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Approach to an Unusual Case of Refractory Immune Thrombocytopenic Purpura with a Zero Platelet Count Level

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The average range in an adult's platelet count is 150,000 to 450,000/microL, with a mean value of 266,000 in females and 237,000/microL in males. When platelet count levels are below the normal limit it is termed thrombocytopenia. Thrombocytopenia can be manifested in a variety of diseases, which makes diagnosing a patient's underlying condition challenging.

We present for your consideration the case of a 69 year old woman with a history of HTN, DM, colon adenocarcinoma, and tongue squamous cell carcinoma, who presented an incidental finding of low platelet count during her initial evaluation by her primary care physician in June 2020. Patient had extensive bleeding after a lip bite, and petechial rash development. Outpatient CBC revealed 0 platelet count level. Two days later, the patient was transferred from another hospital facility to our ER with a platelet count value of 5. She was admitted with a diagnosis of primary thrombocytopenia. On the 5th day, management with methylprednisolone IV and platelet transfusion was started, and laboratory tests were ordered. The next day, laboratory test results showed refractory thrombocytopenia and leukocytosis of 21.26. Aggressive treatment with IVIG was started for suspected immune thrombocytopenic purpura (ITP), to be confirmed by bone marrow (BM) biopsy. BM biopsy revealed Chronic lymphocytic leukemia (CLL) with normal megakaryocytes consistent with concomitant ITP, a combination seen in only 1–5% cases of CLL patients. Management was planned to continue therapy for ITP in hopes of controlling the disease without the use of chemotherapy. However, steroid-refractory ITP is an indication for induction chemotherapy for CLL. Rituximab was added to the treatment regimen, and results showed a progressive increase in platelet count in the following weeks until the patient was discharged home with oral Prednisone.

This case presents a rare relationship between two conditions that require immediate action, where suspecting and managing one of the conditions may lead to the diagnosis of the other. CLL is the most common adult leukemia in Western populations, comprising 25% to 30% of all leukemias in the United States. It is characterized by monoclonal B cell proliferation. Patients with B-chronic lymphocytic leukemia/small lymphocytic lymphoma have a 5–10% risk of developing autoimmune complications that result in cytopenia. The relationship between CLL and ITP is rare. In CLL patients the reported risk of ITP ranges from 1–5%; being the highest proportion in patients whose bone marrow studies are routinely used to determine the etiology of thrombocytopenia. Therefore, early detection of this co-diagnosis allows prompt delivery of appropriate management, and prevention of refractory disease or further disease progression.