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= Abstract =

Clinicial Significance of Spontaneous Pneumomediastinum in Dermatomyositis/Polymyositis

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Objective: Pneumomediastinum (PnM), a rare complication of dermatomyositis and polymyositis (DM/PM), is sporadic and has an unclear pathogenesis. PnM is almost always associated with interstitial lung disease (ILD), and is a poor prognostic factor in inflammatory myositis patients. We studied the prevalence of PnM in Korean DM/PM and its clinical significance.

Methods: We retrospectively studied the medical records of 161 patients diagnosed with DM/PM meeting Bohan-Peter's criteria at Hanyang University Hospital for Rheumatic Diseases from 1995 to 2010. We collected following findings; demographic data, diagnosis, lung involvement, cause of death, and duration from diagnosis to death.

Results: One hundred nineteen patients (73.9%) were DM and 42 patients (26.1%) were PM. Eighty three patients (51.6%) developed ILD at diagnosis or during follow up. Eighteen patients (11.2%) died because of ILD aggravation, infection, or malignancy. The mean duration from diagnosis to death was 11.5 months, with 10 patients (6.2%) dying from from ILD aggravation but none with spontaneous PnM. 6 patients (3.7%) presented with PnM, and it was associated

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with ILD worsening in all cases. PnM resolved with O_2 inhalation, corticosteroids, and/or immunosuppressive agents after 11 weeks (mean) of therapy

Conclusion: PnM is rare but associates with DM and aggravation of ILD. PnM does not usually cause fatalities and can be cured by appropriate therapy.

Key Words: Pneumomediastinum, Dermatomyositis/polymyositis, Interstitial lung disease, Prognosis

서 론

/ (dermatomyositis/polymyositis)
, ,
. 10～22%
, (interstital lung disease)
(1-3). /
(3).
(pneumomediastinum)
, ,
(4). / ,
, ,
(5-7). /
10～43%
. /
(8,9),
Kuroda cyclosporine
(10,11),
(12).
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대상 및 방법

1. 연구대상

1995 1 2010 3
ICD 10 M33, M330, M331, M332, M339 . 161
- (Bohan-Peter's criteria)
/ (13).
161
. , ,
, , CK/LDH , aldolase , (ANA) Jo-1,
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2. 간질폐질환의 방사선학적 분류 및 경과

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. (reticulonodular opacity), (linear opacity)
(ground-glass opacity), (honeycombing),
(traction bronchiectasis)
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,
(idiopathic pulmonary fibrosis),

(bronchiectasis oblitrans with organizing pneumonia, BOOP),

(non-specific interstitial pneumonitis, NSIP),

(acute interstitial pneumonitis, AIP) (1) (14).

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3. 자발종격동기종의 진단

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X

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X

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결 과

1. 일반적인 특징

161 31 130

. 43.2 (6～75) , 30.4 (1～155) . 119 (73.9%), 42 (26.1%) .

1 : 3.8 1 : 6 .

Table 1. Classification and patterns of Interstitial lung disease

Morphologic pattern	Clinical diagnosis	Histologic features	Imaging features
Usual interstitial pneumonia	Idiopathic pulmonary fibrosis	Spatial and temporal heterogeneity, dense fibrosis, fibroblastic foci, honeycombing	Basal, peripheral predominance, often patchy, reticular abnormality, honeycombing
Nonspecific interstitial pneumonia	Nonspecific interstitial pneumonia	Spatially and temporally homogeneous lung fibrosis or inflammation	Basal predominance, ground-glass abnormality, reticular abnormality
Desquamative interstitial pneumonia	Desquamative interstitial pneumonia	Diffuse macrophage accumulation in alveoli	Basal, peripheral predominance ground-glass attenuation; sometimes cysts
Respiratory bronchiolitis	Respiratory bronchiolitis-associated interstitial lung disease	Peribronchiolar macrophage accumulation, bronchiolar fibrosis; macrophages have dusty, brown cytoplasm	Centrilobular nodules, ground-glass attenuation
Organizing pneumonia	COP, BOOP	Patchy distribution of intraluminal organizing fibrosis in distal airspaces; preservation of lung architecture; uniform temporal appearance; mild interstitial chronic inflammation	Ground-glass attenuation; consolidation basal, peripheral predominance
Diffuse alveolar damage	Acute interstitial pneumonia	Diffuse distribution, uniform temporal appearance, alveolar septal thickening due to organizing fibrosis, airspace organization, hyaline membranes	Diffuse, ground-glass attenuation, consolidation
Lymphoid interstitial pneumonia	Lymphoid interstitial pneumonia	Diffuse lymphoplasmacytic infiltration of alveolar septa	Ground-glass attenuation, cysts

COP: cryptogenic organizing pneumonia, BOOP: bronchiectasis obliterans with organizing pneumonia

43.6 (6～75), 38.7 (16～75) . 5
13 (8.1%) . 78 14 (17.9%),
. 83 (51.6%) 30 (38.5%), 25 (32.1%),
69 (58.0%), 1 (1.3%),
14 (33.3%) (2). 8 (10.3%) (3).

2. 예후

7 (9.0%),
18 (11.2%) 1
11.5 (1～ (1.3%) . 51 (65.4%)
37) . , 17 (21.8%)
(10 , 6.2%), (5 , , 10
3.1%), (3 , 1.9%) . (12.8%) 1
(2), (1), ,
(1), T (NK/T cell (4).
lymphoma)(1) .

4. 자발성 종격동기종 발생 환자들의 특징

(2). 6 (3.7%) 7
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1

3. 간질폐질환의 진단과 경과

83 (51.6%) . 9

Table 2. Characteristics of dermatomyositis/polymyositis patients

	Dermatomyositis	Polymyositis
Numbers of patients	119 (73.9%)	42 (26.1%)
Mean age at diagnosis (years)	43.6 (6～75)	38.7 (16～75)
Mean period of follow up (months)	26.6 (1～155)	32.6 (1～121)
Female/Male	94/25	36/6
Interstitial lung disease	69 (42.9%)	14 (33.3%)
Malignancy	11 (9.2%)	2 (4.8%)
Death causes	16 (13.4%)	2 (4.8%)
Interstitial lung disease	9	1
Malignancy	4	1
Sepsis	3	

Table 3. Radiologic classification of interstitial lung disease

	IPF	BOOP	NSIP	AIP	Mixed	Unknown
Numbers	14	30	25	1	8	5
Result						
Alive (PnM)	14	26 (4)	23 (1)	0	4 (1)	5
Expire	0	4	2	1	4	0

IPF: idiopathic pulmonary fibrosis, BOOP: bronchiectasis obliterans with organizing pneumonia, NSIP: non-specific interstitial pneumonitis, AIP: acute interstitial pneumonitis, PnM: pneumomediastinum

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(42～59) . . 3 ,
5 CK , 6
, LDH 1 .
Jo-1 , 3 X . X
speckled pattern (1), cyto-
plasmic pattern (2) . 1 . 6
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4 (collar incision) (squeezing)
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Table 4. Clinical course of interstitial lung disease among dermatomyositis/polymyositis patients

	No interval change	Slow progression	Rapid progression	Unknown
Numbers	51	17	10	5
Result				
Alive (PnM)	51	16 (6)	0	5
Expire		1	10	0

PnM: pneumomediastinum

Table 5. Characteristics of 6 cases of spontaneous pneumomediastinum at dermatomyositis/polymyositis patients

Age /sex	CK	LDH	Aldo.	ANA	Anti Jo-1	CV	ILD agg.	Treatment before PnM	PM/DM, time to PnM	Treatment after PnM	Recovery
59/F	24	142	18.4	(+)*	(−)	(−)	(+)	PDS	DM, initial.	Steroid pulse, CsA	5 month
51/F	32	305	5.4	(−)	(−)	(−)	(+)	PDS, CsA, HCQ	DM, 1 month	Steroid pulse, CsA, O_2	2 month
42/F	166	267	37.6	(+)†	(−)	(−)	(+)	PDS, CsA, HCQ	DM, 2 years	Steroid pulse, CsA, O_2	2 weeks
43/F	125	448	12.9	(−)	(−)	(−)	(+)	PDS, CsA	DM, 6 month	IVIG, PDS, CsA, HCQ, O_2,	5 month
44/F	49	251	3.7	(+)‡	(−)	(−)	(+)	PDS, AZA	PM, initial.	PDS, CsA, O_2, Collar	2 month
recur	290	346	7.1				(+)	PDS, CsA	DM, 1 week	incision Unaltered, O_2	1 month
54/F	26	122	7.1	(−)	(−)	(−)	(+)	PDS, CsA	DM, 1 year	IVIG, O_2, PDS, Tacrolimus	2 weeks

Aldo.: aldolase, CV: cutaneous vasculopathy, agg.: aggravation, PDS: prednisolone, CsA: cyclosporine A, HCQ: hydroxychloroquinolone, AZA: azathioprine, IVIG: intravascular immunoglobulin. *Speckled pattern 1 : 640, †Cytoplasmic pattern 1 : 320, ‡Cytoplasmic patthern 1 : 160

Table 6. Comparison of clinical features between Interstitial lung disease patients with and without pneumomediastinum

	Pneumomediastinum (+)	Pneumomediastium (−)
Numbers of patients	6 (7.1%)	77 (92.9%)
Age	48.8 (42~59)	45.4 (27~75)
Male/Female	0/6	14/63
DM/PM	6/0	63/14
Follow up duration (months)	22 (2~57)	30.3 (1~155)
ANA (+)	3 (50%)	47 (61.0%)
CK/LDH (mean)	101.7/268.7	596.4/358.3

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10~43%

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(mucosal barrier)

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Tong

(17,23).

(9). Benoit Le Goff

(DLCO) (vital capacity)

Table 7. Review of reported spontaneous pneumomediastinum cases with dermatomyositis/polymyositis in Korean

Ref.	Age /Sex	CK	Aldolase	Anti Jo-1	CV	ILD agg.	Treatment before PnM	PM/DM, time to PnM	Treatment after PnM	Recovery
11	40/F	1,521	22.6	(−)	(+)	(+)	PDS, IVIG	DM, 3 weeks	Steroid IV, CsA, O_2	3 weeks
25	45/F	NL	?	(−)	(+)	(+)	PDS, AZA, CPM	DM, 2 months	Unaltered	3 month
26	38/F	131	13.2	(−)	(+)	(+)	PDS, HCQ	DM, 4 months	Unaltered, O_2	1 month
27	18/M	4.6	13.8	(−)	(−)	(+)	PDS, HCQ, IVIG	DM, 1 months	PDS, CPM, O_2, thoracostomy	Expire (2 m.)
28	56/F	1,405	?	(−)	(−)	(+)	PDS	DM, 2 weeks	PDS, O_2	Expire (1 m.)
29	53/F	20	?	(−)	(−)	(+)	PDS, AZA	DM, ?	Unaltered, O_2	?
29	41/F	38	?	(−)	(−)	(+)	PDS	DM, 1 month	PDS, AZA, O_2	2 weeks
30	36/F	73	?	(−)	(+)	(+)	PDS, MTX, HCQ	DM, 4 months	PDS, HCQ, MTX, IVIG, O_2	Expire (1 m.)
30	45/F	156	?	(−)	(+)	(+)	PDS, HCQ	DM, 11 months	Steroid IV, CsA IV, O_2	Expire (1 m.)

Ref.: reference, CV: cutaneous vasculopathy, agg.: aggravation, NL: normal, ?: unknown, PDS: prednisolone, CsA: cyclosporine A, HCQ: hydroxycloroquinolone, AZA: azathioprine, IVIG: intravascular immunoglobulin, M: month

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(24). Jo-1

50% Jo-1

(24). (31).

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Jo-1 , Yoshida

(7) (11,25-30). 21 16

9 Jo-1

(12). Jo-1

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CK/LDH ,

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