

Thymectomy in Red Cell Aplasia - A Case Report

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1. Abstract

1.1. Background: Pure Red Cell Aplasia (PRCA) is a very rare syndrome responsible for severe anemia due to absence or significant reduction of erythroid precursors in the bone marrow. It's etiology is variable, ranging from a congenital disease to an acquired one. Secondary acquired PRCA may be associated with several conditions, including Thymic Tumors.

1.2. Case Presentation: A 42-year-old male presented to the Internal Medicine department with constitutional symptoms: non-intentional weight-loss, palpitations, somnolence. Clinical investigation showed a severe normocytic normochromic anemia, with significant reticulocytopenia. Due to clinical status, several blood transfusions were needed. Further investigation with bone marrow biopsy showed intact precursors for platelets and leukocytes, with absence of erythroid precursors; the primary diagnostic hypothesis was Pure Red Cell Aplasia, with no etiology well defined, however. In order to rule out other conditions, imaging scans were performed; the chest CT showed a large mediastinal expansive formation, compatible with Thymoma. Patient was submitted to Thymectomy by the Thoracic Surgery team, with posterior improvement of clinical and hemodynamic status. Even so, for definitive remission of the PRCA, a bone marrow transplant was needed. The patient was successfully treated and remains healthy so far.

1.3. Conclusion: In the presence of Pure Red Cell Aplasia, it's essential a thorough clinical investigation to define different possible etiologies, including, as stated in this case, a Thymic Tumor. It's important, also, to consider PRCA when the patient is diagnosed with Thymoma. The surgical treatment is essential in such cases.

2. Introduction

Pure Red Cell Aplasia (PRCA) is a rare syndrome defined as a normocytic normochromic anemia with severe reticulocytopenia and marked reduction or absence of erythroid precursors from the bone marrow, with intact precursors for platelets and leukocytes, which are normal in number and morphology in the peripheral blood [1]. Due to the rarity of the disease, PRCA can never be evaluated in large controlled clinical trials; as a result, the majority of recommendations are based on small series of patients. It's important to consider the etiology of PRCA in order to define the treatment, even though, most of the times, immunosuppression therapy will be needed. This syndrome can be inherited or acquired; the congenital PRCA is also called Diamond-Blackfan syndrome and most commonly presents during the first year of life; the acquired etiologies are multiple. Primary acquired PRCA is an autoimmune disorder frequently antibody-mediated. Secondary acquired PRCA may be associated with collagen vascular/autoimmune disorders; infections, particularly B19 parvovirus; thymoma and other solid tumors [2]. Considering that scenario, a patient diagnosed with PRCA, generally, has a long way of clinical investigation to define the possible etiology. In this paper, we discuss a clinical case conducted by our department of a patient diagnosed with PRCA, with the finding of a mass in the anterior mediastinum, compatible with Thymoma, who underwent Thymectomy as one of the lines of treatment.

3. Case Report

A 42-year-old male patient was admitted to the Oswaldo Cruz University Hospital in January 2021, presenting with progres

sive non-intentional weight loss, generalized asthenia, exertional breathlessness, hyporexia, palpitations and somnolence starting 5 months prior consultation.

The patient had no known personal medical history, was considered by the family and by himself extremely healthy prior to the beginning of these symptoms.

Physical examination revealed mild tachycardia and tachypnea and extreme pallor, but no other expressive alterations in the cardiovascular/respiratory tract or abdominal physical exam.

The complete blood count revealed a hemoglobin (Hb) level of 5.5 g/dL (> 13.0), hematocrit of 16.1 (39.7 - 52.0), with normal mean corpuscular volume (MCV) and corpuscular hemoglobin concentration (MCHC). White cell count was 3960/mm³ (3,000-8,000), with a platelet count of 186000/mm³ (150,000 - 450,000) and reticulocyte count of 0.1% (0.5-2.5%). Other blood laboratory exams (including iron studies and vitamin B12) and EKG were normal.

Considering laboratory findings and clinical condition, multiple blood transfusions were needed to stabilize patient's status and get Hb > 9 g/dL. When such level was achieved, symptoms improved.

A bone marrow biopsy was made considering the suspicious of Pure Red Cell Aplasia, and it showed markedly reduced erythroid lineage, with no abnormalities in the lymphocytic, megakaryocyte or myeloid lineage. Based on these findings, the diagnosis of PRCA was finally made, with no etiology well defined however.

To elucidate the etiology of the PRCA in this patient, multiple tests were performed, including parvovirus testing; testing for rheumatologic disorders and adenosine deaminase (ADA), which all came back negative. Chest and abdominal CT scans were also performed: the chest CT scan came back with the findings of a large mediastinal expansive formation, in left paracardiac situation, with post-contrast enhancement, in close relation with the pericardial sac, measuring 12.0 x 9.7 x 9.0 cm, exerting compressive effect on the pulmonary artery trunk, being Thymoma the main diagnostic hypothesis.

A multidisciplinary team meeting was held with the Internal Medicine team; with a consultant hematologist and with the Thoracic Surgery team. Since hemoglobin levels were stable at that point, a surgical approach via video-assisted thoracoscopic surgery (VATS) was defined. Surgical specimen was sent to histopathological analysis, which final report concluded Thymoma Type A as the diagnosis of the tumor (Figure 1).

After surgery, patient had tremendous improvement of clinical and hemodynamic status, with initial remission of the disease. Even so, definitive remission was only obtained with bone marrow transplant, made a year after surgery. In clinical follow-ups, till this day, patient lives a healthy life.



Figure 1: surgical specimen of Thymectomy

4. Discussion

The epidemiology of PRCA it's uncertain, due to it's rarity. Some series of cases may give, however, an idea about that matter. A retrospective study conducted at Mayo Clinic between 1980 and 1994 reported a series of 47 cases of PRCA in adults. The disease was associated with some clinical disorders, such as leukemia, lymphoma and thymoma in this particular patients; the median age was 63 (range 22 to 88); the median hemoglobin concentration was 6.3 g/dL (range 3.0 to 8.4G/dL), with a reticulocyte count of 0.1% (range 0.1% to 0.6%). 4/47 patients had thymoma as the presumed cause of PRCA (8.7%) [3]. Even though it's described in the literature that 25-30% of the patients with PRCA caused by thymoma who underwent thymectomy had full remission of the anemia [4], an important study, also at Mayo Clinic, evaluated 13 adult patients with PRCA and Thymoma who underwent thymectomy, and none of these patients had definitive remission with the surgical approach alone, being necessary, for that, adjuvant immunosuppression therapy or bone marrow transplant [5].

PRCA should always be suspected in front of a patient presenting with anemia, symptomatic or not, with reticulocytopenia in the complete blood count. The clinical investigation to confirm the diagnosis includes a bone marrow biopsy, which should display an important reduction in erythroid lineage, with preservation of other lineages [6, 7]. To elucidate the etiology, a chest CT scan must be made, alongside with iron study, vitamin B12, EKG, and a thorough laboratory exams list. The treatment may vary according to the levels of hemoglobin and the etiology, but blood transfusions and immunosuppression is often required [8, 9].

5. Conclusion

The incidence of PRCA in patients with Thymoma varies between 2-5%. In these cases, the Thymectomy leads to initial remission in about 30% of the cases [10]. The definitive remission, though, may

need adjuvant treatments, such as medications and bone marrow transplant, as seen in the presented case. Is important, then, to be aware of the possibility of PRCA in the face of a Thymoma and other-wise as well; be aware of the possibility of a Thymoma being the etiology for a PRCA in clinical investigation [11, 12].

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7. Author Contributions

ADSS and ABSO gathered all of the informations needed for writing the case report. ADSS, ABSO wrote and reviewed the draft manuscript. DCCN and MCSMF reviewed the second version of the manuscript and reviewed the references. WWSA and CFMV guided the writing of the manuscript, and were responsible for the final review and considerations prior submission.

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