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Review Article

POEMS Syndrome- A Review

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Abstract

This review's objective is to provide you with the most recent data about POEMS syndrome. The authors reviewed, compared, and assessed key findings after conducting a literature search of the accessible sources explaining POEMS syndrome. According to the results of this study, POEMS syndrome is related to a class of diseases called monoclonal gammopathies or plasma cell dyscrasias. The unchecked growth of a single clone (monoclonal) of plasma cells causes an abnormal buildup of M-proteins (also known as immunoglobulin) in the blood, which has a detrimental effect on the quality of life for both the patients and their family. To minimize or even eliminate the symptoms, early and appropriate diagnosis and therapy/Treatment are necessary.

Keywords: POEMS syndrome, Immunoglobulin Plasma dyscrasias, Monoclonal.

INTRODUCTION

Monoclonal gammopathies, also called plasma cell dyscrasias, are connected to POEMS syndrome. The unchecked proliferation of a single clone (monoclonal) of plasma cells leads to the abnormal buildup of M-proteins, commonly known as immunoglobulins, in the blood. A protein in the body called immunoglobulin aids in the defense against infection. Both the precise cause of POEMS syndrome and the precise involvement of M-proteins in it are unknown. According to study, a chemical known as VEGF (vascular endothelial growth factor) may be involved with this illness. The illness was given the name Crow Fukase Syndrome since it was initially characterized by Crow in 1956 and then by Fukase in 1968. The conditions known as PEP syndrome, Takatsuki disease, Crow-Fukase syndrome, and osteosclerotic myeloma are all the same thing.

What is POEMS Syndrome?

POEMS syndrome is a multisystem condition that affects a small number of people. Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy, and Skin Changes are all acronyms for polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes.

Multisystem disease called POEMS syndrome is extremely rare. The acronym POEMS stands for polyneuropathy (disease of many nerves), organomegaly (abnormal enlargement of an organ), endocrinology (disease of some hormone-producing glands that help regulate metabolic function and sexual function), monoclonal gammopathy or M protein (skin abnormalities), and skin abnormalities.

Etiology

- The cause of POEMS syndrome is uncertain.
- Developmental, environmental, and genetic factors all play a part.
- Race: Although numerous instances have been noted in Japanese literature, no conclusive racial connection has been discovered.
- Sex- With a male-to-female ratio of 2.5:1, men are marginally more likely than women to have it.
- Patient age—The onset of POEMS syndrome often occurs in the fifth or sixth decade of life, with a mean patient age of 48 years for men and 59 years for women.

Pathophysiology

The pathophysiology of POEMS syndrome is poorly understood. Unknown mechanisms underlie the malfunction of plasma cells, which is what leads to the condition's onset. Tumor necrosis factor (TNF)-alpha,

interleukin (IL)-1beta, and IL-6 levels have all been reported to be higher. Vascular endothelial growth factor (VEGF) levels have recently been seen to significantly rise. It is believed that elevated VEGF levels cause increased vascular permeability, which in turn causes edema, elevated endo neural pressure, and the deposition of material produced from plasma cells. When myelin is exposed to serum cytokines and complement, demyelization can happen.

Clinical Manifestation

1. Polyneuropathy - Breathing problems and numbness, tingling, and weakness in your legs, potentially over time in your hands as well. This is a vital factor in figuring out if someone has POEMS syndrome or not.
2. Organomegaly - characterized by enlargement of the lymph nodes, liver, or spleen.
3. Endocrinopathy/edema - Symptoms of endocrinopathy/edema include an underactive thyroid (hypothyroidism), diabetes, erectile dysfunction, fatigue, swelling in your limbs, and trouble with metabolism and other critical functions.
4. Monoclonal-protein-producing abnormal bone marrow cells (plasma cells) that create a protein (monoclonal protein) that is present in the bloodstream. This is a crucial element in determining whether or not someone has POEMS syndrome. Unusual bone stiffening or thickening is frequently linked to monoclonal protein.

Other symptoms and signs

1. Ascites, peripheral oedema, and pleural effusion are some of the other symptoms and markers of extravascular fluid excess (accumulation of fluid in the abdominal cavity, limbs and lung lining respectively)
2. Thromboses in the venous and arterial systems (blood clots)Examples of pulmonary diseases include
3. Restrictive lung disease,
4. Pulmonary hypertension, and
5. Weakening of the respiratory muscles.

Diagnostic Evaluation: To diagnose POEMS, need:

- Both mandatory major criteria,
- At least 1 of the other major criteria,
- AND at least 1 of the minor criteria.

Mandatory criteria	Major criteria	Minor criteria
• Peripheral neuropathy • Monoclonal plasma cell disorder	• Osteosclerotic bone lesions • Castleman disease (giant cell or Angio follicular lymph node hyperplasia) • Increased vascular endothelial growth factor (VEGF)	• Endocrinopathy (excluding diabetes or hypothyroidism) • Organomegaly • Extravascular volume overload (most commonly peripheral oedema; also pleural effusion, ascites) • Thrombocytosis/polycythaemia – arterial and venous thrombosis, strokes • Skin changes • Papilloedema

Additional diagnostic procedures include

1. Blood testing.
2. A full blood count, which could show polycythemia and/or thrombocytosis.
3. Endocrine panels may show abnormalities in thyrotropin, hyperglycemia, and oestrogen.
4. Serum or urine electrophoresis to detect monoclonal immunoglobulin
5. Osteosclerotic lesions, which are frequently many, are seen on X-rays of the bones in practically every patient.
6. Biopsy of an enlarged lymph node may show Castleman disease.
7. Those who have neuropathy may have higher levels of protein in their cerebrospinal fluid after lumbar puncture.
8. Studies on nerve conduction may reveal axonal degeneration and demyelination alterations.
9. A bone marrow examination may reveal plasma cell involvement.
10. A biopsy of swollen lymph nodes.
11. Skin biopsy results are usually nonspecific-

- Nonspecific hyperpigmentation of the basal layer with inflammatory infiltration or dermal fibrosis is seen in Scleroderma-like lesions. It differs from scleroderma because sweat glands and collagen are normal.
- Inflammatory infiltration of lymphocytes and plasma cells is seen in hyperpigmented lesions.
- Strawberry naevus, lobular capillary angioma, and glomeruloid haemangioma are examples of angiomas (this is especially characteristic of POEMS syndrome and shows enlarged vascular spaces filled with coiled capillaries surrounded by pericytes, which look like kidney glomerulus).

Management

The underlying plasma cell malfunction is addressed in the same manner as POEMS syndrome. Most patients are treated with a combination of medicine, surgery, and adjuvant therapy. Patients with diffuse sickness are currently treated with corticosteroids, low-dose alkylators, and peripheral blood stem cell transplantation following high-dose chemotherapy.

Prognosis

POEMS Syndrome is a chronic condition. The median survival time for patients with multiple myeloma is eight to fourteen years, which is three times longer. However, many people are restricted to their beds as a result of neuropathy (50 percent).

CONCLUSION

POEMS Syndrome is a rare clinical illness caused by plasma dyscariasis and manifests as polyneuropathy, organomegaly, endocrinopathy monogammppathy, and skin abnormalities. Although there is no recognized cause, this condition has a long family history. Since every patient differs, the person or family should consult a doctor to create a treatment strategy that is appropriate for them.

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