Case Report
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Thymoma with pancytopenia: a case report and review of the literature

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Abstract

Although the most common hematological disturbance in thymoma is pure red cell aplasia, pancytopenia is a very rare paraneoplastic syndrome in this disorder. We only found one case in literature reviews.

We report a 36 year old female with an anterior mediastinal mass and severe pancytopenia. With a mediastinotomy incision, biopsy of the tumor was obtained. The report of the pathologist was thymoma of predominant epithelial type. After a median sternotomy, there was complete resection of the tumor with removal of two segments of the right upper lobe and the involved pericardium (stage 3 of Thymoma). Unfortunately, after complete resection of the tumor there was no improvement in the pancytopenia. The patient died because of sepsis about 33 days after operation.

Keywords: Thymoma, pancytopenia, red-cell aplasia, Thymectomy

Introduction

Forty percent of patients with thymoma have a parathymic or paraneoplastic syndrome (PTS); while one-third have two or more PTS [1].

The incidence of cytopenia but not pancytopenia reported by Verily (1985) and Lewis (1987) was 2% and 2.5%, respectively [2,3]. Hematological syndromes associated with thymoma are red cell aplasia, erythrocytosis, pancytopenia, megakaryocytopenia, T-cell lymphocytosis, acute leukemia and multiple myeloma [1,4].

In literature review we found only one report on pancytopenia and thymoma [5]. Approximately 25-33% of patients with red cell aplasia benefit from resection of thymoma [6].

Case report

A 36 year old female patient referred to our department with severe pancytopenia and anterior mediastinal mass. Preoperative hematological tests showed:

WBC	1.6×10^3
RBC	3.28×10^6
Hb	9 g/dl
HCT	27.4 %
MCV	83.5 fl
MCH	27.4 Pg
MCHC	32 g/dl
Platelet	30000×10^3

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Bone marrow aspiration and biopsy revealed a hypercellular marrow. Additional laboratory tests and images such as anti-nuclear antibody (ANA), abdominal and pelvic CT scans, did not find any cause of pancytopenia such as hypersplenism and autoimmune disease. After ruling out other causes of pancytopenia, thymoma was the perceived cause of pancytopenia. After blood and platelet transfusion and prednisolone administration (20mg) daily, mediastinotomy was performed.

A mass with mediastinal involvement was seen. A wedge resection was performed. The report of the pathologist was thymoma. One week after biopsy and a median sternotomy incision, the tumor, the involved part of the pericardium and two segments of the right upper lobe were radically resected. Four days after operation, CBC was done; the results of which included:

WBC	22×10^3
RBC	3.33×10^6
HB	9 g/dl
HCT	28%
MCV	98 fl
MCHC	32 g/dl
Platelet	60×10^3

Pancytopenia did not improve postoperatively. Thirty-three days after resection, the patient died because of sepsis.

Hematological tests thirty days after operation included:

WBC	1.9×10^3
RBC	2.22×10^6
Hb	6.6 g/dl
HCT	71.7 %
MCV	97.7 fl
MCHC	31.4 g/dl
Platelet	38000×10^3

Discussion

Patients with thymoma are often clinically asymptomatic [1]. The symptomatic patients may have only local symptoms related to the presence of the tumor within the mediastinum or only symptoms related to systemic disease states that are frequently associated with the presence of thymoma or a combination of both [1]. In most reviews in the literature, approximately 30 to 40% of patients have local symptoms and 30 to 50% have an associated systemic PTS [1]. Myasthenia gravis (MG) is the most common PTS encountered [1,7,8,9]. This syndrome (MG) is present in approximately 30 to 59% of patients with thymoma [8,9].

In Rosenow and Hurley's review, 40% of patients with thymoma had one PTS, and one-third had two or more PTS [10].

Hematological syndromes associated with thymoma include red cell aplasia, erythrocytosis, pancytopenia, megakaryocytopenia, T-cell lymphocytosis, acute leukemia, and multiple myeloma [1].

The incidence of cytopenia but not pancytopenia, published by Souadjian, Verily and Lewis are 15%, 2% and 2.5%, respectively [2,3,11].

Association of thymoma with severe anemia

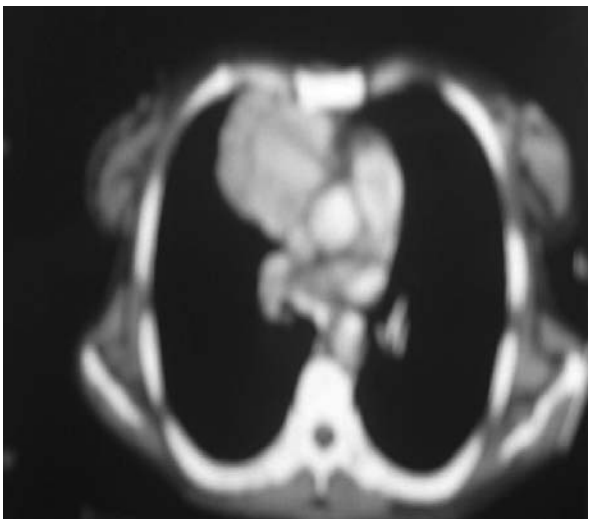

Fig. 1. CT scan of the patient showing an anterior mediastinal mass in the right hemithorax.

caused by suppression of erythrogenesis in the bone marrow is well established [1, 4].

When red cell precursors are greatly reduced or absent, myelo- and megakaryocytic poiesis usually are normal or increased [2, 3].

Burrows and Carrol however reported the occurrence of pancytopenia [5], the only one we found in the literature [5]. In this report the mechanism of erythrocyte aphasia and pancytopenia is not clear [5]. But, Tepson and Vas suggested that it is immunologically mediated [12].

The presence of IgG antibodies that are cytotoxic for erythroblasts and inhibit erythropoietin or hemoglobin synthesis, or both have been observed in these patients [1,4].

The immunodeficiency is generally attributed to a decreased number of B-lymphocytes in the bone marrow and peripheral blood, as noted by Cooper and Butler. The T-cells are usually normal in number [13].

Out of 56 patients with red cell aplasia reviewed by Hirst and Robertson, seven also had myasthenia gravis and two had hypogammaglobulinemia[14].

Of interest was the report of Suzuki and colleagues on the occurrence of both pure red cell aplasia and myasthenia gravis after resection of an invasive thymoma [15].

According to Beard and colleagues, fifty percent of the patients with red cell aplasia have associated thymoma. 70% of these patients have noninvasive spindle cell variant of the epithelial subgroup of thymoma [1].

Zeok and associates reported that approximately 25 to 33% of patients with red cell aplasia benefit from excision of the thymoma [6]. And most as noted are associated with the spindle cell subtype of thymoma, which normally has a good prognosis [1].

Our patient had pancytopenia with invasive thymoma of the epithelial type. Thirty-three days after resection, our patient died because of sepsis.

Acknowledgment

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