

POEMS Syndrome

Ling CSY¹; Komahen C¹; Khan ES²; VA Jacob¹

¹Department Of Orthopaedics, Hospital Tengku Ampuan Afzan, Pahang

² Orthopaedics, Traumatology And Rehabilitation, International Islamic University Malaysia

INTRODUCTION:

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS syndrome) is a multiorgan disease with the setting of plasma cell dyscrasia. POEMS syndrome is a disease that remains a challenge to the treating doctor due to its rarity and its debilitating condition.

MATERIALS & METHODS:

A 31 year old gentleman who had history of L3 pathological burst fracture with posterior spinal instrumentation and fusion three years ago presented with bilateral lower limb weakness MRC grade 1-2. There were presence of pleural effusion, ascites, enlarged cervical lymph nodes with skin lesions.

RESULTS:

CT scan showed a mass at the level of L3 with calcified foci and lytic lesion causing spinal stenosis. Biopsy of the tumour showed plasma cell neoplasm. Patient was diagnosed to have POEMS syndrome. He underwent a debulking surgery of the tumour, posterior shortening of the spinal column with Ponte osteotomy and spinal decompression via wide laminectomy and exchange of rods.

DISCUSSIONS:

POEMS syndrome is diagnosed when the patient fulfills 2 mandatory criterias which are plasma cell neoplasm and polyneuropathy, 1 major criteria and 1 minor criteria. The patient had polyneuropathy with plasma cell tumour over the level of L3. The tumour was associated with abnormally dense bone structures (calcified foci) which fulfilled one of the major criteria. The other symptoms fulfilled the minor criterias. The patient developed neurological deficit due to the progression of the disease causing vertebral body collapse. The loss of anterior support of the spinal column caused the failure of implant. Post operatively, the patient

received radiotherapy as a treatment for the disease. At latest follow up patient was able to ambulate without walking aid.

CONCLUSION:

POEMS syndrome is a multisystemic disease which requires multidisciplinary approach to achieve the best outcome for the patient.

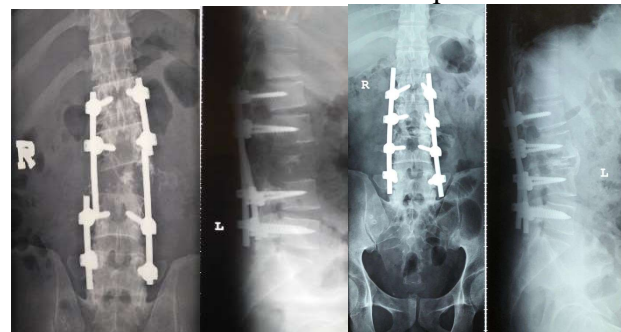


Figure 1: X ray before and after revision surgery



Figure 2: MRI scan

REFERENCES:

1. Dispenzieri A. POEMS syndrome: 2011 update on diagnosis, risk-stratification, and management. *Am J Hematol* 2011;**86**(7):591-601.
2. Dispenzieri A, Kyle RA, Lacy MQ, et al. POEMS syndrome: definitions and long-term outcome. *Blood* 2003;**101**(7):2496-2506.