional biopsy, although the latter yielded no positive results in this case. This outcome intensifies the controversial results of fine-needle aspiration biopsy, when it is used as an alternative.

The important distinction to be made is whether the lesion is invasive or not, since it determines the treatment plan and the patient’s outcome. Therefore, beyond the macroscopic observations made at surgery, the pathologist should also search for microscopic evidence of capsular invasion, extra-capsular extension, implantation or metastasis involving pericardium, pleura or lung parenchyma. Many pathologists prefer the term “invasive thymoma”, rather than “malignant thymoma” because the classic features of malignancy, such as nuclear atypia and high mitotic activity, are not seen in invasive lesions.

In this case, the thymic component of the lesion was adherent to the pericardium but the latter was easily removed during operation and no invasive element in any site of the removed tissue was found. Treatment of thymomas is determined after the staging of the lesion, which is based upon the presence of invasion and/or metastatic disease. In general, complete surgical resection is the preferable treatment. Radiotherapy may be used as adjuvant treatment in patients with incomplete resection or advanced disease. The prognosis of patients with thymoma is related to the stage of the tumor. Several other factors

Figure 2. A, B. Dilated vascular channels transversing the lesion, containing several phleboliths, with dilated subclavian vein.
also influence prognosis, including degree of surgical resection and the presence of parathymic syndromes.  

Since the coexistence of cystic thymoma and vascular dysplasia has not been reported in the literature, we can only speculate that they both originate from a common primitive tissue.

The benign histological characteristics of the removed thymic and dysplastic tissue, the fact that the patient remains free of symptoms over the last 5 years, and the stable imaging appearance favor the aspect that the lesions were benign and not invasive.

The patient is under annual follow-up.

References


