UAMS myeloma institute

Information on POEMS SYNDROME

What is POEMS Syndrome?

POEMS syndrome is a rare, multi-system blood disorder.

The acronym "POEMS" stands for:

P – Polyneuropathy
O – Organomegaly
E – Endocrinopathy
M – Monoclonal protein
S – Skin changes

A diagnosis of POEMS is determined by the presence of a monoclonal plasma cell disorder, peripheral neuropathy, and one or more of the following: osteosclerotic bone lesions, organomegaly, endocrinopathy, skin changes, increased levels of vascular endothelial growth factor, and swelling.

Key Statistics

POEMS syndrome is extremely rare. It affects more men than women, and typically occurs in one's 50's. The exact incidence is unknown.

Causes

The cause of POEMS syndrome is not known. However, chronic overproduction of proinflammatory and other cytokines (proteins made by the immune system that act as chemical messengers) seems to be a main feature of the syndrome. Individuals with POEMS syndrome have an increased number of plasma cells.

Symptoms

Polyneuropathy is characterized by chronic, progressive disease affecting the peripheral nervous system. The peripheral nervous system consists of the motor and sensory nerves that connect the brain and spinal cord (central nervous system) to the rest of the body. Individuals with POEMS syndrome experience numbness, tingling, feelings of coldness, and weakness, starting in the toes and feet and progressively working its way upwards. Over time, the hands can be affected.

Organomegaly refers to abnormal enlargement of the spleen, liver and lymph nodes. Swelling, especially of the lymph nodes, may be present. Biopsy of enlarged lymph nodes may reveal Castleman Disease (a rare disease of lymph nodes and related tissues that results from the overgrowth of the cells in the body's lymphatic system).

Endocrinopathy refers to abnormalities in the endocrine system (the network of glands that secrete hormones into the circulatory system). Abnormal hormone levels can lead to hypothyroidism (underactive thyroid), metabolism problems, improper functioning of the ovaries in females or testes in males, and diabetes.

Monoclonal protein (M-protein), caused by a monoclonal plasma cell disorder, can be detected in the blood serum and/or urine. Monoclonal plasma cell disorders are characterized by uncontrolled growth of abnormal plasma cells. The overproduction of plasma cells in individuals with POEMS syndrome can result in the formation of tumors known as plasmacytomas; these tumors appear like sclerotic lesions (thickening of the bone) on x-ray. Overproduction of plasma cells can also result in the formation of osteosclerotic lesions in bone.

Skin abnormalities in POEMS syndrome can include abnormal darkening of the skin, hardening and thickening of the skin (sclerosis), increased facial or leg hair, and excessive sweating.

Additional symptoms may include fluid buildup in the lungs, chronic renal failure, fluid accumulation in the skin of the arms and legs, and cardiovascular disease.

Diagnosis

A diagnosis of POEMS syndrome begins with a complete medical history and physical exam.

Laboratory tests of blood and urine are conducted to check protein and hormone levels. Often vascular endothelial growth factor (VEGF) levels are elevated.

Additional diagnostic testing may include:

- **Skeletal imaging tests, such as CT (computerized tomography) and PET (positron emission tomography) scans, to check for osteosclerotic lesions and enlargement of lymph nodes, liver and spleen**
- **Biopsy of an osteosclerotic lesion and/or bone marrow biopsy to check for abnormal plasma cells**
- **Lymph node biopsy to diagnose co-existent Castleman Disease**
- **Pulmonary function testing**
- **Echocardiogram (ultrasound of the heart)**
- **Endocrine evaluation**
- **Nerve conduction studies to evaluate the neuropathy**

Treatment

Treatment is aimed at the underlying plasma cell disorder and relieving symptoms. Every patient's disease is different. Myeloma Institute physicians are dedicated to defining the nuances of each individual's disease and customizing treatment regimens that have the most promise for effective results.

Autologous Stem Cell Transplantation (high-dose chemotherapy with stem cell rescue) is the treatment of choice for transplant eligible patients and often results in dramatic improvement.

Treatments that are similar to those for myeloma are utilized for non-transplant eligible patients and include:

- **Steroids,** such as dexamethasone, to trigger destruction of abnormal plasma cells
- **Thalidomide or Revlimid® (lenalidomide),** which are immunomodulatory drugs that act on cells involved in the body's immune system
- **Velcade® (bortezomib) or Kyprolis® (carfilzomib),** a proteasome inhibitor that causes abnormal cells to die (Proteosomes are protein complexes in cells that degrade unneeded or damaged proteins.)
- **Radiation therapy** to destroy localized bone lesions
- **Monoclonal antibodies,** such as rituximab, for patients with significant organomegaly

Antibodies which target interleukin 6, e.g., tocilizumab and siltuximab, are often not effective in the treatment of POEMS syndrome. Plasma exchange and intravenous immunoglobulin treatment do not result in a lasting improvement of the neuropathy.

Patients with POEMS syndrome often need treatment to alleviate the symptoms of neuropathy, extravascular volume overload, and endocrine abnormalities.

- **Neuropathy** - Physical therapy to strengthen legs and appropriate use of ankle/foot orthotics and walking aids can improve lifestyle.

- **Extravascular volume overload** – Diuretics may be used to decrease the amount of excess fluid and swelling

- **Endocrine abnormalities** – Hormone replacement may be appropriate for the treatment of hypothyroidism (underactive thyroid) and other hormone-related symptoms.

The effectiveness of treatment is monitored on a regular basis.

Prognosis

POEMS syndrome can be effectively treated and significant improvement in overall functioning and quality of life can be expected. Autologous Stem Cell Transplantation (high-dose chemotherapy with stem cell rescue), the treatment of choice for transplant eligible patients, often results in dramatic improvement.

A multi-disciplinary approach to the treatment of POEMS syndrome is essential. The physician team should include myeloma specialists, radiologists, neurologists, nephrologists, cardiologists and other specialists as needed.

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