

Thymoma Associated with Myasthenia Gravis and Sjögren Syndrome

Y-M Tsai¹, Y-C Lin², C-H Chen³, C Tzao¹

ABSTRACT

Thymoma is the most common neoplasm of the anterior mediastinum but thymoma with Sjögren syndrome (SS) is rare. Sjögren syndrome is a systemic autoimmune inflammatory disorder. It is characterized by lymphocyte-mediated destruction of exocrine glands, which leads to absent glandular secretion. Here, we present the case of a 63-year-old man with thymoma and concurrent myasthenia gravis and SS, who achieved remission after thymectomy.

Keywords: Autoimmune disease, myasthenia gravis, Sjögren syndrome, thymoma

Timoma Asociado con Miastenia Grave y Síndrome de Sjögren

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RESUMEN

El timoma es la neoplasia más frecuente del mediastino anterior, pero un timoma acompañado del síndrome de Sjögren (SS) constituye una ocurrencia rara. El síndrome de Sjögren es un trastorno inflamatorio autoinmune sistémico. Se caracteriza por la destrucción – mediada por linfocitos – de las glándulas exocrinas, lo cual conduce a la ausencia de secreción glandular. Aquí presentamos el caso de un hombre de 63 años de edad con timoma, y miastenia gravis y SS concurrentes, que logró la remisión después de una timectomía.

Palabras clave: Enfermedad autoinmune, miastenia gravis, síndrome de Sjögren, timoma

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INTRODUCTION

Thymoma is the most common neoplasm of the anterior mediastinum. It is often associated with various autoimmune diseases such as myasthenia gravis (MG), rheumatoid arthritis, scleroderma, polymyositis, and systemic lupus erythematosus. Myasthenia gravis is the most important association, and it can be found in 30–50% of patients with thymoma (1). Sjögren syndrome (SS) is an autoimmune rheumatic disease; it refers to keratoconjunctivitis sicca and xerostomia, which result when immune cells attack and destroy the lacrimal and salivary glands (2). The clinical symptoms of SS can be highly variable. It may present with atypical features such as peripheral neuropathy and pulmonary insufficiency, but dry eyes and dry mouth are

common conditions (3). Diagnosis can be made according to results of the Schirmer test, radiologic nuclear medicine salivary scans or biopsy samples of the salivary-gland tissue. Patients with early-stage thymoma are often asymptomatic, and it can be found incidentally on a chest film or a chest computed tomography (CT) scan. The incidence of both MG and SS in a patient with thymoma is rare. We present the case report of a patient who had chronic cough and an incidental finding of a thymoma combined with MG and SS.

CASE REPORT

A previously healthy 63-year-old man presented with productive cough and easy fatigability for two to three months prior to admission. He had a history of dry eyes with artificial tear-drops treated for one year. Physical examination revealed drooping of the upper eyelid. Laboratory findings showed that white blood cell count was 6700/mm³, haemoglobin, 13.8 g/dL, platelets, 168 000/mm³ and C-reactive protein, 0.17 mg/dL. The liver and renal function tests were normal. Chest radiograph revealed a huge mass lesion over the left anterior mediastinum. Chest CT scan showed a

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heterogeneous lobulated mass, which was mainly located at the left aspect of the prevascular space (Figure A). Myasthenia gravis was diagnosed by clinical examination

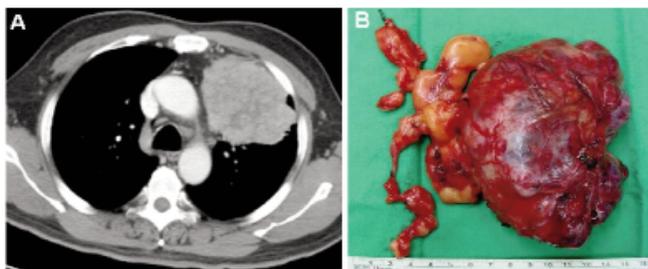


Figure: (A) Chest computed tomography showed a 10 cm lobulated mass in the left upper mediastinum. (B) An encapsulated tumour about 11.5 x 8 x 5 cm in size and 255 g in weight was excised.

acetylcholine receptor antibodies > 4.0 nmol/L (normal range, 0–0.5 nmol/L) and repetitive nerve stimulation test was positive. Blood tests for rheumatic diseases revealed that the antinuclear antibody titer was 1: 80, SSA/RO antibody was 0.3 U/ml, and SSB/LA antibody was 0.1 U/ml. Schirmer test was positive, and sialo-scintigraphy showed impaired function of the right parotid gland and both submandibular glands. The clinical diagnosis was thymoma with MG and SS. The patient underwent thymectomy and the postoperative specimen showed a well-encapsulated mass measuring 11.5 × 8 × 5 cm in size and 255 g in weight (Figure B). The histological diagnosis was type-A thymoma with spindle-shaped epithelial cells and minimal amount of small lymphocytes in the stroma. The patient was discharged on the 10th postoperative day. The dry eyes had improved without artificial tear-drops treatment at two months after operation. The patient had no tumour recurrence or metastasis at the 24-month follow-up.

DISCUSSION

Thymomas are rare epithelial neoplasms. They are usually encapsulated, slow-growing, space-occupying tumours associated with symptoms of dyspnoea, chest pain, cough, and symptoms of MG (4). They can present as lesions, locally invasive diseases, or metastatic diseases and account for about 45% of masses of the anterior mediastinum (1). Thymomas have been reported with many autoimmune diseases including MG, systemic lupus erythematosus,

rheumatoid arthritis, polymyositis, and progressive systemic sclerosis (5). There were approximately 50–70% of thymomas associated with autoimmune or paraneoplastic disorders (1), and MG was observed to occur in 47% of non-malignant thymoma patients (6). Sjögren syndrome is the second most common autoimmune rheumatic disease and is characterized with xerostomia and xerophthalmia [sicca symptoms] (1). The prevalence of SS is estimated to be 1–3%. It affects women nine times more than men and typically occurs between 40 and 60 years of age (7). Few cases of SS and concurrent thymoma with MG have been reported (8, 9). Surgical excision is the recommended treatment because of the potential occurrence of malignancy (1). In the index case, the symptoms and signs persisted for one year and eventually improved without tumour metastasis, after thymectomy. Although the current case is rare and the aetiology of this association remains controversial, careful history taking and physical examination are most important for early diagnosis and treatment. Thymoma should be included in the differential diagnosis when a patient complains of xerostomia and xerophthalmia, although thymoma associated with MG and SS is rare and surgical excision is the recommended therapy.

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