ARTICLE IN PRESS

REV COLOMB REUMATOL. 2016; xxx(xx): xxx-xxx



Revista Colombiana de REUMATOLOGÍA



www.elsevier.es/rcreuma

Case Report

Haemolytic anaemia secondary to thymoma without myasthenia gravis as parathymic syndrome: A case report[☆]

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ARTICLE INFO

Article history:
Received 18 December 2015
Accepted 15 March 2016
Available online xxx

Keywords: Thymoma Haemolytic anaemia Autoimmunity

Palabras clave: Timoma Anaemia hemolítica Autoinmunidad

ABSTRACT

The case is presented of a 41 year-old male patient with chest pain, asthenia and adynamia. The imaging studies showed a mass in the anterior mediastinum, which according to the pathology report, was a thymoma. Also, the patient also had haemolytic anaemia and autoimmune hypothyroidism, and with no associated myasthenia gravis.

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Anaemia hemolítica secundaria a timoma sin miastenia gravis como síndrome paratímico: presentación de un caso

RESUMEN

Describimos el caso de un paciente masculino de 41 años que cursa con cuadro clínico de dolor torácico, astenia y adinamia, con estudios imagenológicos que evidencian masa en mediastino anterior que corresponde a timoma, de acuerdo con el reporte de patología. Además cursa con anemia hemolítica e hipotiroidismo autoinmune, sin miastenia gravis asociada.

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PII of original article: S0121-8123(16)30020-2

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^{*} Please cite this article as: Hurtado Amézquita C, Páez Ardila HA, Pabón Duarte L, Tiusabá Rojas PC. Anemia hemolítica secundaria a timoma sin miastenia gravis como síndrome paratímico: presentación de un caso. Rev Colomb Reumatol. 2016. http://dx.doi.org/10.1016/j.rcreu.2016.03.001

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Introduction

Thymoma is an infrequent neoplasm with an incidence of 1–5 cases per every million people/year, it occurs in all ages, with a peak incidence between 55 and 65 years.¹

The initial diagnostic approach of thymoma is accomplished through computed axial tomography of the chest with contrast; however, the surgical resection with pathological study is the most important diagnostic tool and in addition is the treatment of choice, especially in stages III and IV. Adjuvant chemotherapy with cyclophosphamide, doxorubicin and cisplatin can be used in the case of unresectable tumour.¹

Other pathological entities, so called parathymic syndrome, such as myasthenia gravis, bone marrow aplasia, hypogammaglobulinemia, among others, may coexist associated with the neoplastic picture.²

As evidenced in the case reports and review articles in the literature, the association between thymoma, haemolytic anaemia, autoimmune hypothyroidism and lupus-like syndrome is quite uncommon as parathymic syndrome, unlike the association between thymoma and myasthenia gravis, which is the most common and best studied association between this tumour and autoimmune disease.^{3–9}

Autoimmune haemolytic anaemia is defined as the destruction of erythrocytes secondary to the presence of antibodies directed against erythrocyte membrane antigens. It has an incidence of 0.61-1.3 per 100,000 inhabitants per year. The first diagnostic approach is made with a complete blood count with evidence of decrease in the haemoglobin and haematocrit values, usually characterized as being an anaemia with normal corpuscular volumes, elevation of lactic dehydrogenase (associated with greater severity of the haemolysis), decrease in haptoglobin, high reticulocyte count, presence of positive direct Coombs test (which indicates autoimmune aetiology of the anaemia), presence of warm or cold antibodies (immunoglobulin G and immunoglobulin M, respectively). Finally, the study is based on the search for the aetiology, primary cause (idiopathic) or secondary (neoplasms, collagen diseases, or pharmacological). 10,11

Autoimmune hypothyroidism or Hashimoto's thyroiditis is a chronic inflammation of the thyroid gland associated with an autoimmune component, being the most frequent autoimmune disease. Anti-peroxidase antibodies are the best serological marker for the diagnosis, being positive in 95% of patients, while the anti-thyroglobulin antibodies exhibit lower sensitivity (60–80%) and specificity than the abovementioned.¹²

The lupus like or lupus imitators refer to a group of entities with clinical and paraclinical characteristics, including the profile of antibodies, similar to those described in patients with systemic lupus erythematosus (SLE).¹³

Below we present the case report of a patient with thymoma, haemolytic anaemia, autoimmune hypothyroidism and lupus like as parathymic syndrome, with resolution of the haemolytic anaemia and negativity of antinuclear antibodies (ANA) after the surgical resection of the neoplasia at the mediastinal level.

Case presentation

A 41-year-old male patient, who consulted due to a clinical picture of 8 days of evolution consisting of epigastric throbbing pain, associated with asthenia and adynamia, without fever or other symptoms. With a history of hypothyroidism of difficult management under hormone replacement therapy and a family history of SLE and rheumatoid arthritis. On physical examination the patient was tachycardic, with mucocutaneous pallor. The rest of the physical exam did not show relevant findings.

During hospitalization, the initial blood count shows bicytopenia (leukocytes 2660 cells/mm³, haemoglobin 7.1 g/dl, MCV 97.7 fL), liver profile with hyperbilirubinemia at the expense of the indirect bilirubin (total bilirubin: 2.5 mg/dl, indirect bilirubin 1.54 mg/dl), positive Coombs test, with high corrected reticulocyte count (19.43%) and elevated lactate dehydrogenase 352 IU/l; and therefore a picture of autoimmune haemolytic anaemia is considered, requiring doses of systemic steroid. In addition, the chest X-ray (Fig. 1) evidences mediastinal widening, and therefore a chest CT scan is requested (Fig. 2) finding a mass in the right cardiophrenic angle, solid, with smooth contours that suggests infiltration of the pericardium, and diameters greater than $70 \, \text{mm} \times 65 \, \text{mm} \times 64 \, \text{mm}$, compatible with thymoma, without being able to rule out lymphoma. Given the finding of bicytopenia and a mass which could correspond to lymphoma, a bone marrow aspiration is carried out, which reported hypercellularity with haematopoiesis of the three cell lines, erythroid hyperplasia with megaloblastic change and negative study for neoplastic infiltration.

Subsequently it is decided to perform a trucut biopsy of the mediastinal mass which documents ovoid cells intermingled with small lymphocytes, with immunohistochemical studies positive for CKAE1AE3, with a lymphoid population of T phenotype immunoreactive for CD5,

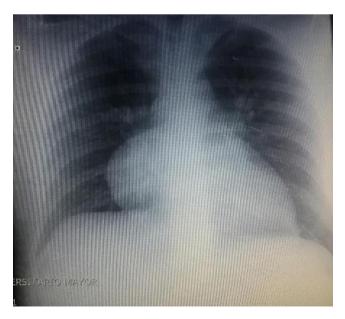


Fig. 1 - Chest X-ray.

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