ECRONICON OPEN ACCESS

EC PULMONOLOGY AND RESPIRATORY MEDICINE

Case Report

# First Case of Association of Thymoma and Red Cell Aplasia after a Twenty Years Career: Case Report

**H Harmouchi[1]*, Y Ouadnouni[1,2], M Lakranbi[1,2] and M Smahi[1,2]**

*[1]Department of Thoracic Surgery CHU Hassan II of Fez-Morocco*

*2Faculty of Medicine and Pharmacy, Sidi Mohamed Ben Abdallah University, Fez-Morocco*

***Corresponding Author:** H Harmouchi, Department of Thoracic Surgery CHU Hassan II of Fez-Morocco.

**Received:** December 22, 2021; **Published:** February 23, 2022

## Abstract

Pure red cell aplasia (PRCA) is an uncommon pathology associated with normochromic normocytic anemia without leukopenia or thrombocytopenia. PRCA is defined by the absence or rarefaction of erythroblasts in the bone marrow, with normal myeloid and megakaryocytic lines, and the diagnosis is paraclinical, evoked on the hemogram. In this observation, we report a man of 63 year-old, who presents a PRCA associated to a thymoma and an infection by barvovirus B19. The patient was operated by vertical sternotomy, a total thymectomy enlarged to the mediastinal fat.

***Keywords:*** *A First Case; Association of Thymoma; Red Cell Aplasia*

## Introduction

Pure red cell aplasia (PRCA), first described in 1922 by Kaznelson [1], is defined by the absence or rarefaction (less than 5%) of erythroblasts in the bone marrow, with normal myeloid and megakaryocytic lines. The diagnosis of this pathology is paraclinical, evoked on the hemogram in front of a normochromic normocytic aregenerative anemia, and confirmed on the myelogram by objectifying a decrease or an absence of erythroid precursors, contrasting with a normal maturation of the myeloid and megakaryocytic lines. PRCA is uncommon, although its etiologies remain multiple, and can be constitutional (predominated by Diamond-Blackfan disease) or acquired (infectious, tumor, systemic or idiopathic etiology). This observation describes the association of an infectious (parvovirus B19) and autoimmune (thymoma) etiologies in a patient with PRCA.

## Case Report

This is a 63-year-old patient, with no particular medico-surgical history and not taking long-term medication, who consults a hematologist for isolated anemia, demonstrated following general fatigue with moderate dyspnea. Apart from these functional signs, the patient's history and clinical examination were unremarkable.

During the first consultation (05/06/2020), the complete blood count (CBC) showed anemia (Hb at 6.3 g / dl) normochromic normocytic, with a normal level of white blood cells and platelets (respectively 8690 / mm3 and 258000 / mm3), and the patient was transfused with three packets of red blood cells. This anemia was aregenerative according to a reticulocyte count at 5940 / mm3 (0.18%), and a bone marrow biopsy was performed in the sternum to read the myelogram after exclusion of deficiency, metabolic and inflammatory causes (serum iron, vitamin B12, folic acid, CRP, urea, creatinine). This reading of the myelogram revealed a granular rich marrow presenting notable qualitative abnormalities, a clear aplasia of the erythroblastic line (0%), and a relative excess in the lymphoid line at the level of lymphocytes (18%).

Faced with this relative increase in lymphocytes, lymphocyte immunophenotyping was performed showing an isolated increase in CD8 + T lymphocytes, interpreted as reactive lymphocytosis following infection with parvovirus B19 which came back positive (anti-parvovirus B19 antibody positive). Complement of the etiological assessment objectified a negative value of anti-nuclear antibodies and anti-native DNA antibodies, with correct values of protein electrophoresis, and negative serologies for hepatitis A, B, C, and Epstein Bar virus (EBV). The etiological assessment also consisted of performing a chest CT scan which demonstrated the presence of a resectable anterior mediastinal mass (Figure 1), a priori in favor of a thymic epithelial tumor (TET).

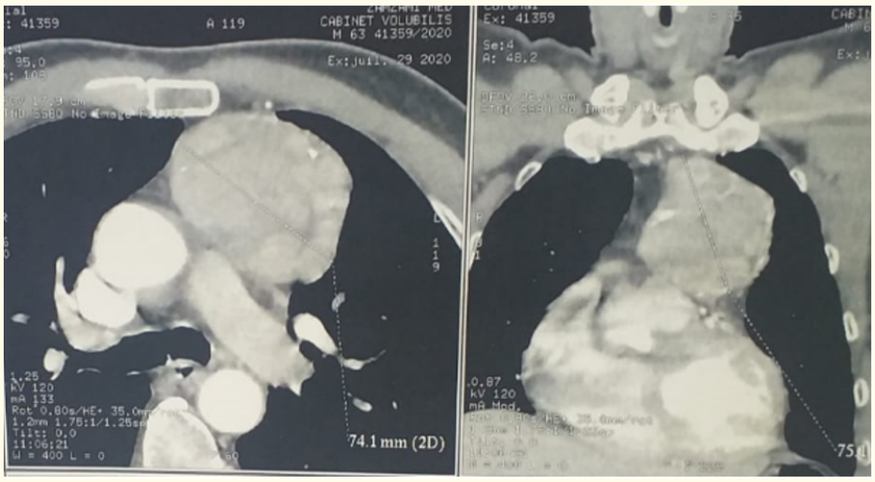


***Figure 1:*** *Thoracic CT scan showing a resectable anterior mediastinal mass in favor of thymoma.*

In the meantime, the patient beneficied of a control of blood cell count objectifying anemia (Hb at 5.5g / dl), with correct levels of white blood cells and platelets, and retransfusion with four packets of red blood cells was performed. By vertical sternotomy, a total thymectomy enlarged to the mediastinal fat was performed (Figure 2). The immediate postoperative follow-up was marked by transfusion of the patient with two packets of red blood cells since he was operated with an hemoglobin calculated at 8g / dl.

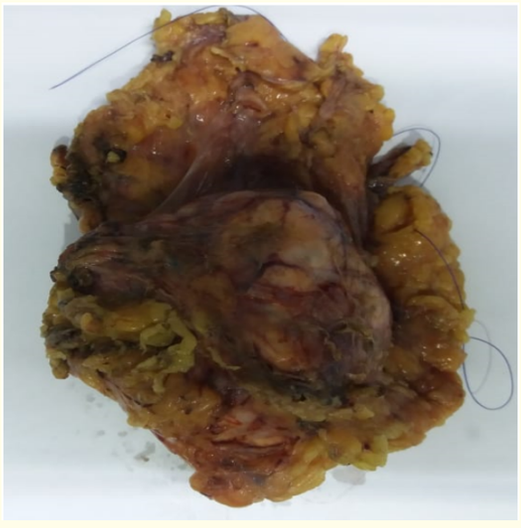

***Figure 2:*** *Specimen showing the thymoma and the mediastinal fat.*

## Discussion

PRCA is an uncommon pathology associated with normochromic normocytic anemia without leukopenia or thrombocytopenia. This is a condition that results from an interruption in the maturation of erythroid precursors in the bone marrow, at a more or less early stage depending on the etiology. This etiology may be primary predominated by Diamond-Blackfan disease which is characterized by anemia occurring in the neonatal period or infancy associated with morphological abnormalities involving the hands, face, or heart [2]. However, the most frequent etiologies of PRCA remain acquired, secondary to infection, blood disease, autoimmune disease, or without a detectable cause (idiopathic PRCA). This case report demonstrates the presence of a double causality of PRCA, on the one hand infection with parvovirus B19, and on the other hand the presence of a thymoma. This is the first case of PRCA associated with thymoma in our department after 12 years of performing thoracic surgery.

The most recognized infectious etiology responsible of PRCA is infection with parvovirus B19. This virus directly attacks human erythroid progenitors via the red blood cell surface P antigen (globoside) [3]. It is a virus whose inter-individual transmission is primarily respiratory, in a community context. However, maternal-fetal transmission or transmission through blood transfusion can occur. In immunocompetent people with a normal lifespan of red blood cells (120 days), parvovirus B19 causes erythropoietic aplasia of 5 to 10 days, and therefore does not cause significant anemia, and the infection is in order resolving after neutralization of the virus through a specific humoral-mediated immune reaction [4]. However, in immunocompromised patients, the infection may persist and lead to chronic anemia [4].

The presence of PRCA requires performing of chest CT scan to look for a TET that is present in around 10% of patients with PRCA, while this latter is only reported in 2 to 5% of patients with TET [5]. The mechanism by which a TET causes PRCA is not fully understood. Some authors speculate that this aregenerative anemia results from immune-mediated paraneoplastic destruction of erythroid precursors [6]. In the literature review, different histological types of the thymus were reported, including thymic hyperplasia, thymoma (all types), thymic carcinoma, but also thymolipoma [7,8].

The treatment of PRCA is symptomatic and etiological. Given the severity of the anemia, transfusions by red blood cells are usually necessary. For parvovirus B19 infection, there is no antivirals currently established clinical activity. Intravenous immunoglobulin (IVIG), which contains a large amount of anti-parvovirus B19 IgG, is the treatment of choice for erythroblastopenia secondary to this infection occurring in immunocompromised patients. The largest series studying this therapeutic option included 136 patients all immunocompromised, and had shown that treatment with IGIV seems to be effective in the short term [9]. However, further controlled studies are needed to better define the optimal dose to be administered, as well as the factors predicting relapse of erythroblastopenia. Surgical resection is generally recommended in combination with TFT, which will lead to a return to normal hematopoiesis in 25% to 30% of cases [10-12]. However, some authors have suggested that remission from surgery alone is actually quite rare and that most patients will require some other form of treatment [13]. On the other hand, erythroblatopenia may not appear until after a thymectomy, which was found in the series by Suzuki., *et al* collecting 136 myasthenic patients (all benefiting from a thymectomy) and of which 4 patients developed erythroblastopenia [14].

## Conclusion

The peculiarities of this observation is the presence of PRCA secondary to an association of an infectious etiology by parvovirus B19 and an autoimmune etiology (thymoma). PRCA is a rare disease, and this association of these etiologies is exceptional, but in front of any thymic epithelial tumor, the diagnosis of PRCA must be investigated.

## Conflict of Interest

The authors declare that they have no conflict of interest with this manuscript.

## Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

## Author Contributions

All the authors contributed substantially to the authorship of this manuscript.

## Ethics

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

## Informed Consent

Written informed consent was obtained from the patients for publication of this manuscript and any accompanying images.

## Bibliography

1. Kaznelson P. "Zur entstehung der blutplättchen". *Verh Disch Verhandlungen der Deutschen Gesellschaft* 34 (1922): 557.
2. Marije Bartels and Marc Bierings. "How I manage children with Diamond-Blackfan anaemia". *British Journal of Haematology* 184.2 (2019): 123-133.
3. Brown KE., *et al*. "Erythrocyte P antigen: Cellular receptor for B10 parvo virus". *Science* 262 (1993): 114-117.
4. Fisch P., *et al*. "Pure red cell aplasia". *British Journal of Haematology* 111 (2000): 1010-1022.
5. Thompson CA and Steensma DP. "Pure red cell aplasia associated with thymoma: clinical insights from a 50-year single-institution experience". *British Journal of Haematology* 135 (2006): 405-407.
6. M Lacy., *et al*. "Pure red cell aplasia: association with large granular lymphocyte leukemia and the prognostic value of cytogenetic abnormalities". *Blood* 87.7 (1996): 3000-3006.
7. Suto Y., *et al*. "Myasthenia gravis with thymus hyperplasia and pure red cell aplasia". *Journal of the Neurological Sciences* 224 (2004): 93-95.
8. David Ferreira., *et al*. "Pure Red Cell Aplasia Associated with Thymolipoma: Complete Anaemia Resolution following Thymectomy". *Case Reports in Hematology* (2018): 8627145.
9. Yoann Crabol., *et al*. "Intravenous immunoglobulin therapy for pure red cell aplasia related to human parvovirus b19 infection: a retrospective study of 10 patients and review of the literature". *Clinical Infectious Diseases* 56.7 (2013): 968-977.
10. Jacolien van der Marel., *et al*. "Thymoma with paraneoplastic syndromes, Good's syndrome, and pure red cell aplasia". *Journal of Thoracic Oncology* 2.4 (2007): 325-326.

11. D Jiang and M Wu. "Thymectomy resulted in complete remission of pure red cell aplasia when associated with thymoma". *The Journal of Thoracic and Cardiovascular Surgery* 58.4 (2010): 235-236.

12. Zeok JV., *et al*. "The role of thymectomy in red cell aplasia". *The Annals of Thoracic Surgery* 28 (1979): 257-260.

13. C Rosu., *et al*. "Pure red cell aplasia and associated thymoma". *Clinics and Practice* 1.1 (2011): 1.

14. Shigeaki Suzuki., *et al*. "Initial predictors of development of pure red cell aplasia in myasthenia gravis after thymectomy". *Clinical Neurology and Neurosurgery* 106.1 (2003): 16-18.

**Volume 11 Issue 3 March 2022**