Case Reports in Oncology

Case Rep Oncol 2020;13:76–78

DOI: 10.1159/000505533
Published online: February 6, 2020

Case Report

Case Report: Pure Red Cell Aplasia due to Angioimmunoblastic T-Cell Lymphoma

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Keywords

Pure red cell aplasia · Angioimmunoblastic lymphoma · Anemia

Abstract

Pure red cell aplasia (PRCA) is a rare bone marrow failure characterized by a progressive normocytic anemia and reticulocytopenia without leukopenia and thrombocytopenia. It can be associated with various hematological disorders but exceedingly rarely with angioimmunoblastic T-cell lymphoma (AITL). We report the case of a 72-year-old woman with PRCA associated with AITL. The patient presented with severe anemia (hemoglobin 2.6 g/dL) and a low reticulocyte count 0.7%. Direct and indirect Coombs tests were positive. A CT scan of the chest, abdomen, and pelvis revealed multiple lymphadenopathies. A cervical lymph node biopsy was compatible with AITL. A bone marrow biopsy showed medullary involvement by AITL and a severe erythroid hypoplasia with a myeloid:erythroid ratio of 19.70. The patient was started on CHOP and after 6 cycles the PET scan confirmed complete remission.

Introduction

Pure red cell aplasia (PRCA) is a rare bone marrow failure with a normocytic normochromic anemia and reticulocytopenia with a normal count of white blood cells and platelets. It can be associated with various diseases such as thymoma, lympho- and myeloproliferative disorders, autoimmune diseases, infections, and drugs. The hematological malignancies are an important group of causes of PRCA. Although various types of lymphomas may also be

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associated with this condition, the case of PRCA with angioimmunoblastic T-cell lymphoma (AITL) has rarely been reported [1, 2]. AITL is a rare condition that counts for 1–2% of all the cases of non-Hodgkin lymphoma; its diagnosis can be delayed for weeks or months with the majority of patients presenting in an advanced stage of disease. This type of lymphoma has an aggressive course. Occasional remissions are seen but with frequent relapses [3].

Case Report/Case Presentation

A 72-year-old woman presented to the emergency room with complaints of a 1-month history of progressive exertional dyspnea and fatigue. The patient denied blood loss, jaundice, fever or weight loss. The patient has a medical history of high blood pressure and dyslipidemia which was properly controlled. Epidemiological and social history was negative.

On physical examination severe pallor was observed, enlarged lymph nodes were identified at axillary, cervical, and inguinal levels; hepatosplenomegaly was not present. Laboratory blood examination showed the following: hemoglobin 2.6 g/dL, hematocrit 8.8%, mean corpuscular volume 85.1 fL, low reticulocyte count 6×10^9/L (0.7%), peripheral white cell count 7.5×10^9/L, platelet count 274×10^9/L, C-reactive protein 2.56 mg/dL, erythrocyte sedimentation rate 89 mm/h, lactate dehydrogenase 398 U/L, total bilirubin 0.80 mg/dL, ferritin 1,027 mg/mL, and haptoglobin 87.74 mg/dL. Direct and indirect Coombs tests were positive, and peripheral blood smear showed microspherocytes. Protein electrophoresis revealed hypergammaglobulinemia 33.5% with immunoglobulin G 1,669 mg/dL and immunoglobulin M 517 mg/dL. Hepatitis B and C virus, Epstein-Barr virus, human immunodeficiency virus, and human T-cell lymphotropic virus I and II serologies were negative. IgG and IgM CMV were positive and DNA quantification negative. IgG and IgM parvovirus B19 were positive but the DNA quantification (polymerase chain reaction testing) was negative. Renal and hepatic function were normal as well as levels of vitamin B12 and folic acid. The computed tomography of the chest, abdomen, and pelvis identified axillar, cervical, periaortic, and inguinal lymphadenopathies. The biopsy of a cervical lymph node was compatible with AITL with immunophenotype CD3+, CD2+, CD7+/−, CD10+, BCL6+, with lymphoid population CD20+ and EER+. An osteomedullary biopsy was performed, which revealed medullary involvement by AITL and showed severe erythroid hypoplasia with a myeloid:erythroid ratio 19.70; the immunohistochemistry of B19 parvovirus was negative. Chemotherapy with CHOP (cyclophosphamide, adriamycin, vincristine, prednisone) was started with dose adjustment for anemia. After 3 cycles of chemotherapy an image of positron emission tomographic (PET) scan was made, showing no hypermetabolic changes. In the end of the treatment (6 cycles) a new PET scan was made showing complete remission.

Discussion/Conclusion

In the case reported the patient presented with severe anemia and evidence of enlarged lymph nodes, evoking a lymphoproliferative disease. The immune anemia Coombs positive with normal white-cell and platelet counts and a low reticulocyte index gave rise to concern about red-cell aplasia. The final diagnosis was made with an immunohistochemistry technique from the biopsy of a peripheric lymphatic node that was compatible with AITL. The osteomedullary biopsy, up with lymphoma infiltration, confirmed severe red-cell hypoplasia.

The positive serology for parvovirus B19 led to the consideration that this agent is a cause of PRCA and yet motivated a therapeutic course with immunoglobulin. However, the DNA quantification and, mainly, the negative result of immunohistochemistry for B19 parvo-

virus in osteomedullary biopsy excluded the involvement of parvovirus B19. It can be presumed that positive parvovirus B19 and CMV serologies are related to cross-reactivity caused by hypergammaglobulinemia.

The mechanisms behind the association with PRCA and lymphoproliferative diseases are not known. It is believed that there is a heterogenous ground that associates humoral factors from the tumor with the inhibitory action in the development of the erythroid lineage. PCRA may precede, occur simultaneously or after the diagnosis of a lymphoproliferative disease [4].

Besides aggressiveness and poor prognosis of AITL associated with PRCA, this patient achieved a complete remission after the treatment and remains asymptomatic and without signs of active disease 1 year after the diagnosis.

Nevertheless, the risk of relapse is high and the long-term prognosis is poor, requiring a close follow-up.

Statement of Ethics

The patient gave written informed consent to publish the case.

Disclosure Statement

The authors have no conflicts of interest to declare.

Funding Sources

The authors received no financial support for this study.

Author Contributions

Marina Vitorino drafted the manuscript and contributed on the interpretation of data. Filipa Nunes, Mariana Costa, and Beatriz Porteiro revised the manuscript and contributed to the interpretation of data. Alexys Reis Borges and João Machado performed the pathology analysis and revised the manuscript, contributed to the interpretation of data, and gave final approval to the version to be published. All authors have read and approved the final manuscript.

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