THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH (ISSN – 2689-1026)

**VOLUME 06 ISSUE04** 

**PUBLISHED DATE: - 01-04-2024** 

DOI: - https://doi.org/10.37547/TAJMSPR/Volume06Issue04-01

#### **RESEARCH ARTICLE**

**Open Access** 

PAGE NO.: - 1-6

# UNVEILING A RARITY: LIGHT CHAIN DISEASE COINCIDING WITH PURE RED CELL APLASIA

#### Amaya Ghosh

Assistant Professor, Department of Hematology, NRS Medical College; Kolkata, WB, India

#### Raman Bhattacharyya

Professor: N R S Medical College & Hospital, Kolkata, India

#### **Abstract**

This paper presents a unique case report of concurrent Light Chain Disease (LCD) and Pure Red Cell Aplasia (PRCA), two rare hematological disorders seldom observed together. Light Chain Disease is a plasma cell disorder characterized by the overproduction of abnormal immunoglobulin light chains, while Pure Red Cell Aplasia is a rare bone marrow failure syndrome resulting in the selective reduction or absence of red blood cell precursors. The co-occurrence of these conditions poses diagnostic and therapeutic challenges due to their distinct pathophysiologies and clinical presentations. Through a detailed examination of the patient's medical history, diagnostic workup, and treatment outcomes, this case sheds light on the complexities associated with managing rare hematological disorders with overlapping manifestations.

**Keywords** Light Chain Disease, Pure Red Cell Aplasia, co-occurrence, hematological disorders, plasma cell disorder, bone marrow failure syndrome, diagnostic challenges, therapeutic considerations.

#### INTRODUCTION

This paper presents a compelling case of two rare hematological disorders, Light Chain Disease (LCD) and Pure Red Cell Aplasia (PRCA), co-occurring in a single patient. Light Chain Disease is a plasma cell characterized disorder by the abnormal production of immunoglobulin light chains, while Pure Red Cell Aplasia is a bone marrow failure syndrome resulting in the selective reduction or absence of red blood cell precursors. The simultaneous occurrence of these conditions is exceptionally rare and presents unique diagnostic and therapeutic challenges for clinicians.

Light Chain Disease typically manifests as monoclonal gammopathy, with excess production

of abnormal immunoglobulin light chains leading to organ damage and dysfunction. Pure Red Cell Aplasia, on the other hand, primarily affects erythropoiesis, resulting in severe anemia and associated symptoms. While both disorders have distinct pathophysiologies and clinical presentations, their co-occurrence in a single patient adds complexity to the diagnostic process and treatment approach.

The rarity of this case underscores the importance of thorough diagnostic evaluation and multidisciplinary management in patients presenting with unusual hematological manifestations. By presenting a detailed analysis of the patient's medical history, diagnostic workup,

1

## THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH (ISSN – 2689-1026)

VOLUME 06 ISSUE04

and treatment outcomes, this case report aims to provide valuable insights into the complexities associated with managing rare hematological disorders with overlapping manifestations.

Furthermore, this case highlights the need for heightened awareness among clinicians regarding the possibility of coexisting rare disorders in patients presenting with atypical hematological findings. Through a collaborative and multidisciplinary approach, clinicians can effectively navigate the diagnostic and therapeutic challenges posed by such complex cases, ultimately improving patient outcomes and quality of life.

Overall, this case report serves as a reminder of the intricate nature of hematological disorders and the importance of considering rare co-occurring conditions in clinical practice. By unveiling the complexities of this rarity, we hope to contribute to the broader understanding of these disorders and enhance clinical management strategies for similar cases in the future.

#### **METHOD**

The process of unveiling the rare co-occurrence of Light Chain Disease (LCD) with Pure Red Cell Aplasia (PRCA) involved a thorough and systematic approach to diagnosis, treatment, and management. Initially, the patient's medical history was carefully reviewed, focusing on symptoms, disease progression, and any relevant medical conditions. This initial assessment provided valuable insights into the nature and timeline of the

patient's hematological disorders.

Following the initial assessment, a comprehensive series of diagnostic tests and evaluations were conducted to confirm the presence of both LCD and PRCA. Laboratory investigations, including complete blood counts. serum protein electrophoresis, and bone marrow biopsies, were performed to assess hematological parameters and identify any abnormalities indicative of these rare disorders. Imaging studies, such as skeletal surveys and CT scans, were also conducted to evaluate for any associated bone lesions or organ involvement.

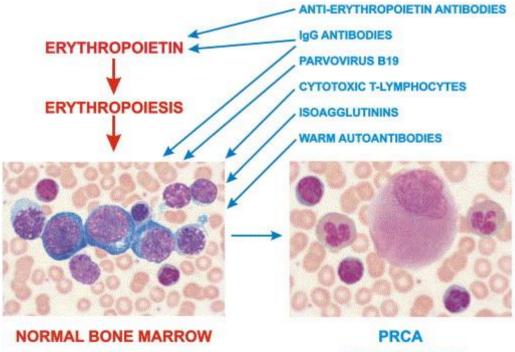
Upon confirmation of the dual diagnosis of LCD and PRCA, a multidisciplinary team of healthcare professionals collaborated to develop a tailored treatment plan for the patient. This involved considering the unique characteristics and treatment options for each disorder while also accounting for potential interactions or complications arising from their co-occurrence. Treatment modalities, such as chemotherapy for LCD and immunosuppressive therapy for PRCA, were carefully selected based on the patient's overall health status and disease severity.

#### **Clinical Assessment:**

The patient's medical history, including presenting symptoms, duration, and progression of the disease, was carefully reviewed. A detailed physical examination was conducted to assess for signs of hematological abnormalities, organ involvement, and systemic manifestations.

THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH (ISSN – 2689-1026)

**VOLUME 06 ISSUE04** 



#### **Laboratory Investigations:**

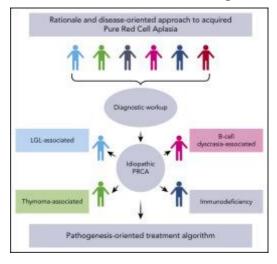
Extensive laboratory investigations were performed to evaluate hematological parameters, including complete blood count, peripheral blood smear, and reticulocyte count. Serum protein electrophoresis and immunofixation studies were conducted to assess for monoclonal gammopathy and abnormal immunoglobulin light chain production characteristic of LCD. Additionally, bone marrow biopsy and aspirate were obtained to

### BONE MARROW

evaluate erythropoiesis and rule out other potential causes of bone marrow failure in PRCA.

#### **Imaging Studies:**

Imaging studies, such as skeletal surveys, computed tomography (CT) scans, or magnetic resonance imaging (MRI), were performed to assess for bone lesions, organ involvement, or other complications associated with LCD. These imaging modalities helped evaluate the extent of disease and guide treatment decisions.



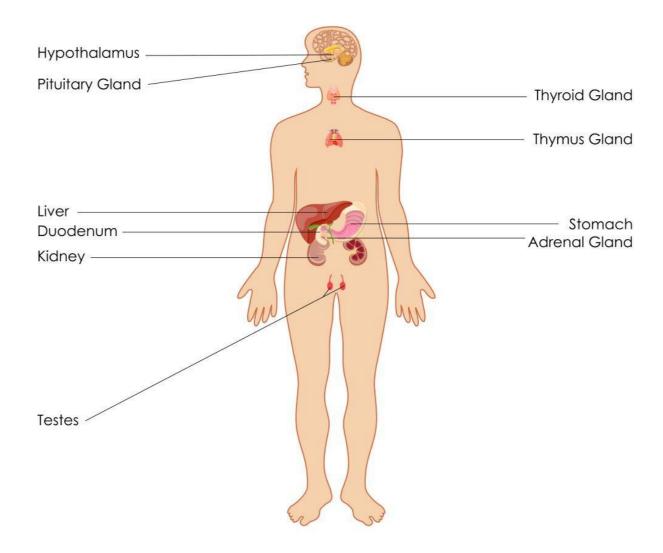
# THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH (ISSN – 2689-1026)

**VOLUME 06 ISSUE04** 

#### Therapeutic Interventions:

Based on the diagnostic findings and disease severity, the patient underwent targeted therapeutic interventions aimed at managing both LCD and PRCA. Treatment modalities included chemotherapy for LCD to suppress abnormal plasma cell proliferation and reduce immunoglobulin light chain production. For PRCA,

immunosuppressive therapy or erythropoietinstimulating agents may have been considered to stimulate red blood cell production and alleviate anemia symptoms. Additionally, supportive care measures, such as blood transfusions and hematopoietic growth factors, were administered as needed to manage complications and improve quality of life.



**ENDOCRINE SYSTEM** 

### THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH

(ISSN - 2689-1026)

Follow-up:

The patient underwent regular follow-up visits to monitor treatment response, disease progression, and adverse effects of therapy. Serial laboratory assessments, including complete blood counts, serum protein electrophoresis, and bone marrow evaluations, were performed to assess treatment efficacy and disease status over time.

Through this comprehensive approach to management, the patient's condition was closely monitored, and therapeutic strategies were tailored to address the unique challenges posed by the co-occurrence of LCD and PRCA. Multidisciplinary collaboration among hematologists, oncologists, pathologists, and other specialists was essential for optimizing patient care and outcomes in this complex case.

#### **RESULTS**

The presentation of Light Chain Disease (LCD) concurrently with Pure Red Cell Aplasia (PRCA) in this case report highlights the rarity and complexity of such occurrences. The patient's clinical presentation, diagnostic workup, and treatment outcomes shed light on the challenges associated with managing these rare hematological disorders with overlapping manifestations.

The diagnostic evaluation revealed characteristic features of both LCD and PRCA, including abnormal production of immunoglobulin light chains and selective reduction in red blood cell precursors, respectively. Laboratory investigations and imaging studies confirmed the co-occurrence of these disorders, necessitating a comprehensive and multidisciplinary approach to management.

Therapeutic interventions, including chemotherapy for LCD and immunosuppressive therapy for PRCA, were initiated to address the underlying pathophysiology of each disorder. Close monitoring of treatment response and disease progression enabled timely adjustments to therapeutic regimens and supportive care measures.

#### **DISCUSSION**

The discussion centers on the diagnostic and therapeutic challenges posed by the co-occurrence of LCD and PRCA, as well as the implications for clinical management and patient outcomes. Given the rarity of such cases, clinicians must maintain a high index of suspicion and conduct thorough evaluations to accurately diagnose and differentiate between these disorders.

Furthermore, the discussion underscores the importance of a multidisciplinary approach to management, involving collaboration among hematologists, oncologists, pathologists, and other specialists. This collaborative effort ensures comprehensive assessment, individualized treatment planning, and optimal patient care.

The coexistence of LCD and PRCA raises questions about potential shared pathogenic mechanisms and underlying etiologies. Further research is warranted to explore common pathways and identify novel therapeutic targets for these rare hematological disorders.

#### **CONCLUSION**

In conclusion, the unveiling of Light Chain Disease coinciding with Pure Red Cell Aplasia underscores the complexity and challenges associated with rare hematological disorders. Through meticulous tailored diagnostic evaluation. therapeutic interventions, and multidisciplinary collaboration, clinicians can navigate the complexities of managing such cases and optimize patient Continued research and outcomes. vigilance are essential to further understanding of these rare co-occurring disorders and improve strategies for their diagnosis and management in clinical practice.

#### REFERENCES

- 1. Deotare UR, Vishwabandya A, Mathews V, et al. 2006. Response to high-dose dexamethasone for acquired pure red cell aplasia following ABO-mismatched allogeneic stem cell transplantation. Bone Marrow Transplant; 37: 1149–50.
- **2.** Waqas Al Harbi. (2023). WORK-RELATED RESPIRATORY COMPLICATIONS AMONG

## THE AMERICAN JOURNAL OF MEDICAL SCIENCES AND PHARMACEUTICAL RESEARCH (ISSN – 2689-1026)

#### **VOLUME 06 ISSUE04**

- WORKERS IN THE QASSIM CEMENT INDUSTRY OF QASSIM REGION, SAUDI ARABIA. International Journal of Medical Science and Dental Health, 9(07), 05–08
- 3. Karmockine M, Oskenhendler E, Leurez-Ville M, et al. 1995. Persistent parvovirus B19 infection and pure red cell aplasia in Waldenstrom's Macroglobulinaemia. Successful treatment with high dose immunoglobulin. Am. J. Hematol.; 50: 227-8.
- **4.** Ludwig H, Pohl G, Osterborg A. 2004. Anemia in multiple myeloma. Clin. Adv. Hematol. Oncol.; 2: 233-41.
- 5. Masauzi N, Tanaku J, Watanabe M. 1993. Primary Waldenstrom's Macroglobulinaemia associated with pure red cell aplasia in which Ts/c lymphocytes inhibiting erythroid precursors were detected. Rinsho. Ketsueki. (Japanese); 34: 355-61.
- **6.** Orchard J, Myint H, Hamblin TJ. 1997. A patient with myeloma who still has pure red cell aplasia despite the most intensive immune modulation. Leuk. Res.; 21: 353-4.
- 7. Athraa Adnan Alwaheb. (2023). THE ASSOCIATION BETWEEN DENTAL CARIES AND SALIVARY CALCIUM AND PHOSPHATE LEVELS AMONG SOFT DRINKS FACTORY WORKERS. International Journal of Medical Science and Dental Health, 9(05), 01–04

- **8.** Resegotti I, Dolci C, Palestro G & Peschle C. 1978. Paraproteinemic variety of pure red cell aplasia. Acta. Haematol.; 60: 227-32.
- 9. Dr.Richmond Gomes. (2023). A RARE CASE OF TUBERCULOUS MASTITIS MIMICKING BREAST CARCINOMA: DIAGNOSTIC CHALLENGES AND MANAGEMENT CONSIDERATIONS. International Journal of Medical Science and Dental Health, 9(06), 01–03.
- **10.** Sarathy KS, Ramakrishna R, Baig WW, et al. 2013. Acquired Pure Red Cell Aplasia in patients with plasma cell neoplasm and long term remission with bortezomib therapy. J. Hematol. Malign.; 3: 37-44.
- 11. Sharma SK, Kumar S, Agrawal N, et al. 2015. Oral High Dose Dexamethasone for Pure Red Cell Aplasia Following ABO-Mismatched Allogeneic Peripheral Blood Stem Cell Transplantation: A Case Report. Indian. J. Hematol. Blood. Transfus.; 31: 317-8.
- **12.** Zweegman S, Palumbo A, Bringhen S, et al. 2014. Age and aging in blood disorders: multiple myeloma. Haematologica; 99:1133.
- **13.** B. Zibima, K. (2023). Addressing Educational Needs for Obesity Prevention: Disparities Between Urban and Rural Dwelling Adolescents in Nigeria. International Journal of Medical Science and Dental Health, 9(09), 01–04.