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Perspective

Characteristics and Prognosis of Plasma Cell Leukemia

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DISCRIPTION

Plasma Cell Leukemia (PCL) is a plasma cell dyscrasia, or a disease characterized by the malignant degeneration of plasma cells, a subtype of white blood cells. It is the most advanced and aggressive form of these dyscrasias, accounting for 2% to 4% of all cases of plasma cell malignancies. PCL may show either as primary plasma cell leukemia (in patients with no prior history of a plasma cell dyscrasia) or secondary plasma cell dyscrasia (in patients with a history of its predecessor dyscrasia, multiple myeloma). The two PCL forms appear to be at least partially distinct from each other. In all cases, however, PCL is a severe, life-threatening, and therapeutically challenging disease.

The clinical presentation of primary PCL (pPCL) reveals a significantly more aggressive disease than that of a typical multiple myeloma case, with clinical features that are a combination of those found in multiple myeloma and acute leukemia. In 28%-56%, 4%-7%, 23%-44%, or 0%-12% of cases, pPCL patients exhibit pathologically high levels of monoclonal plasma cells in their bone marrow, as well as a malignant plasma cell-secreted circulating monoclonal myeloma protein, either IgG, IgA, a light chain, or none. pPCL patients exhibit splenomegaly, lymphadenopathy, hepatomegaly, kidney failure, bone marrow failure (thrombocytopenia, anemia, and/or, rarely, leukopenia), similar to B cell leukemia but not multiple myeloma.

Defects in the central nervous system and peripheral neuropathies caused by plasma cell invasion and/or deposition of their circulating monoclonal immunoglobulin in these tissues. pPCL patients have higher rates of developing a hypercalcemic crisis, i.e. a potentially life-threatening episode of high ionic calcium (Ca^{2+}) levels in the blood due to excess bone re-absorption and/or renal failure, than multiple myeloma patients; b) higher levels of serum lactate dehydrogenase and Beta-2 microglobulin; and c) lower rates of bone but higher rates of soft tissue plasma cell tumors.

Secondary PCL (sPCL) is diagnosed in 1%-4% of patients with multiple myeloma over a median of 21 months. It is the final phase of these patients' blood cancer disease. Patients with sPCL are typically highly symptomatic due to extensive disease defined by malignant plasma cell infiltrations and failures of not only the bone marrow but also other organs. They have failed or discontinued one or more treatment regimens and may show some of the toxic effects of these treatments. PCL is caused by an abnormally high number of genetic abnormalities in plasma cells or, more particularly, their precursor B cells and plasma blasts.

This genetic instability is caused by a variety of acquired abnormalities, including gene mutations; single nucleotide polymorphisms; depletions and duplications of parts of a gene, larger portions of a chromosome, or even an entire arm of a chromosome; translocations, deletions, and duplications of entire chromosomes; and increases and decreases in the expression of intact genes due to, for example, methylation of gene promoters and These gene mutations have an effect on the Wnt signaling system, cell cycle regulation, RNA metabolism, protein folding, and cadherin-related cell adherence to extracellular matrix. These effects, in turn, control plasma cell proliferation, survival, apoptosis, bone marrow adhesion, genome stability, and monoclonal immunoglobulin secretion.

Secondary Plasma Cell Leukemia (sPCL) is caused by the comparatively slow development of plasma cell/plasma cell precursor genetic defects, which initially produce a clone of cells that produces the premalignant condition of monoclonal gammopathy of unknown etiology. In a very tiny percentage of these cases, the development of additional genetic abnormalities creates a clone(s) of plasma cells, leading in the more serious but still premalignant disorder of smouldering multiple myeloma, overt myeloma cancer, and, ultimately, sPCL. In contrast to sPCL, pPCL starts from start with a wide range of genetic abnormalities. For example, at the time of diagnosis, advanced methods for examining the genome, such as whole-exome sequencing and gene expression profiling, identified 166 non-silent gene variants per pPCL patient sample.

These abnormalities are similar but not identical to those detected in sPCL, whereas those detected in sPCL more closely resemble those detected in multiple myeloma than those detected in pPCL: the genetic data support the clinical data in suggesting that sPCL and pPCL are distinct diseases, with sPCL being more closely related to multiple myeloma. Examination of plasma cell

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immunophenotype by measuring certain cell surface antigens, particularly Cluster of differentiation. CD markers on plasma cells from patients with pPCL differ from those observed in people who have multiple myeloma or sPCL. For example, pPCL plasma is more likely to express CD20 antigen, which is believed to be important in anchoring plasma cells to the bone marrow stroma.

The International Myeloma Working Group defines plasma cell leukemia as the presence of more than 2×10^9 plasma cells per liter of blood or, alternatively, more than 20% of nucleated blood cells being plasma cells. Recently, the Group has suggested that values of 0.5×10^9 or 5%, respectively, may be more appropriate from a therapeutic viewpoint and, as being such, should be studied as a definitive criterion for the disease. A recent study discovered that multiple myeloma patients with >5% circulating plasma cells had a much worse prognosis than those with multiple myeloma and similar to those with plasma cell leukemia.

Flow cytometry immunophenotyping of blood cells to detect clonal phenotypes of plasma cells seen in multiple myeloma (for example, the $CD138^+$, $CD38^+$, $CD19^{+/-}$ phenotype) may be a more sensitive way to count circulating clonal plasma cells and diagnose plasma cell leukemia. Prior to the use of newly developed drugs and treatment regimens, the median survival rates for pPCL and sPCL from the time of diagnosis were 8-11 months and 2-8 months, respectively, even when treated very aggressively with the VAD regimen of vincristine, doxorubicin, and dexamethasone or the VCMP regimen of vincristine, carmustine, melphalan, and prednisone alternating with vincristine, carmustine, The use of newer methods for treating PCL patients, particularly pPCL patients, appears to have led in modest improvements in survival rates.