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Spontaneous Tumor Lysis Syndrome in Myeloproliferative Disorder: A Rarely Reported Clinical Entity

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Abstract Tumor lysis syndrome (TLS) is an oncologic emergency resulting from a massive breakdown of tumor cells. It causes electrolyte imbalance and acute renal failure. It occurs after the initiation of chemotherapy. Spontaneous tumor lysis syndrome is seen in Burkitt's lymphoma, acute lymphoid leukemia, diffuse large B-cell lymphoma, and solid tumors like breast and prostate cancer. Spontaneous tumor lysis syndrome is exceedingly rare in patients with myeloproliferative disorders. We report a rare case of spontaneous tumor lysis in an 82-year-old female patient with a myeloproliferative disorder.

Keywords: *tumor lysis, myeloproliferative, spontaneous*

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1. Case Presentation

An 82-year-old female had leukocytosis (white blood cell counts 179000/microL) on routine lab work up ordered by primary care physician. As the patient had no symptoms, she declined further evaluation and treatment. After 6 months, she presented with complaints of generalized weakness, erythema, and pain in the right leg for 3 days. Laboratory works up showed white cell count (WBC) >250000/microL with differentials of neutrophils 36% lymphocytes 2%, band 21%, myelocytes 14%, and metamyelocytes 15%. Patient had blood urea nitrogen 125 mg/dl, creatinine 3.3mg/dl, potassium 6.6 mmol/L, phosphorus 10.2 mmol/L, uric acid 16.6 mg/dl and lactate dehydrogenase level of 1668 U/L. During initial treatment in the emergency department, the patient developed torsade's de points and, subsequently, cardiac arrest from ventricular fibrillation. The patient had a return of spontaneous circulation (ROSC) after defibrillation. The patient was intubated and transferred to the intensive care unit. The patient received single-dose rasburicase and started on hydroxyurea for cytoreduction. The patient became anuric and started on continuous renal replacement therapy. Electrolyte abnormalities improved with continuous renal replacement therapy. Peripheral blood smear showed neutrophilia with left-sided shift and circulating blast. Due to the patient's critical condition, bone marrow biopsy was postponed. BCR-ABL and JAK2V617F tests were sent in consultation with hematologist/ oncologist. BCR-ABL turned out to be positive, and JAK2 V617F was negative. The patient developed inferior wall ST-segment elevation myocardial infarction while being on Ventilator and renal replacement therapy. The family opted for comfort measures only.

2. Discussion

Chronic myeloid leukemia is one of the myeloproliferative disorders. It accounts for 15-20% of all leukemia's in adults. It is mainly diagnosed with a Philadelphia chromosome or one of its products. [1] Tumor lysis syndrome is a metabolic complication that occurs after starting the patient on chemotherapy or spontaneously. It is mainly characterized by hyperuricemia, hyperkalemia, hyperphosphatemia, and hypocalcemia. These electrolyte abnormalities can lead to acute renal failure, seizure, cardiac arrhythmias, and death. [2] According to Cairo and Bishop's definition of tumor lysis syndrome, diagnosis can be clinical or laboratory. Laboratory tumor lysis syndrome is defined as the presence of severe electrolyte abnormalities without clinical manifestation. Such patients should be treated. Clinical tumor lysis syndrome is defined as biochemical changes associated with severe clinical manifestation and needs aggressive treatment. [3]

According to Cario and Bishop's laboratory criteria of tumor lysis syndrome, two or more of the following abnormalities should be present either 3 days before or 7 days after starting on chemotherapy: Uric acid >476 mmol/L, potassium>6 mmol/L, phosphorus > 2.1 mmol/L, calcium<1.75 mmol/L or 25 % increase in uric acid, potassium or phosphorus and 25% decrease in calcium level from baseline. Clinically it can be defined as

one or more of the following abnormalities: cardiac arrhythmias, sudden death or seizure, or increase in creatinine >1.5 from baseline. [2]

Later on, Cario and Bishop's criteria were modified by omitting a 25 % change from baseline in 2011 by Howard et al. According to Howard et al., and colleagues 25% change from baseline in not important clinically. So tumor lysis syndrome was redefined as the presence of two or more following labs—abnormalities during the same 24 hours period within 3 days before or 7 days after starting chemotherapy. On the other hand, lab abnormalities plus clinical manifestations, including death, increase in creatinine, cardiac arrhythmias, and seizure constitutes tumor lysis syndrome. [4]

Treatment of tumor lysis syndrome includes correction of specific electrolyte abnormalities, acute renal failure, and use of allopurinol vs rasburicase to decrease the conversion of nucleic acid to uric acid. [5] Decrease production of uric acid is helpful against the development of oliguric renal failure. The use of rasburicase is associated with decrease stay in the intensive care unit. [6]

3. Conclusion

We describe a case of spontaneous tumor lysis syndrome in a patient with chronic myeloid leukemia a rarely reported entity with this diagnosis. Clinicians should consider this diagnosis while dealing with the myeloproliferative disorder as this complication is preventable.

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