POEMS syndrome: A Case Report

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BACKGROUND

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes (POEMS) syndrome is a rare plasma cell dyscrasia. A national survey conducted in Japan in 2015 estimated the prevalence of POEMS syndrome to be 0.3 per 100,000 [1].

A predominant symptom of POEMS syndrome is an ascending, symmetric, sensorimotor polyneuropathy that is progressive and often results in significant functional impairment [2]. Another predominant finding, and one of the main diagnostic criteria for POEMS syndrome, is the presence of a monoclonal protein, often lambda type [2].

The syndrome is frequently underrecognized and delays in diagnosis are not uncommon. Early diagnosis and treatment maximize the potential for neurological recovery and preservation of the patient's quality of life [1,2,3,].

Regular utilization of serum protein electrophoresis, immunofixation electrophoresis, and serum free light chain testing in patients with peripheral neuropathy could help prevent misdiagnosis [3].

This case highlights the key clinical features, diagnostic criteria, and potential for neurologic improvement after treatment in a patient with POEMS syndrome.

CASE PRESENTATION

HISTORY

- 60-year-old male with diabetes mellitus and solitary plasmacytoma referred by hematology for evaluation of ascending painful paresthesias and subsequent lower extremity weakness progressing over the course of 11 months
- He also reported sensory changes and weakness to bilateral hands, fatigue, frequent falls, and now required the use of a wheelchair and feeding assistance
- Review of systems revealed recent 30lb unintentional weight loss, worsening eczema, and orthopnea
- Sacral solitary plasmacytoma had been treated with 27 fractions of radiation three weeks prior to admission

PHYSICAL EXAM

- Significant muscle atrophy of bilateral lower extremities
- Severe distal greater than proximal upper and lower extremity weakness
- Absent proprioception and vibratory sense at bilateral ankles
- Diffuse areflexia

DIAGNOSTICS

- Skeletal survey: stable 6.0 cm sacral lytic lesion
- MRI lumbar spine: thickening and enhancement of the ventral cauda equina nerve roots
- EMG: severe polyradiculoneuropathy with features of axon loss and demyelination
- CSF analysis: elevated protein of 138 mg/dL and glucose of 198 mg/mL
- Immunofixation electrophoresis: monoclonal gammopathy, IgG lambda type
- Elevated vascular endothelial growth factor (VEGF): 638 pg/mL
- Low total testosterone 96 ng/dL and low free testosterone at 1.44 ng/dL

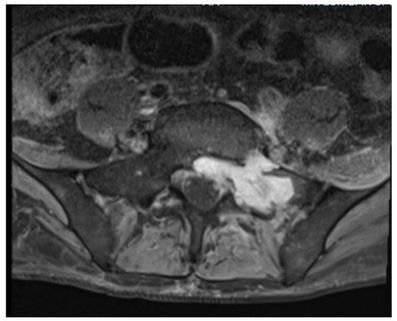
TREATMENT

- Had received 27 fractions of radiation for solitary plasmacytoma in outpatient setting
- Gabapentin 300mg TID and tramadol 50mg PRN for neuropathic pain
- Discharged to inpatient rehabilitation

OUTCOMES

- Solitary plasmacytoma successfully treated with radiation
- At four-month outpatient neurology follow up:
  - functional improvement; able to stand without assistance
  - sensory loss now only up to bilateral knees and wrists
  - neuropathic pain adequately controlled on gabapentin and tramadol

IMAGE 1



MRI L spine with contrast; axial view – sacral lytic lesion post radiation

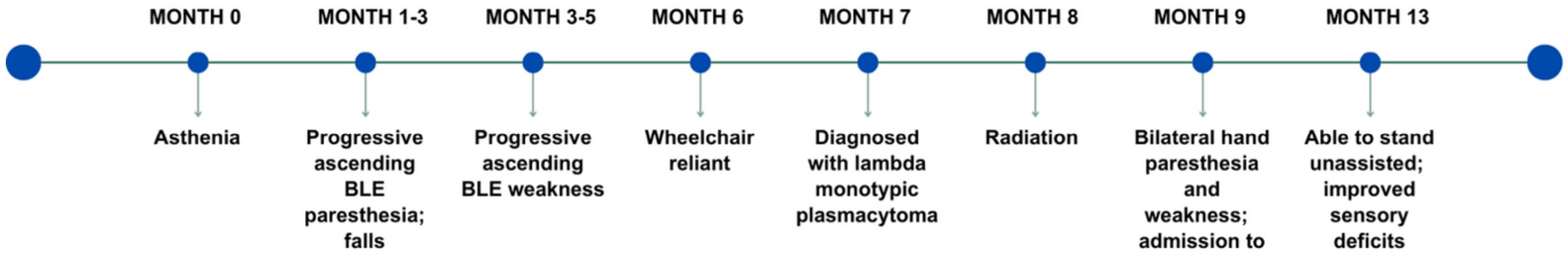


FIGURE 1: Timeline of Clinical Course

DISCUSSION

This cases highlights three major criteria required for the diagnosis of POEMS syndrome including peripheral neuropathy, monoclonal gammopathy and elevated VEGF. It also demonstrates two of the minor criteria: endocrinopathy and skin changes.

Neurological recovery after treatment of the underlying plasma cell disorder is possible but it is expected to be delayed and occur over the course of months to years [1].

TABLE 1: POEMS syndrome diagnostic criteria

Mandatory major criteria (both required)	Other mandatory criteria (one required)	Minor Criteria (one required)
- Polyneuropathy	- Castleman disease	- Organomegaly
- Monoclonal plasma cell proliferative disorder	- Sclerotic bone lesions	- Extravascular volume overload
	- Elevated vascular endothelial growth factor	- Endocrinopathy
		- Skin changes
		- Papilledema
		- Thrombocytosis/polycythemia

CONCLUSIONS

Timely diagnosis and prompt treatment of POEMS syndrome are critically important in maximizing the potential for neurologic recovery

Serum protein electrophoresis, immunofixation electrophoresis, serum free light chains, and vascular endothelial growth factor testing can aid in distinguishing POEMS syndrome from other conditions.

REFERENCES

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