

- Abstract -

A Case of POEMS Syndrome Presented as CIDP

**Eun-Yeon Joo, M.D., Jee-Young Oh, M.D., Hye-Soo Ku, M.D.*,
Kee-Duk Park, M.D., Kyung-Gyu Choi, M.D.**

Department of Neurology, Department of clinical pathology, Ewha Womans University College of Medicine*

POEMS syndrome is a rare multisystem disorder manifests as polyneuropathy, organomegaly, endocrinopathy, M-proteins and skin changes. We report here the case of 54-year-old woman with POEMS syndrome that was followed by a 1-year history of symptoms that mimicked a chronic inflammatory demyelinating polyradiculoneuropathy(CIDP). The laboratory examination revealed multiple osteosclerotic bone lesions and IgG paraproteinemia later in the course of the disease. Initial treatment with IVIg showed marked improvement of generalized weakness from bedridden state to walking with minimal assistance. In spite of the substantial improvement of peripheral neuropathy with steroid treatment the patient died suddenly at home 5 months after discharge from hospital.

Key Words : POEMS syndrome, CIDP, Steroid

thy ,
POEMS 1980 Bardwick
.1 1

POEMS (polyneuropathy), 1
(organomegaly), (endocrinopathy),
(monoclonal gammopathy),

POEMS

, , , , , , , ,
, , , , , , , ,
, , , , , , , ,
가 .2,3 Crow- 53 98 3 , 10
Fukase , osteosclerotic myeloma with polyneuropathy,

Address reprint requests to Kyoung-Gyu Choi, M.D.
Department of Neurology, Ewha Womans University College of Medicine, # 911-1 Mok-dong Yangcheon-ku, Seoul, 158-701, Korea
Tel : 82-2-650-5036, Fax : 82-2-650-5162, e-mail : kgyuchoi@hotmail.com

CIDP

POEMS

1 :

가
86mg/dl, 0/mm³,
395mmH₂O

immunog-
(Fig. 1).
lobulin G- type
, 510mmH₂O,
0/mm³,
184mg/dl, oligoclonal band가

가
Pelvic MRI pelvis, iliac bone sacrum
osteosclerotic lesions

가
, 7, 12 , 1 ,
hot uptake
가 (Fig. 2).

(Fig. 3).

7
120mg/dl, 0/mm³, 300mmH₂O

가
, Luxol fast
blue Masson trichrome 가
가 , Bodian
(Fig. 4).

LE cell, ANA, cryoglobulin, vitamin B12, folate, por-
phyria screening test,
VDRL titer, 가

0.6% ,
POEMS , prednisone

가
.98 12
,99 3

99 4 10
, , 2

가
T₃
56.8(80~200ng/dl), T₄ 4.2(4.5~12.5ng/dl) TSH 3.3(0.3~5
ng/dl)
M-peak

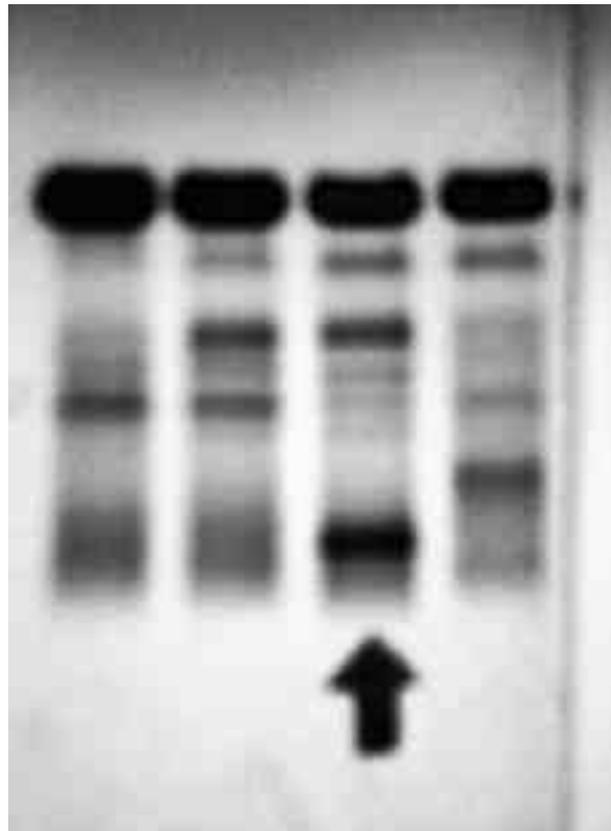


Fig. 1. Immunoelectrophoresis
: Immunoglobulin G- type gammopathy



Fig. 2. Whole body bone scan
: Multiple hot uptake on manubrium, T7, T12, L1, cranium, both tibiae, both knees and ankle joints

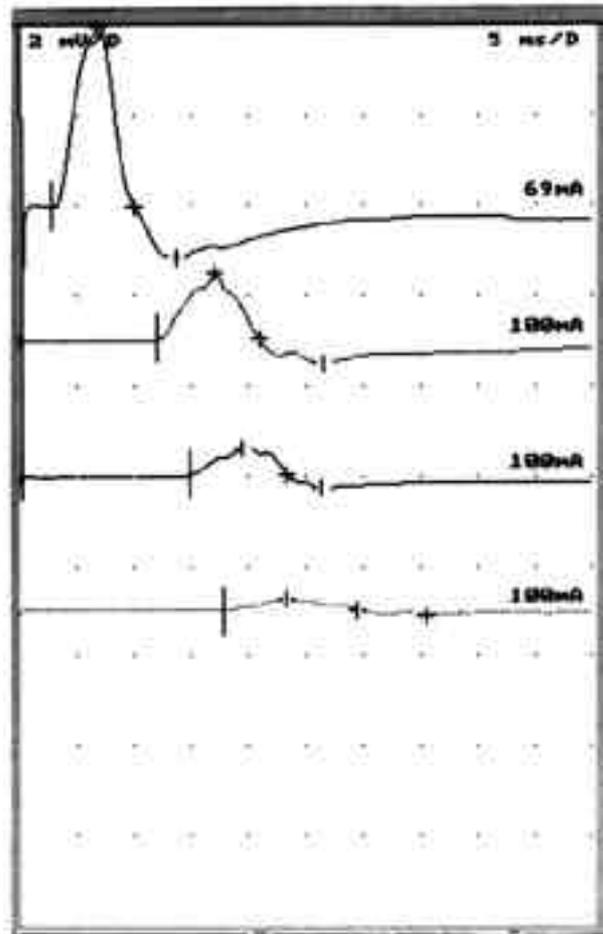


Fig. 3. Nerve conduction study(2nd study, 5/7/98)
: Conduction block on right ulnar nerve motor conduction study

60mg/d melphalan , 가
 ,
 ,
 , 2
 . 3
 ,
 5

POEMS 40~50
 , 2:1
 .^{2,4}
 myeloma, Castleman , Waldenstrom macroglobulinemia,
 extramedullary plasmacytoma, M protein alone, polyclonal
 protein alone, , nonhematologic malignan-
 cies .²

가 . Crow
 , 1938 Scheinker가 sternum myeloma가

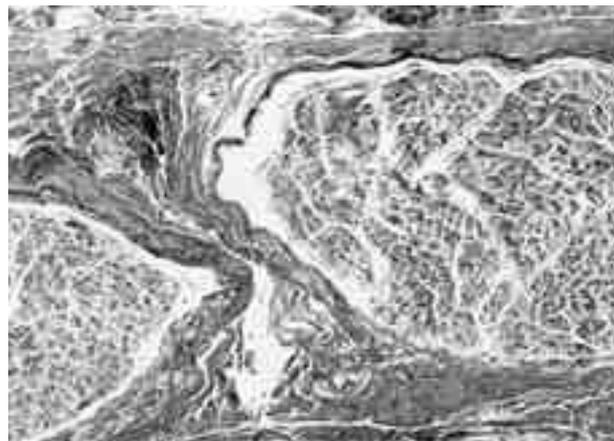


Fig. 4. Sural nerve biopsy
: Cross section of sural nerve, showing a marked increase of endoneurial connective tissue.
Masson trichrome stain * 100

Table 1. Nerve Conduction Study

motor NCV	1st study(98/3)			2nd study(98/5)			3rd study(99/4)		
	m/sec	m/sec	Amp(mV)	msec	m/sec	Amp(mV)	msec	m/sec	Amp(mV)
Median nerve									
TL	3.9			3.9			5.0		
Wrist -elbow		5.5	39.5		2.1	29.6		1.6	25.0
Elbow-axilla		4.4	40.4		1.8	25.7		1.3	25.7
F-wave latency	29.6			NP			NP		
Ulnar nerve									
TL	2.7			2.8			3.9		
Wrist-elbow		7.8	36.9		5.1	25.3		4.1	19.5
Elbow-axilla		6.4	40.9		2.0	31.5		3.0	17.8
Peroneal nerve									
TL	4.8			NP			NP		
Ankle-knee		0.4	25.6						
F-wave latency	NP			NP			NP		
Sensory NCV									
Median nerve									
	m/sec Amp(μV)								
Finger-wrist		3.9	31.3	NP			NP		
Wrist-elbow		7.1	33.6						
Elbow-axilla		17	33.3						
Sural nerve			NP	NP			NP		

39 M-protein
 , Crow가 2 osteos-
 clerotic & osteolytic multiple myeloma .
 .¹² Fukase IgG-
 extrameduallary plasmacytoma IgA-^{4,6}
 POEMS . 1984 Nakan- M-protein, -light chain ,
 ishi Crow-Fukase 102 (plasma cell) .³ TNF- , Interleukin-1, 6 Interfer-
 (M-protein) 27% .² on- pro-inflammatory cytokine vascular endothe-
 , 가 lial growth factor(VEGF)가 .^{7,8} -
 POEMS ,⁹ pro-inflammatory cytokines POEMS
 가 plasma cell dyscrasia 가
 .⁵ 1993 Scully POEMS .
 , POEMS, Crow-Fukase ,
 .¹³ ,
 1980 Bardwick POEMS cyclophosphamide, melphalan prednisolone,
 가 .^{1,2,4} 7
 가

POEMS
1987

5 12

가
가 (,)
가
POEMS

가

REFERENCES

(CIDP) (conduction block,
prolonged distal latency & F-wave latency)

POEMS

CIDP

가

POEMS

1 Guillian-Barre

가, 2

CIDP

10

POEMS

3

20

CIDP

POEMS

, 6

cyclophosphamide

.¹⁰ Benito-Leon

IVIg

.¹¹

prednisolone(60mg/day)

가

가

, 가

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