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Between 1975 and 1992, 78 patients with polymyositis/dermatomyositis (PM/DM) who met the criteria for diagnosis of PM/DM according to Bohan and Peter, were admitted to the Medical Service Section of Chulalongkorn Hospital, Bangkok. Polymyositis and dermatomyositis were found in 70 and 30 percent of the total cases, respectively. The female-to-male ratio was 3:1 among the total number of cases; however, the ratio was 11:1 in the group of PM/DM patients associated with other connective tissue diseases. Idiopathic PM/DM (55.1%) was more commonly observed than PM/DM associated with other connective tissue disease (30.8%) and PM/DM associated neoplasm (14.1%). Childhood type PM/DM was not found among the patients studied. Age distribution was equally similar, except for those at younger and older ages in PM/DM associated with other connective tissue disease and neoplasm, respectively. Proximal muscle weakness was the most common feature (96%) followed in frequency by myalgia (59%), arthralgia (44%), dysphagia (32%), respiratory symptoms (28%), Raynaud's phenomenon (24%) and arthritis (15%). Abnormal findings in muscle enzymes, electromyelogram results, and muscle histopathology were evident in 83, 88 and 74 percent of the cases, respectively. Carcinomas of the nasopharynx and of the lung were common neoplasms.

In conclusion, the clinical features of PM/DM in our series of Thai patients are similar to those previously reported elsewhere.

Key words: *Polymyositis, Dermatomyositis.*

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ผู้ป่วย Polymyositis (PM) และ Dermatomyositis (DM) ที่เข้าเกณฑ์การวินิจฉัยของ Bohan & Peter ที่เข้ารับการรักษาแบบผู้ป่วยใน ในโรงพยาบาลจุฬาลงกรณ์ ในระหว่าง ค.ศ. 1975-1992 ได้ถูกนำมาศึกษาลักษณะทางคลินิก ผลการศึกษาพบว่ามีผู้ป่วยทั้งหมด 78 คน โดยเป็น PM 70% และ DM 30% อัตราส่วนเพศชายต่อเพศหญิงเท่ากับ 3:1 แต่ในกลุ่มที่สัมพันธ์กับโรคทาง Connective tissue จะเท่ากับ 11:1 เมื่อศึกษาถึงกลุ่ม (Classification) พบว่าอยู่ในกลุ่ม Idiopathic PM/DM มากที่สุดคือ 55.1% รองลงมาเป็นกลุ่มที่สัมพันธ์กับโรคทาง Connective tissue 30.8% และกลุ่มที่สัมพันธ์กับมะเร็ง 14.1% เมื่อศึกษาถึงกลุ่มอายุพบว่าในแต่ละกลุ่มใกล้เคียงกัน ยกเว้นในกลุ่มที่สัมพันธ์กับโรคทาง Connective tissue พบอายุน้อยกว่า และในกลุ่มที่สัมพันธ์กับมะเร็งจะพบอายุมากกว่าในด้านอาการแสดงพบว่าอาการกล้ามเนื้ออ่อนแรงพบมากที่สุดคือ 96% ตามด้วยปวดกล้ามเนื้อ 59% ปวดข้อ 44% กลืนลำบาก 32% อาการทางระบบหายใจ 28% Raynaud's phenomenom 24% และข้ออักเสบ 15% การศึกษาการตรวจทางห้องปฏิบัติการพบว่ามีความผิดปกติในเอนไซม์กล้ามเนื้อ 83% คลื่นไฟฟ้ากล้ามเนื้อ 88% และพยาธิวิทยากล้ามเนื้อ 74% และเมื่อศึกษาในกลุ่มที่สัมพันธ์กับมะเร็งพบว่า มะเร็งปอด และมะเร็งของ Nasopharynx พบมากที่สุด

โดยสรุป พบว่าลักษณะทางคลินิกของผู้ป่วย PM/DM ที่ทำการศึกษานี้ไม่แตกต่างไปจากรายงานเดิมที่เคยทำการศึกษาจากต่างประเทศ

Polymyositis (PM) and dermatomyositis (DM) are chronic inflammatory myopathies caused by unknown causative agents, although the cell mediated immune mechanism may be the main pathophysiologic disease mechanism.[1] The clinical features of PM/DM have been studied extensively elsewhere,[2-5] whereas they have rarely been reported in Thailand.

The purpose of this study is to present the clinical spectrum of PM/DM observed during a 17-year period at the Medical Service Section of Chulalongkorn Hospital, Bangkok, Thailand.

Patients and methods

A total of 78 patients diagnosed as having PM/DM were studied. All were in-patients at the Hospital's Medical Service Section during the period January 1975 to December 1992. The criteria for diagnosis of PM/DM were as according to Bohan and Peter[1] and they included the following:

1. Symmetric weakness of the limb-girdle muscle and anterior neck flexor,progressing over weeks to months, with or without dysphagia or respiratory muscle involvement.

2. Skeletal muscle histology showing evidence of necrosis of type 1 and 2 muscle fibers, phagocytosis, regeneration with basophilia, large sarcolemmal nuclei and prominent nucleoli, atrophy in a perivascular distribution, variation in fiber size and inflammatory exudate.

3. Elevation of serum skeletal muscle enzyme (CPK, aldolase, SGOT, SGPT and LDH).

4. EMG triad of short, small, polyphasic motor units, positive fibrillation waves, insertional irritability and bizzar high-frequency discharge.

5. Dermatologic features included a lilac (heliotrope) discoloration of the eyelids with periorbital edema, a scaly erythematous dermatitis over the dorsa of the hands, especially over the MCP and PIP joints (Grotton's sign) and involvement of the knee, elbows and medial malleoli, face, neck and upper trunk.

The difference between polymyositis and dermatomyositis is observed in dermatologic manifestations. The spectrum of PM/DM[1] can be classified as follows:

Group I : Primary idiopathic PM
Group II : Primary idiopathic DM
Gruop III : PM or DM associated with neoplasia
Group IV : Childhood PM or DM associated with vasculitis
Group V : PM or DM associated with collagen vascular disease (overlap group)

The documentation included sex, age at onset, clinical features, classification and laboratory findings.

Results

Of the 78 patients who met the criteria for diagnosis of PM/DM, 55 of them (70 percent of the total) were polymyositis cases, and 23 patients (30 percent of the total) were dermatomyositis cases. Idiopathic PM/DM (55.1%) was more commonly evident than PM/DM associated with other connective tissue diseases (30.8%) and PM/DM associated neoplasms (14.1%). Among the total number of cases the female-to-male ratio was 3:1; and the female-to-male ratio was even more prominent (11:1) in the group of

PM/DM associated with other connective tissue diseases as compared with the PM/DM associated neoplasm group (3:1) and the idiopathic group (2:1) (as shown in Table 1).

Table 1. Sex and type of PM/DM.

			Idiopathic		Neoplasm		CTND	
	No.	%	43	(55.1%)	11	(14.1%)	24	(30.8%)
	78		F	M	F	M	F	M
Polymyositis (PM)	55	70.5	17	11	3	2	21	1
Dermatomyositis (DM)	23	29.5	12	3	5	1	1	1
			29	14	8	3	22	2
Female : Male			2:1		3:1		11:1	
			3:1 (all)					

Table 2 shows that the age distribution for the total group of patients was between 20 and 59. Ages between 20 and 49 years were more commonly seen in patients with connective tissue disease whereas patients with neoplasm tended to be slightly older.

Table 2. Type of PM/DM by age distribution.

Age group (years)	Idiopathic PM/DM		Neoplasm PM/DM		CNTD PM/DM		Total PM/DM	
	No.	%	No.	%	No.	%	No.	%
10-19	2	4.6	1	9.1	1	4.2	4	5.1
20-29	7	16.3	3	27.3	5	20.8	15	19.2
30-39	10	23.3	1	9.1	9	37.5	20	25.6
40-49	5	11.6	-	-	6	25.0	11	14.2
50-59	12	27.9	2	18.2	2	8.3	16	20.5
> 60	7	16.3	4	36.3	1	4.2	12	15.4
	43	100	11	100	24	100	78	100

Muscle weakness, particularly in the proximal group of muscles, was the most common manifestation (96%) followed in frequency by myalgia (59%), arthralgia (44%), dysphagia (32%), respiratory symptoms (28%), Raynaud's phenomenon (24%) and arthritis (15%). The details of the manifestations are shown in Table 3.

Table 3. Clinical manifestations among 78 cases of PM/DM.

Type	Idiopathic		Neoplasm		CNT		Total	%
Symptoms	PM	DM	PM	DM	PM	DM		
Muscle weakness								
- Proximal	27	14	5	5	22	2	75	96.1
- Distal	7	4	-	2	5	1	19	24.4
Articular								
- Arthralgia	11	4	2	-	16	1	34	43.5
- Arthritis	4	4	-	-	3	1	12	15.4
Raynaud's phenomenon	3	1	1	1	12	1	19	24.4
Myalgia	13	10	2	5	14	2	46	59.0
Dysphagia	2	8	2	4	7	2	25	32.1
Respiratory symptoms	4	4	2	2	9	1	22	28.2

Anemia, defined by a hematocrit level of less than 30, was observed in 68 percent of the total number of patients. Antinuclear antibody (by indirect immunofluorescent study) and rheumatoid factor were observed in 46 and 36 percent, respectively, of the total number of patients, (as shown in Table 4).

Table 4. Laboratory findings among 78 cases of PM/DM.

Type	Idiopathic		Neoplasm		CNT		Total	%
Lab	PM	DM	PM	DM	PM	DM		
Anemia	15	11	5	2	19	1	53	68
Elevated ESR	9	7	2	1	17	1	37	47.4
ANA +ve	7	5	2	-	14	2	30	38.5
RF +ve	5	4	1	-	5	1	16	20.5

Abnormal findings in muscle enzymes, electromyelogram, and muscle histopathology were observed in 83, 88 and 74 percent of the patients,respectively, (as shown in Table 5).

Table 5. Findings according to diagnostic criteria of PM/DM.

Type / Criteria Dx.	No. of cases examined	Idiopathic PM	Idiopathic DM	Neoplasm PM	Neoplasm DM	CNT PM	CNT DM	Total	%
1. Proximal muscle weakness	78	27	14	5	5	22	2	75	96.2
2. ↑ Muscle enzyme	78	24	12	4	3	21	1	65	83.3
3. EMG	72	21	14	3	5	19	2	64	88.8
4. Muscle Bx	58	18	9	-	3	11	2	43	74.1
5. Skin lesion	23								
- Heliotrope		-	11	-	4	-	1	16	69.6
- Grotton		-	8	-	4	-	1	13	56.5

In cases of dermatomyositis, heliotrope and Grotton's signs were found in 69 and 56 percent of the cases respectively.

Systemic sclerosis (12/24 cases) was more commonly observed in the group of PM/DM cases associated with connective tissue disease than in those with systemic lupus erythematosus (9/24), Sjogren's syndrome (1/24) and rheumatoid arthritis (1/24).

In the group of PM/DM with associated neoplasms (11 cases), carcinoma of the nasopharynx was evident in three cases, carcinoma of the lung in three cases, and one case of each of multiple myeloma, chronic myelocytic leukemia, malignant histiocytosis and carcinoma of the cervix.

Table 6. Frequency of PM and DM associated with a second CNT disorder or malignancy.

Study, Year	Total No. of cases	Asssociated with CNT disease No. (%)	Associated with malignancy No. (%)
Walton & Adams,1958	40	8(20)	6(15)
Pearson, 1963	48	2(4)	9(19)
Barwick & Walton, 1963	52	16(31)	6(12)
Vignos et al., 1964	38	2(5)	5(13)
Rose & Walton, 1966	89	17(19)	14(16)
Devere & Bradley, 1975	118	30(25)	9(8)
Bohan et al., 1977	153	32(21)	13(8)
Rowland et al.,1977	83	20(24)	1(1)
	47	15(32)	2(4)
Henriksion & Sandstedt,1982	107	16(15)	7(7)
Hochberg et al., 1983	76	18(24)	6(8)
Total	**851**	**176(21)**	**78(9)**

Discussion

A few classifications of PM/DM have been proposed;[6] however, Bohan and Peter's classification[1] is still practical and has been widely used. The incidence of PM/DM has been reported to be 0.5 to 8.4 cases per million.[5] Although no survey has been taken among the Thai population, our study shows that the prevalence of PM/DM, as gunged by admission to Chulalongkorn Hospital, is about five patients per year, and that polymyosis is more common than dermatomyosis, and this is similar to the result of studies reported elsewhere.[2,3,5]

In other series,[2,3,5] idiopathic PM/DM has been more commonly observed than PM/DM associated with other connective tissue diseases and PM/DM associated with neoplasms. Neoplasms have been reported in 15-20 percent[4,7-10] of other studies, which is not different from our study (14%).[4] Carcinoma of the lung and breast have been reported in association with PM/DM;[7-11] however, carcinoma of the nasopharynx and carcinoma of the lung fairly were common in our cases, which is similar to that reported in a series in Singapore.[12] Dermatomyosis has been more frequently seen in the group of PM/DM patients associated with neoplasms, and also more often in older age groups which is the same as in previous reports.[9,11]

There were no differences in terms of the female sex being prevalent in our study as compared with other series.[1,2,6] There was evidence of a

higher female-to-male ratio in the group of PM/DM associated with other connective tissue diseases. Sex distribution was not different compared with other series; also, involvement of patients at younger ages was observed in the group of patients with PM/DM associated with other connective tissue diseases.

Muscle weakness, particularly in the proximal group of muscles, was the main feature of PM/DM. Other features such as dysphagia, Raynaud's phenomenon, articular involvement, heliotrope and Grotton's signs were similarly observed as in other series.[1,2,6,13-15]

In other studies,[16-19] pulmonary fibrosis was reported in 5-10 percent of the cases and pulmonary function tests showed a restrictive pattern. In our study, respiratory symptoms were evident in 28 percent of the cases.

Abnormalities in muscle enzymes, EMG and histopathologic findings in PM/DM have been reported,respectively, in 95, 75, and 70 percent of the cases reported.[20-23] Those results were not different from our own cases.

Thus, proximal muscle weakness, abnormalities in muscle enzymes, EMG and histopathology are the main features of PM/DM.

Antinuclear antibodies and rheumatoid factor were observed as in other series; antinuclear antibodies were more commonly evident in cases of systemic lupus erythematosus.[20,21,24]

In conclusion, the clinical features of polymyositis/dermatomyositis in Thai patients are similar to those reported in studies elsewhere.

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