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Thymectomy and Immunosuppression in Thymoma-Associated Pure Red Cell Aplasia: A Case Report in a Rare and Treatable Paraneoplastic Syndrome

Rajkrishna Sarkar[1*], Ajit Kumar Pegu[2], Kuldeep Deka[3], Anish Hazra[4], Mohd Uveash[5]

[1,3,4,5]Post Graduate Trainee, Department of General Medicine, Assam Medical College and Hospital

[2]Professor Department of General Medicine, Assam Medical College and Hospital

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[*]**Corresponding author:** Rajkrishna Sarkar, Post Graduate Trainee, Department of General Medicine, Assam Medical College and Hospital

ABSTRACT

Pure red cell aplasia (PRCA) is a rare hematological disorder characterized by normocytic normochromic anemia, severe reticulocytopenia, and a marked reduction or absence of erythroid precursors in the bone marrow. Secondary PRCA is often paraneoplastic in origin, with thymoma being a well-recognized but uncommon cause. We report the case of a 70-year-old non-diabetic, non-hypertensive female who presented with fatigue, exertional dyspnea, and palpitations. Clinical examination revealed severe pallor without lymphadenopathy or organomegaly. Laboratory workup showed profound anemia (Hb 3 g/dL), reticulocytopenia (0.2%), and a normocytic normochromic picture on peripheral smear. Bone marrow biopsy confirmed suppression of erythroid precursors, consistent with PRCA. Extensive evaluation for infectious, autoimmune, and other hematologic causes was negative. Contrast-enhanced CT thorax revealed an anterior mediastinal mass suggestive of thymoma. The patient was initiated on immunosuppressive therapy with cyclosporine and responded clinically. She subsequently underwent a successful thymectomy, with follow-up indicating hematologic recovery. This case highlights the paraneoplastic association between thymoma and PRCA, emphasizing the importance of a thorough diagnostic approach in patients presenting with unexplained anemia. Early identification and a combination of immunosuppression and surgical management can lead to favourable outcomes. This report adds to the existing body of literature supporting thymoma-associated PRCA as a rare but treatable condition.

Keywords: Pure red cell aplasia; Thymoma; Paraneoplastic syndrome; Cyclosporine; Thymectomy; Normocytic anemia; Bone marrow suppression

INTRODUCTION

An uncommon cause of anaemia is pure red cell aplasia, which results from the bone marrow's lack of red blood cell precursors. It is frequently a paraneoplastic condition that could have a thymoma as a contributing factor.[1]

A normocytic normochromic anaemia accompanied by significant reticulocytopenia and a notable decrease in erythroid precursors in the bone marrow is referred to as "pure red cell aplasia" (PRCA). A prevalent type of secondary PRCA, observed in 5–13% of these patients, is thymoma-associated PRCA. Less than 10% of patients with thymomas will go on to develop PRCA, while less than 5% of people with PRCA also have thymomas.[2] In most cases, thymectomy is recommended for these patients. While 30% of instances result in an initial remission of the aplasia following surgery, further drugs such as corticosteroids, cyclosporine, or cyclophosphamide may be necessary to maintain the remission.

CASE PRESENTATION

A 70 years old non-hypertensive, non-diabetic female presented to our internal medicine OPD with symptoms of palpitation, generalized weakness, dyspnea on exertion that had been worse over the previous three months. Palpitation is not associated with orthopnea, chest pain, syncope, weight loss, diarrhea, heat intolerance, anxiety, insomnia. Dyspnea is not associated with fever, persistent cough, bipedal edema, paroxysmal nocturnal dyspnea, oliguria, rash, joint pains or deformity. No history of similar illness in her family.[3]

Upon physical examination, severe pallor is present with brittle nails. There is no icterus, cyanosis, clubbing, edema, palpable lymph nodes, engorged neck veins or thyroid swelling her chest is normal and murmur is not heard in cardiovascular system examination. Abdominal examination shows no heapato-splenomegaly. Investigations are planned to rule out hematological or malignant causes of severe anemia.[4]

On laboratory evaluation, Hemoglobin levels of 3g/dl(reference range:12-15 g/dl),0.2% of reticulocytes (reference range:1-2%), a total leucocyte count of 7000/cumm(reference range:4000-11000/cumm), Platelet count: 3.5 lakhs/cumm(reference range:1.5-4.5 lakhs/cumm). PBS revealing normochromic normocytic anemia. Iron profile came to be within normal limit. Ultrasound whole abdomen and 2D echocardiography shows no abnormality. Occult stool and Coombs test turned out to be detrimental. Aspiration and biopsy of Bone Marrow shows suppressed erythroid series with myeloid, lymphoid, megakaryocytic, and plasma cell series are found within the normal range. Infectious and viral panel including Parvo B19, EBV came to be negative. Moreover, ANA Profile is negative. On CECT Thorax, In the anterior mediastinum, a well-defined, hyperdense thymoma with calcification and a central necrotic region is visible in the thorax.[5]

Severe normochromic normocytic anemia, no fever, icterus, edema, hepatosplenomegaly, lymphadenopathy with pure red cell aplasia picture in bone marrow study and anterior mediastinum mass in CT suggests possibility of Thymoma is the cause of Pure red cell Aplasia.[6]

Patient was started on Cyclosporine 100 mg twice daily, which she responds well and discharged. CTVS consultation taken and Thymectomy was done. Patient was on regular follow-up.

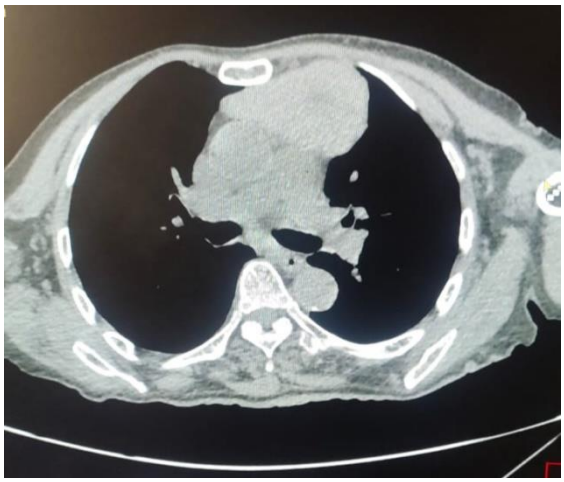

Figure1: Thymoma in CECT Thorax

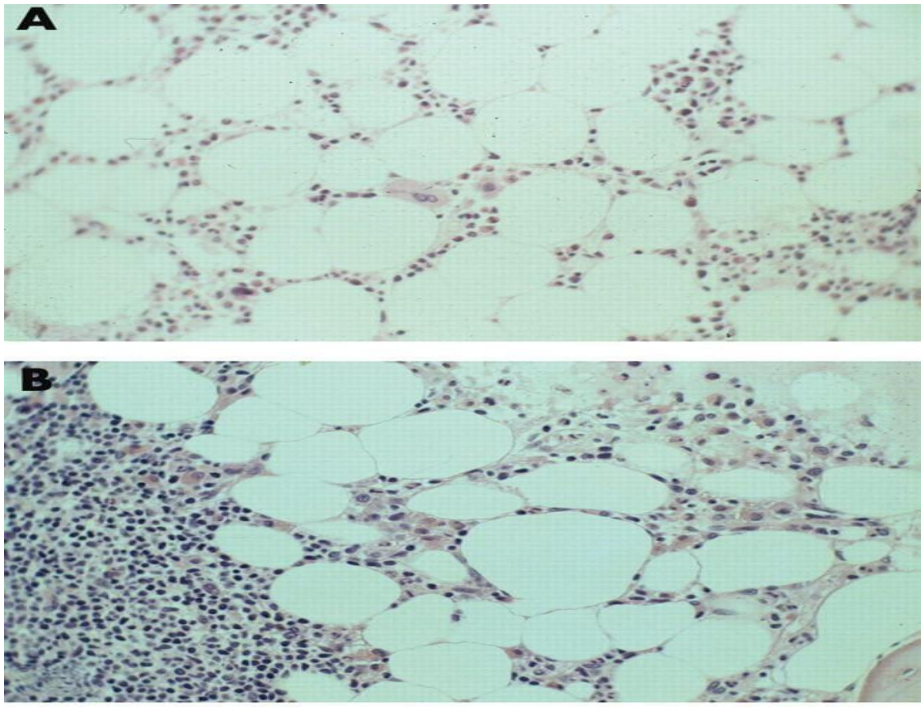

Figure 2: Bone Marrow Biopsy of Pure Red Cell Aplasia

DISCUSSION

Erythropoietic failure with intact megakaryopoiesis and granulopoiesis is a characteristic of PRCA. The pathophysiology of thymoma and PRCA appears to be connected to an autoimmune mechanism where antibodies against erythroblasts prevent erythroid progenitor differentiation; in autoimmune PRCA cases unrelated to autoantibodies, T lymphocytes appear to be the mediating factor in the suppression of erythropoiesis.[7]

PRCA is mostly acquired, though it can occasionally be congenital and linked to medications such as recombinant erythropoietin, Chlorpropamide, immune checkpoint inhibitors (monoclonal antibodies that target CTLA4, PD1, or PDL1), Mycophenolate mofetil, Phenytoin, Cotrimoxazole, Valproate, and Zidovudine. ABO-incompatible hematopoietic stem cell transplantation occurs when hematopoietic stem and progenitor cells from blood group A donors are transplanted into blood group O recipients. Viral diseases include Parvo B19, hepatitis A, HCV, HIV, EBV, and CMV. Immune illnesses include AIHA, SLE, and RA. PRCA linked to lymphoid neoplasms such as non-Hodgkin lymphoma, LGL leukemia, CLL, and Myeloid Neoplasms such as PMF and CML, multiple myeloma.[2,8] Pregnancy-related reports of it typically resolve with delivery. Overall, 5 percent of PRCA patients have a thymoma, while some studies have reported a frequency as high as 22 percent.

Pure Red Cell Aplasia (PRCA) is characterized by severe anemia and symptoms such as fatigue, pallor, shortness of breath, and dizziness. Secondary PRCA may be associated with thymoma, leading to additional symptoms like chest pain or myasthenia gravis. Diagnosis involves CBC, bone marrow biopsy, autoantibody tests, and imaging studies. Treatment includes thymectomy for thymoma-related PRCA, supportive care for parvovirus B19, and immunosuppressive therapy for autoimmune-induced PRCA.[9,10] Blood transfusions may be required for severe anemia. Prognosis varies: thymoma-related PRCA has a good outlook after thymectomy, while autoimmune PRCA may need long-term immunosuppressive therapy.[11,12]

CONCLUSION

This case emphasizes the importance of early recognition and treatment of thymoma-associated PRCA, with thymectomy and immunosuppressive therapy being critical components of management for successful patient outcomes.

DISCLOSURES

CONFLICT OF INTEREST DECLARATION

The writers say they have no conflicts of interest.

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