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# Poems syndrome: A review

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**Abstract**
The purpose of this review is to provide the latest information on POEMS syndrome. The authors conducted a literature search of available sources describing the issue of POEMS syndrome with special focus on syndrome and made a comparison and evaluation of relevant findings. The results of this review indicate that POEMS syndrome is associated with a group of disorders known as monoclonal gammopathies or plasma cell dyscrasias. These disorders are characterized the uncontrolled growth of a single clone (monoclonal) of plasma cells, which results in the abnormal accumulation of M-proteins (also known as immunoglobulin) in the blood which has a significant impact on the quality of life of both the patients and his/her family. Therefore, early and proper diagnosis and treatment are necessary in order to reduce or even eliminate both symptoms and social burden of the patient.

**Keywords:** POEMS syndrome, plasma dyscrasias, immunoglobulin

## Introduction
POEMS syndrome is associated with a group of disorders known as monoclonal gammopathies or plasma cell dyscrasias. These disorders are characterized the uncontrolled growth of a single clone (monoclonal) of plasma cells, which results in the abnormal accumulation of M-proteins (also known as immunoglobulins) in the blood. Immunoglobulin in health fight infection. However, the specific role M-proteins play and the exact cause of POEMS syndrome is unknown. Research would suggest that a chemical called VEGF (vascular endothelial growth factor) plays an important role in this disease. The syndrome was also termed as crow Fukase Sydrome as it was described by Crow in 1956 and then Fukase in 1968. POEMS syndrome (also termed osteosclerotic myeloma, Crow–Fukase syndrome, Takatsuki disease, or PEP syndrome)

## What is POEMS Syndrome?
POEMS syndrome is a rare multisystem disorder. POEMS stands for: Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal gammopathy and Skin changes.
POEMS syndrome is an extremely rare multisystem disorder. POEMS is an acronym that stands for polyneuropathy, disease affecting many nerves; organomegaly, abnormal enlargement of an organ; Endocrinology, disease affecting certain hormone –producing gland that help to regulate sexual function, and metabolic function, Monoclonal gammopathy or M protein; and skin abnormalities.
POEMS syndrome is paraneoplastic syndrome caused by plasma cell dyscrasia related to changes in the level of a cytokine or a growth factor.

## Etiology
- The cause of POEMS syndrome is Unknown
- Genetics
- Environmental and developmental factors
- Race-no specific racial association has been identified, although a preponderance of cases have been reported in the Japanese literature.
- Sex- Slightly more prevalent among men than women, with a male-to-female ratio of 2.5:1.
- Age-The onset of POEMS syndrome occurs most frequently in the fifth or sixth decade of life, with a mean patient age at onset of 48 years for men and 59 years for women.

**Pathophysiology**

The pathophysiology of POEMS syndrome is not clearly understood. A a plasma cell disorder underlies the development of the syndrome; however, the mechanism by which this occurs is unknown. Elevations of cytokines, such as interleukin (IL)–1beta, IL-6, and tumor necrosis factor (TNF)–alpha, have all been noted. Most recently, significant elevations in vascular endothelial growth factor (VEGF) levels have been noted. Increases in VEGF levels have been postulated to lead to enhanced vascular permeability, leading to the associated edema, increased endo neural pressure, and deposition of plasma cell–derived material. As myelin is exposed to serum cytokines and complement, demyelization can occur.

**Clinical Manifestation**

1. **Polyneuropathy:** Numbness, tingling and weakness in your legs — and over time, maybe in your hands — and difficulty breathing. This is an essential feature in the diagnosis of POEMS syndrome.
2. **Organomegaly:** Enlarged spleen, liver or lymph nodes.
3. **Endocrinopathy/edema:** Abnormal hormone levels that can result in an underactive thyroid (hypothyroidism), diabetes, sexual problems, fatigue, swelling in your limbs, and problems with metabolism and other essential functions.
4. **Monoclonal-protein:** Abnormal bone marrow cells (plasma cells) that produce a protein (monoclonal protein) that can be found in the bloodstream. This is an essential feature in the diagnosis of POEMS syndrome. Monoclonal-protein is often associated with unusual bone hardening or thickening.
5. **Skin changes:** More colour than normal on your skin, red spots, possibly thicker skin, and increased facial or leg hair.

**Other symptoms and signs**

- Weight loss and fatigue
- Extravascular fluid overload with ascites, peripheral oedema and pleural effusion (accumulation of fluid in the abdominal cavity, limbs and lung lining respectively)
- Arterial and venous thromboses (blood clots)
- Pulmonary disease:
  - Restrictive lung disease
  - Pulmonary hypertension
  - Respiratory muscle weakness.

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

**Other diagnostic test**

- Blood tests.
  - Full blood count: which may show thrombocytosis +/- polycythaemia
  - Endocrine panels may show abnormalities including deranged thyrotropin, glucose, and oestrogen
  - Serum or urine electrophoresis to test for the monoclonal immunoglobulin
- X-ray of bones in almost all patients show osteosclerotic lesions, which are often multiple
- Biopsy of an enlarged lymph node may show Castleman disease
- Lumbar puncture in those with neuropathy may show increased protein in the cerebrospinal fluid
- Nerve conduction studies may show changes of demyelination and axonal degeneration
- Bone marrow examination may show involvement with plasma cells
- Lymph node biopsy of enlarged nodes
- Skin biopsy results are usually nonspecific
  - Scleroderma-like lesions show nonspecific hyperpigmentation of the basal layer with inflammatory infiltrate or dermal fibrosis. Sweat glands and collagen are normal, which differentiates it from scleroderma.
  - Hyperpigmented lesions show nonspecific inflammatory infiltrate of lymphocytes and plasma cells
  - Angiomas include strawberry naevus, lobular capillary angioma, and in around 3%, glomeruloid haemangioma (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 treatment of POEMS syndrome depends on the treatment of the underlying plasma cell disorder. Most patients are treated with a combination of medical, surgical, and adjuvant therapies. The current mainstays of treatment for patients with diffuse disease include combinations of corticosteroids, low-dose alkylators, and peripheral blood stem cell transplantation following high-dose chemotherapy.

**Prognosis**

POEMS Syndrome is a chronic disease. The median survival is around a decade (8-14 years), which is three times longer than patients with multiple myeloma. However, many patients are bedridden due to neuropathy (50%).

**Conclusion**

POEMS Syndrome is a rare clinical entity caused due to plasma dyscariasis manifest a group of symptoms such as polyneuropathy, organomegaly, endocrinopathymonogammppathy, and skin changes. Cause is remain unknown but family history is prevalent in this

disorder. Each patient is unique, the individual or family should work with a clinician to determine an appropriate treatment plan.

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