Update in the primary plasma cell leukemia

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Plasma cell leukemia (PCL) is a rare and highly aggressive plasma cell neoplasm developing in 0.5-4% of patients with multiple myeloma (MM). The diagnostic criterion was recently revised from 20% to ≥5% circulating plasma cells in peripheral blood smears. PCL is classified as primary or secondary; primary PCL is that of patients lacking prior MM. Primary PCL shows clinical and laboratory features at presentation that differ from those of MM and exhibits a dismal prognosis even with the use of agents that are effective against MM. Intensive chemotherapy should commence immediately after diagnosis, and autologous stem cell transplantation is recommended for transplant-eligible patients. Maintenance therapy after transplantation may reduce early relapse. We review the definitions of PCL, the revised diagnostic criteria, the clinical features, and appropriate initial treatments for primary PCL.