Case Report

Good Syndrome with Thymic Adenosquamous Carcinoma — Report of a Case

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A 68-year-old man with recurrent bilateral severe pneumonia and invasive thymic carcinoma was admitted to our hospital. An extended thymo-thymectomy with lymph nodes dissection was performed for an irregular shaped anterior mediastinum mass. The tumor was mainly composed of type C, adenosquamous carcinoma, and found to have a small area of types B2 and B3 thymoma. History and laboratory findings were compatible with the diagnosis of Good syndrome. Although there are some reports of thymic carcinoma arising from thymoma, this is the first report of co-existence of adenosquamous carcinomas and thymoma with Good syndrome as far as reviewed articles. Thymic carcinoma with severe infection should be examined carefully for co-existence of thymoma, and co-existence of thymoma and thymic carcinoma suggests a close histogenetic relationship between the 2 tumors. (Ann Thorac Cardiovasc Surg 2007; 13: 50–2)

Key words: thymic carcinoma, thymoma, adenosquamous carcinoma, Good syndrome

Introduction

Good syndrome is classified as a large entity, immunodeficiency with thymoma, not thymic carcinoma. On the other hand, thymoma and thymic carcinoma represent different points within a spectrum of differentiation and are histogenetically closely related entities. We report on a case of Good syndrome with adenosquamous thymic carcinoma, and with a small area of thymoma. Complete surgical excision and detailed pathological examination should be performed as the treatment for thymic tumors to find invasive or malignant potential.

Case Report

A 68-year-old man was admitted to our hospital with a week history of cough, dyspnea, fever and general fatigue. The chest X-ray showed severe pneumonia of bilateral lower lobes. Laboratory examination showed severe inflammatory findings (WBC 19,100/µL, CRP 22 mg/dl). A computed tomographic scan revealed bilateral pneumonia, 10×10 mm and 60×40 mm heterogeneous masses and invasion to the left innominate vein and left upper lung (Fig. 1). The patient recovered from pneumonia after 3 weeks of mechanical ventilator using antibiotics. Thymic carcinoma was strongly suspected considering the high level of SCC (33 ng/ml). Brain and an abdominal scan and bone scan were also performed to rule out metastasis from this tumor.

An extended thymo-thymectomy with a lymph node dissection was performed. As the tumor was massively invading the left lung and left brachiocephalic vein with no pleural or pericardial dissemination, partial resection of the left upper lung and left brachiocephalic vein was performed. Macroscopically, the resected tumor was 70×45×30 mm in diameter and histological examination confirmed that the tumor was directory invasive to the left upper lung and left brachiocephalic vein with multiple intrathymus metastasis. On macroscopic and microscopic serial section examinations one tumor appeared to be separated from the others. Furthermore, the tumor was mainly composed of type C (Fig. 2B), adenosquamous...
Adjuvant radiation (40 Gy) was performed postoperatively. The patient was admitted to the hospital several times with recurrent pneumonia in both lungs and treated successfully with antibiotics. Laboratory examination demonstrated hypogammaglobulinemia (6%). The serum immunoglobulins were markedly decreased (IgG, 520 mg/dl; IgA, 35 mg/dl; IgM, 3 mg/dl). History and laboratory findings were compatible with the diagnosis of Good syndrome, and there has been no finding of recurrence in 5 years.

Discussion

Primary thymic epithelial neoplasms exhibit a wide spectrum of morphologic and biological characteristics. WHO classification was proposed in 1999, as an agreement to the classification system based on the morphology of epithelial cells as well as the lymphocyte-to-epithelial cell ratio.\(^1\) In rare cases, thymic carcinoma can arise in thymoma. Only 36 cases of thymic carcinoma and thymoma were reported from the review of the literature.\(^2\)–\(^5\) Thymoma and thymic carcinoma represent different points within a spectrum of differentiation and are histogenetically closely related entities. Additionally, as the thymic tumor often consists of various areas of carcinoma or several thymoma types, it is difficult to diagnose on a limited biopsy, and complete surgical excision and detail pathological examination should be performed as the treatment for these tumors to find invasive or malignant potential.

Suster and Rosai reported on the largest series of 60 patients with thymic carcinoma, which did not include adenocarcinoma.\(^3\) The overall survival rates were reported 57, 40, or 33% at 1, 3, or 5 years respectively, and 30 patients (50%) developed metastasis, most commonly to the lymph nodes, bone, lung, and liver. Truong et al. reported on 13 cases of thymic carcinoma which included 1 case of adenocarcinoma.\(^2\) Although thymic adenocarcinoma is very rare with a poor prognosis, there were no findings of recurrence in 5 years in our case.

Thymoma has been associated with both humoral and cellular immunodeficiency and 10% of patients with thymoma have hypogammaglobulinemia, as Good syndrome, which is characterized by remarkably severe infections such as recurrent sinopulmonary infection due to encapsulated bacteria.\(^6\) Good syndrome refers to a rare group of thymoma and adult-onset immunodeficiency, often in 4th or 5th decade, that is characterized by low or absent B cells in the peripheral blood, hypogammaglobulinemia, and variably defects in cell-mediated immunity with CD4+ T lymphopenia and inverted CD4:CD8+ T-cell ratio.

On the other hand, a common variable immunodefici-
ciency, the age of onset which shows a biphasic age which peaks at 1–5 years and 16–20 years, is a heterogenous group of primary antibody deficiency syndromes with recurrent infections that is associated with cellular immune defects. The reduced number of peripheral blood B cells is typical in Good syndrome but altered B cell maturation is found in the majority in common variable immunodeficiency patients. While a formal diagnostic definition of Good syndrome has not been developed, Good syndrome is classified as a large entity, immunodeficiency with thymoma, separated from common variable immunodeficiency. Moreover, thymoma has also been associated with cellular immunodeficiency without hypogammaglobulinemia and presents with several infections such as chronic mucocutaneous candidiasis, cytomegalovirus or herpes virus infections, and pneumocystis carinii pneumonia. Thymectomy has not resulted in effective therapy for cellular immunodeficiency.

The co-existence of thymoma and thymic carcinoma suggests a close histogenetic relationship between 2 tumors and this is very important not only for understanding of these tumors but it may play a significant role in the assessment of the biological behavior and treatment of these tumors, and thymic carcinoma with several infections should be carefully examined to rule out Good syndrome.

References