

Information on PLASMA CELL LEUKEMIA

What is Plasma Cell Leukemia?

Plasma cell leukemia (PCL) is an aggressive form of multiple myeloma characterized by high levels of abnormal plasma cells circulating in the peripheral (circulating) blood. Normal plasma cells in the bone marrow produce antibodies that fight infection. In myeloma most of the abnormal plasma cells remain in the bone marrow and are not found in the peripheral blood. In PCL, the abnormal plasma cells are in the peripheral blood. Therefore, PCL is considered to be an advanced form of myeloma. PCL can either originate as the primary manifestation of the disease (primary PCL with no history of myeloma) or as a transformation of myeloma (secondary PCL with progression of previously diagnosed myeloma).

Key Statistics and Risk Factors

Primary PCL is rare, with an estimated 1 per million of the general population diagnosed each year. Secondary PCL occurs in one to four out of 100 cases of myeloma and is becoming more common as myeloma patients are living longer.

As with myeloma, PCL is more common in African Americans than in Caucasians and is slightly more common in men than in women. As new insights and knowledge about the biology of myeloma and PCL are gained, it may be possible to determine which myeloma patients are at increased risk for developing PCL.

The causes of PCL are similar to those of myeloma. A series of genetic alterations during the development of a plasma cell may lead to the cell's uncontrolled growth. However, what triggers these alterations is not fully known. Risk factors, such as age and exposure to industrial and environmental elements, are thought to play important roles.

Signs and Symptoms

While PCL patients often have symptoms and complications similar to those of myeloma patients, they tend to be more severe. Symptoms and complications include:

- Bone pain
- Fatigue
- Recurring infections
- Bleeding
- High levels of calcium (hypercalcemia)
- Kidney damage
- Enlarged liver or spleen resulting from large numbers of abnormal plasma cells deposited in these organs.

Diagnosing PCL

PCL is diagnosed by the number of abnormal plasma cells circulating in the blood. In PCL patients, abnormal plasma cells make up more than 20% of the total number of white blood cells present in the peripheral blood. PCL is diagnosed in the same way as myeloma.

Treatment

Current treatments for primary PCL are the same as those for myeloma. Most secondary PCL patients have already had several anti-myeloma treatments, and for some patients their disease may have become resistant, or refractory, to the treatments. For these patients, more intensive treatments using combinations of chemotherapy drugs, steroids and new agents may be considered. Treatments may include:

- Chemotherapy agents, such as cisplatin, adriamycin, and cyclophosphamide
- Proteasome inhibitors, such as Velcade® (bortezomib)
- Immunomodulatory agents, such as Revlimid® (lenalidomide) and thalidomide
- High-dose therapy and stem cell transplantation for younger and/or healthier patients
- Newer agents, such as Kyprolis® (carfilzomib) and Imnovid® (pomalidomide), for those whose disease is refractory to Velcade and Revlimid
- Other novel agents

Prognosis

Because of the aggressive nature of the PCL, it is usually associated with a poor prognosis and a survival expectancy that is shorter than that of patients with typical myeloma.

Future

Scientists at the Myeloma Institute are investigating the characteristics of and risk factors for PCL. Understanding the biological and genetic features that enable development of primary PCL and progression from myeloma to secondary PCL will lead to improved therapies for better patient outcomes.

Patients are encouraged to consult with their physicians about the most current therapies available.

This information about PCL and other patient education materials are available at www.myeloma.uams.edu