Case Report

Thymoma Associated with Hypergammaglobulinemia: A Case Report

Diana Taheri 1, Hamid Reza Suzangar 1, Roya Sherkat 2, Sina Biria 2, Abbas Tabatabai 3, Mehdi Fesharakizadeh 4

1- Dept of Pathology, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.
2- Dept of Infectious Disease, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.
3- Dept of Surgery, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.
4- Dept of Surgery, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran.

ABSTRACT

Thymoma is a term that should be restricted to neoplasms of thymic epithelial cells, irrespective of the presence or the number of lymphocytes. The usual location of thymoma is the anterosuperior mediastinum; however, this tumor can also occur in other mediastinal compartments, in the neck, within the thyroid gland, in the pericardial cavity, the pulmonary hilum, within the lung parenchyma, or the pleura itself (sometimes coating it in a mesothelioma-like fashion). The association of thymoma with hypogammaglobulinemia, Muco-cutaneous candidiasis, systemic lupus erythematosus and myasthenia gravis is well known. Here we present a patient admitted in our hospital with buttock rashes regarded to be zoster of the buttock. In his past medical history he had recurrent cutaneous infections, lupus erythematosus, hypergammaglobulinemia, recurrent oral candidiasis and B-cell dysfunction. During the patient evaluation, a mass was found in the mediastinum and percutaneous needle biopsy confirmed the diagnosis of thymoma (type B3). This is the second case reported having thymoma in association with hypergammaglobulinemia.

Keywords: Thymoma, Hypergammaglobulinemia, Systemic lupus erythematosus

Introduction

Thymoma is a term that should be restricted to neoplasms of thymic epithelial cells, independently of the presence or number of lymphocytes. Seminoma, Carcinoid tumor, Hodgkins lymphoma, and non-Hodgkins malignant lymphoma, can all involve the thymus; thus they can be viewed as thymic tumors but should not be regarded as variants of thymoma. Nearly all thymomas present in adult life. Thymomas in children are exceptional; most of the cases so diagnosed in the past actually represent lymphoblastic lymphomas of the thymus. However, some well- documented cases exist, most of them occurring near the age of puberty, with an appearance and behavior equivalent to that of their adult counterpart, including an occasional association with myasthenia gravis. Thymoma is divided into A, B1, B2, B3 and C types, and is associated with a variety of diseases (1).

The most important of which are: Systemic lupus
erythematous (2-6), Mucocutaneous candidiasis (7,8), hypogammaglobulinemia (9-12), myasthenia gravis (13), T-cell and B-cell immune deficiency(14-16) and pure red cell aplasia (17).

In our case report, thymoma is associated with hypergammaglobulinemia , which is the second case report having thymoma in association with hypergammaglobulinemia.

**Case report**

A 43 year-old man admitted in Alzahra Hospital (Esfahan, Iran) for zoster of the buttock region. He was found to have Systemic lupus erythematosus previously treated with Prednisolon and Chloroquine.

Also he had a history of recurrent oral candidiasis, mean while he was a known case of B-cell dysfunction with negative CD19 and CD20 markers in the Flowcytometry of peripheral blood (normal=87 to 536) and hypergammaglobulinemia.

Physical examination revealed decreased breathing sounds and in the chest X-ray, a mass was found in the anterosuperior mediastinum.

CT scan of the thorax confirmed the mediastinal mass and also a mass found in the right lung parenchyma.

Lab data: ANA = 3+, ds DNA = 260 (Positive) IgG = 2450 mg/dl (800-1900), IgM = 225 mg/dl (50-250), CD19 = Negative, CD20 = Negative IgA = 265 mg/dl (70-350), C3 = 45 (50-140), C4 = 6 (10-40), CH50 = 115 (120-350)

In the next step, CT guided percutaneous needle biopsy of both the mediastinal and pulmonary lesions were performed.

After microscopic evaluation, the diagnosis of thymoma (type B3) was made (A type of thymoma mainly composed of epithelial cells having a round or polygonal shape and exhibiting no or mild atypia). They are admixed with a minor component of lymphocytes, resulting in a sheet like growth of the euplastic epithelial cells (Fig. 1, Fig. 2).

The Immunohistochemical staining results were as follows: CD20: Negative / CD5: Negative / CD99: Positive / CK: Positive in epithelial cells.

We decided to treat the patient with chemotherapy in order to reduce the tumors size and make them operable.

The chemotherapy regimen, included Cyclophosphamide, Adriamycin, Cysplatin, Bleomycin, Etoposide and Prednisolon, and his treatment is being continued.

**Fig. 1:** Epithelial cells are admixed with lymphocytes. (H & E staining ×400)

**Fig. 2:** Epithelial cells are admixed with lymphocytes. (H & E staining ×100)

**Discussion**

Thymoma is a term that should be restricted to neoplasms of thymic epithelial cells, independently of the presence or number of lymphocytes. Almost all thymomas present in adult life. Thymomas in children are exceptional; most of the cases so diagnosed in the past actually represent lymphoblastic lymphomas of the thymus However, some well-documented cases exist, most of them occurring near the age of puberty, with an appearance and behavior equivalent to that of their adult counterpart, including an occasional association with myasthenia gravis (1).

The usual location of thymoma is the anterosuperior mediastinum; however this tumor can also occur in other mediastinal compartments (although a posterior location is very rare), in the neck (18), within the thyroid, within the pericardial cavity, in the pulmonary hilum, within the lung parenchyma (19) or in the...
pleura itself sometimes coating it in a mesothelioma-like fashion (20,21).

Radiographically, thymoma usually presents as a lobulated shadow that may be calcified. CT scan and MRI are the methods of choice for preoperative diagnosis and evaluation of its extent (22). Fine needle aspiration has been used with success, the diagnosis of thymoma being based on a finding of a dual population of epithelial cells and lymphocytes with the appropriate cytologic feature (23).


In many case-reports the association of thymoma with Systemic lupus erythematosus (2-6), Mucocutaneous candidiasis (7,8), Hypogammaglobulinemia (9-12), Myasthenia gravis(13) and pure red cell aplasia (17) is reported but there was only one article about the association of thymoma and hypergammaglobulinemia, which is reported by San Filippo J in Cutis in 2006 (2).

Case report

As stated above, the association of thymoma with hypogammaglobulinemia is mentioned in a great number of articles and also in the pathology literatures. While in the two cases mentioned above thymoma is associated with hypergammaglobulinemia. In our case thymoma is associated with hypergammaglobulinemia despite negative CD19 and CD20 markers in peripheral blood smear.

Considering the paucity of reported cases of thymoma associated with hypergammaglobulinemia, large multicenter studies are still required to best identify this correlation.

References


