

A review: crow–fukase syndrome/poems syndrome

Abstract

POEMS syndrome is a paraneoplastic syndrome whose acronym stands for less than half of the defining features of the disease, that is, polyradiculoneuropathy, organomegaly, potentially including coexisting Castleman disease, endocrinopathy, monoclonal plasma cell neoplasm, and skin changes. POEMS syndrome is caused by an underlying plasma cell disorder. The diagnosis of POEMS syndrome: The two mandatory criteria PLUS ≥ 1 major AND ≥ 1 minor criterion. Patients with POEMS syndrome are treated with medical, surgical and adjuvant therapies.

Keywords: paraneoplastic, castleman disease, endocrinopathy, organomegaly, polyneuropathy

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Introduction

POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, Skin changes) is characterized by the presence of a monoclonal plasma cell disorder, peripheral neuropathy, and Disease features are: osteosclerotic myeloma, Castleman's disease, elevation of serum vascular endothelial growth factor (VEGF), organomegaly, endocrinopathy, edema, typical skin changes, and papilledema.¹

Definition: POEMS syndrome is a paraneoplastic syndrome whose acronym stands for less than half of the defining features of the disease, that is, polyradiculoneuropathy, organomegaly, potentially including coexisting Castleman disease, endocrinopathy, monoclonal plasma cell neoplasm, and skin changes.²

Causes: POEMS syndrome is a rare paraneoplastic syndrome that is caused by an underlying plasma cell disorder.³

Signs and symptoms:⁴

- Polyneuropathy. Numbness, tingling in hands and difficulty in breathing
- Organomegaly. Splenomegaly, nodes in lymph or liver.
- Endocrinopathy. Hypothyroidism, diabetes, sexual problems weakness and metabolic problems.
- Skin changes. More pigmentation, thicker skin and hair over body.

Diagnosis

The diagnosis of POEMS syndrome: The two mandatory criteria PLUS ≥ 1 major AND ≥ 1 minor criterion.⁵

Mandatory criteria

- Polyneuropathy (typically demyelinating)
- Monoclonal plasma cell proliferation.

Major criteria

- Castleman disease
- Sclerotic bone lesions
- Vascular endothelial growth factor (VEGF) elevation

Minor criteria

- splenomegaly, hepatomegaly, or lymphadenopathy
- edema, pleural effusion, or ascites
- adrenal, pituitary, gonadal, parathyroid, thyroid and pancreatic
- Skin changes
- Papilledema
- Thrombocytosis

In addition laboratory tests such as EMG for neuropathy, CT scan, bone marrow biopsy to detect clonal plasma cells, plasma or serum protein electrophoresis to myeloma proteins, Raised blood levels of VEGF, thrombocytes, and/or erythrocyte parameters these are the supportive diagnosis for POEMS syndrome.⁶

Treatment

The treatment of POEMS syndrome depends on the treatment of the underlying plasma cell disorder. And also treated with medical, surgical and adjuvant therapies. Patients are treated with combination of corticosteroids, low-dose alkylators, and peripheral blood stem cell transplantation following high-dose chemotherapy. Caution should be taken in selecting chemotherapeutic regimen to avoid worsening of the disease. A 2014 multicenter retrospective study in Japan has shown positive results for autologous stem cell transplantation (ACST) in the treatment of patients with POEMS syndrome in terms of long-term survival and quality of life.⁷

POEMS Syndrome at current News

NBC 10 philadelphia-15-Nov-2017: Action News anchor Thomas-Laury suffered from a rare condition called POEMS syndrome that paralyzed her vocal cords, bound her to a wheelchair and required two bone marrow transplants. Then, she became addicted to the opioid prescribed to treat her chronic pain.⁸

Conclusion

By this review article I got an opportunity to review about POEMS Syndrome which is a paraneoplastic syndrome whose acronym stands for less than half of the defining features of the disease, that is, polyradiculoneuropathy, organomegaly, potentially including coexisting Castleman disease, endocrinopathy, monoclonal plasma cell neoplasm, and skin changes.

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Conflict of interest

The author declares that there is no conflict of interest.

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