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## Myopathy in Autoimmune Diseases – Primary Sjögren’s Syndrome and Dermatomyositis

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### 1. Introduction

Myopathy, which clinically shows muscular pain (myalgia), weakness, cramps, stiffness and spasm, is one of neuromuscular disorders due to inflammation and/or dysfunction of muscle fibers. “Myositis”, which is a general term for inflammation of the muscle, is pathologically an inflammatory myopathy seen mainly in autoimmune disorders including dermatomyositis (DM). The myopathy is classified by National Institute of Neurological Disorders and Stroke (NINDS) as indicated in Table 1 (1). We here focus myopathy on primary Sjögren’s syndrome (pSjS) associated with myalgia “mimicking DM”, as previously reported (2), and the inflammatory myopathy of DM (Table 2). Most of SjS is a secondary disorder to systemic autoimmune diseases including systemic lupus erythematosus (SLE), systemic sclerosis, DM, and so on. However, SjS, which is not associated with other autoimmune diseases, is considered to be an idiopathic primary disorder characterized by sicca symptoms. It is known that pSjS may be associated with fever, fatigue, myalgia, arthralgia, cutaneous vasculitis, etc. in addition to sicca symptoms (4-8).

DM is also characterized by myalgia, muscular weakness and fatigue due to inflammatory myopathy that ultimately progresses to muscle degeneration and the cutaneous involvements. The skin manifestations include helio-trope-like colored erythema and swelling on the eye-lids, cheeks, neck and upper extremities of the sun-exposed areas and Gottron’s papules on the dorsa of the hand fingers (3). Although the etiology of DM remains unknown, internal malignant disorders including lung and/or other organ cancers are frequently associated. Generally DM is classified as shown in Table 3 (9).

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<p><b>Congenital myopathies:</b> characterized by developmental delays in motor skills; skeletal and facial abnormalities are occasionally evident at birth</p> <p><b>Muscular dystrophies:</b> caused by progressive weakness in voluntary muscles; sometimes evident at birth</p> <p><b>Mitochondrial myopathies:</b> caused by genetic abnormalities in mitochondria, cellular structures that control energy; include Kearns-Sayre syndrome,</p> <p><b>Glycogen storage diseases of muscle:</b> caused by mutations in genes controlling enzymes that metabolize glycogen and glucose (blood sugar); include Pompe's, Anderson's and Cori's disease</p> <p><b>Myoglobulinurias:</b> caused by disorders in the metabolism of a fuel (myoglobin) necessary for muscle work; include McArdle, Tarui and DiMauro diseases</p> <p><b>Dermatomyositis:</b> an inflammatory myopathy and skin lesions</p> <p><b>Myositis ossificans:</b> characterized by bone growing in muscle tissue</p> <p><b>Familial periodic paralysis:</b> characterized by episodes of weakness in arms and legs</p> <p><b>Polymyositis inclusion body myositis and related myopathies:</b> inflammatory myopathies of skeletal muscles</p> <p><b>Neuromyotonia:</b> characterized by alternating episodes of twitching and stiffness</p> <p><b>Stiff-man syndrome:</b> characterized by episodes of rigidity and reflex spasms</p> <p><b>Common muscular cramps and stiffness and tetany:</b> characterized by prolonged spasms of the arms and legs</p>
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(National Institute of Neurological Disorders and Stroke, National Institutes of Health, USA<sup>1</sup>)

Table 1. Classification of myopathy

Patient	Gender	Age years	Disease	Enzyme	Muscle biopsy	Auto-antibody	Observation term	Complication	Outcome
1.	F	37	pSjS	No abnormalities	myopathy	ANA nv SSA 61.4 SSB 21.6	3.0 y	no	remission
2.	M	32	pSjS	CK 147 ALD 5.8	myopathy	ANA 20 SSB 15.7	8 m	no	remission
3.	F	29	DM	CK 883 ALD 11.2	myositis	nv	2.6 y	no	remission
4.	F	42	DM	CK 212 KL-6 2485	nd	nv	4.3 y	intestinal pneumonia	remission
5.	M	45	DM	CK 302 ALD 8.4	myositis	nv	3.8 y	no	remission
6.	F	55	DM	SLD 5.7	nd	nv	3.11 y	no	remission
7.	F	60	DM	CK 364	nd	nv	2.0 y	no	remission
8.	M	66	DM	CK 1706 ALD 8.2 KL-6 1876 myoglobin 29.7	myositis	ANA 20	2.0 y	pneumonia and cancer	death

ALD: aldolase, ANA: anti-nuclear antibody, CK: creatine kinase, DM: dermatomyositis, F: female, M: male, nd: not done, Nv: negative, pSjS: primary Sjögren's syndrome, SSA: anti-SjS A antibody, SSB: anti-SjS B antibody  
Myopathy: non-inflammatory myopathy, Myositis: inflammatory myopathy

Table 2. Myopathy and myositis in primary Sjögren's syndrome (pSjS) and dermatomyositis (DM)

- **Dermatomyositis**
- Without muscle weakness (amyopathic dermatomyositis or dermatomyositis sine myositis)
- With muscle weakness
- Adult
- associated with cancer
- not associated with cancer
- Pediatric
- **Polymyositis**
- Adult
- Pediatric
- Inclusion-body myositis
- Overlap (myositis associated with a connective tissue disease)

Drake LA, et al<sup>9</sup>: Guidelines of care of dermatomyositis. J Am Acad Dermatol 1996; 34: 824

Table 3. Classification of Dermatomyositis/ Polymyositis

## 2. Cases

**pSjS:** A 37-year-old woman (Case 1 in Table 2) was suffered from fever around 37.5°C, fatigue, proximal muscle pain and weakness in her limbs and arthralgia since a week before her visiting our hospital. She presented herself with swelling and helio-trope-like colored erythema on the eye-lids (Fig. 1a,b), purpurish erythema-spots on the elbows, thin-reddish erythema patches on the legs (Fig.1c) and red palms. On the dorsa of the hand-fingers, the eruption looked like Gottron’s papules was seen and thin purpuric spots were also noted on the paronychia areas. The electromyography showed low amplitude motor units (less than 1 mV) from muscles of the upper extremities that were suggestive myopathy. We suspected the patient had DM and the skin biopsy was taken from the erythematous patches on the left leg. The histology revealed so-called “lymphocytic vasculitis” which showed swollen



a)



b)



c)

Fig. 1. a,b) Close-up view of the right upper eye-lid showing slightly swollen and heliotrope-like colored erythema (Case 1 in Table 2). c) Gottron's nodule-like eruption on the dorsa of the fingers and thin-reddish erythematous patches on the lower legs.

vascular walls surrounded by mainly monocytes and a few of neutrophils in the middle and deep dermis (Fig. 2a,b). No deposit of IgG, IgA, IgM and C3 was seen at the dermo-epidermal (D-E) junction and vascular walls in the dermis by immunofluorescent microscopy. The immunochemical histology revealed CD4<sup>+</sup>>CD8<sup>+</sup>>CD56<sup>+</sup> cells distributed around vessels in the deep dermis. A muscle biopsy was performed from the biceps muscle of the left-upper arm where the patient was complaining of pain. No features of inflammation associated the “muscle fiber degeneration” could be found though slight vascular infiltration was seen in the interstitial tissue.

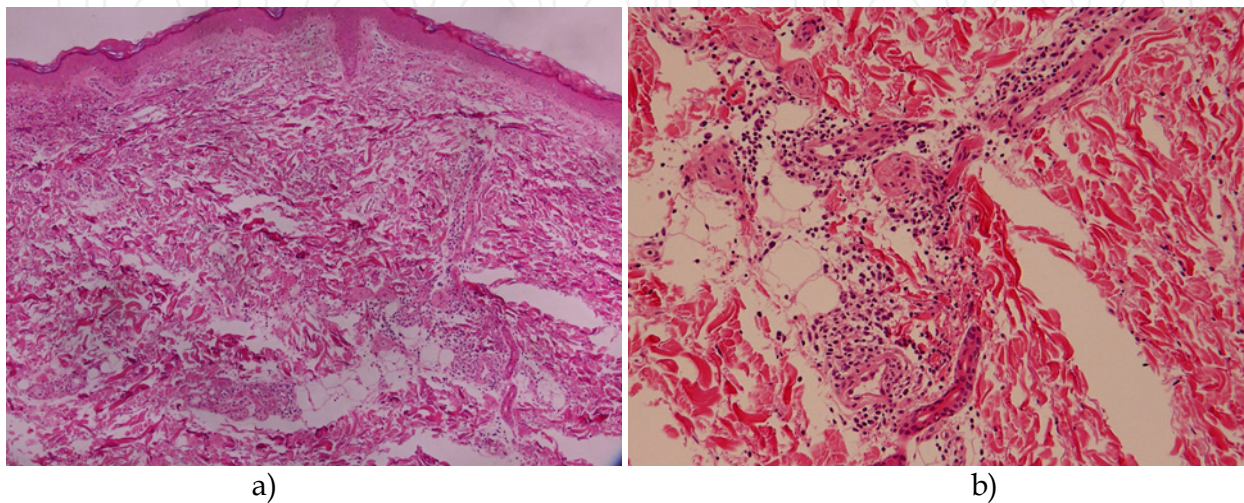


Fig. 2. a) A skin biopsy from the left side of the lower leg revealed, a. vascular infiltration with lymphoid cells and a few neutrophils in the middle and deep dermis (H&E stain, x40). b) The magnified view showed swelling of the vascular endothelial cells with slight degeneration infiltrated by lymphoid cells, suggesting so-called “lymphocytic vasculitis” (x 200)

Laboratory examinations revealed within normal-limit (WNL) ranges of white blood cells (WBC), red blood cells (RBC) and serum AST (aspartate aminotransferase), ALT (alanine aminotransferase), and CK (creatin kinase) (28 IU/L, WNL: 48~259) and aldolase (ALD) (2.2 IU/L, WNL: 2.5~5.6) were rather lower than WNL. The serum levels of complement were high in CH50 (58.4 u/ml, WNL: 31.0~48.0) but levels of C3 and C4 were WNL. Although auto-antibodies (auto-Abs) including anti-nuclear Ab (ANA), anti-Jo-1, anti-DNA and anti-acetylcholine receptor Abs and RA factor were negative, anti-Sjögren’s syndrome A (anti-SSA) and anti-SSB Abs showed the titers of 61.4 and 21.6 (WNL: less than 1.0 ), respectively, which were highly positive in detection by ELISA. Serum carcinoembryonic antigen (CEA) was not detected. Urinalysis revealed no abnormalities except detection of acetone body. The chest X-ray and positron emission tomography (PET) with <sup>18</sup>FDG (2-deoxy-2-fluoro-D-glucose) were performed but no abnormal uptakes were detected except for an enlarged lymphnode at the right-side neck and slight enlargement of the liver and spleen. Ophthalmological examinations revealed positive Schirmer’s test (10mm/6mm) and fluorescein test which showed dry-eyes, although the amount of saliva was 3.5 ml/10 min which seemed to be low But WNL. The pain of visual analog scale (VAS) score was 75 mm when she initially visited our clinic.

The patient was diagnosed as having pSjS with myalgia mimicking DM, which may be classified as the extraglandular type. She was treated with prednisolone (PSL) 20mg/day and non-steroidal anti-inflammatory drugs (NSAIDs). The symptoms including the

cutaneous manifestations completely disappeared and pain VAS score also gradually decreased to 55 mm in a week. The patient was quickly recovered from fatigue, subfebrile state, myalgia and arthralgia two weeks after treatment by oral steroid. However, PSL administration of within 10 mg/day was needed to keep her well condition, although more than two years have been passed since her first visit.

Although the other 32 year-old male patient (Case 2) complained of dry eyes, mild fever, myalgia and muscle weakness of the upper extremities for more than one year, we initially suspected DM and examined for possibility of his having SjS regardless of absence of a DM-like eruption. Ophthalmologically he was suggested to have SjS. The blood examination revealed ALT 40 U/L (WNL: 6-36), ALD 5.8 IU/L, titers of ANA 20x (speckled type) and anti-SSA Ab 15.7 which were relatively high. However, a biopsy from the biceps muscle of the left upper arm was free from inflammation, and no internal malignancy was associated through examinations including PET with  $^{18}\text{F}$ FDG. He has been followed as pSjS similarly to the Case 1 for the duration of more than half a year.

**DM:** A 29 year-old female (Case 3) visited our clinic for helio-trope-like eruptions on the sun-exposed areas including upper eye-lids, cheeks and V-neck area (Fig. 3a), swollen fingers with periungual hemorrhage of the hands (Fig. 3b) and myalgia of the upper extremities. The examination revealed a rise of AST and ALT (65 and 46 U/L; WNL: 35-11 and 39-6, respectively), CK 883 U/L and ADL 11.2 IU/L, but no auto-Abs including ANA, anti-SSA and SSB Abs were found. No internal malignancies were found. However, a biopsy from left biceps muscle revealed the typical myositis with interstitial vascular infiltration (Fig. 4a). The immunochemistry of infiltrated cells around the interstitial vessels of the muscle tissue revealed  $\text{CD4}^+$   $\text{CD8}^+$   $\text{CD68}^+$  cells and little of  $\text{CD20}^+$  cells as similarly seen in the cutaneous findings of Case 1 (Fig.4b). We made a diagnosis of early stage of DM.



Fig. 3. a) A 29 year-old female with dermatomyositis (DM) (Case 3). Swollen helio-trope-like erythema on the upper eye-lids and cheeks. b) Swollen erythema on the dorsa of the hands and periungual hemorrhage of the fingers.

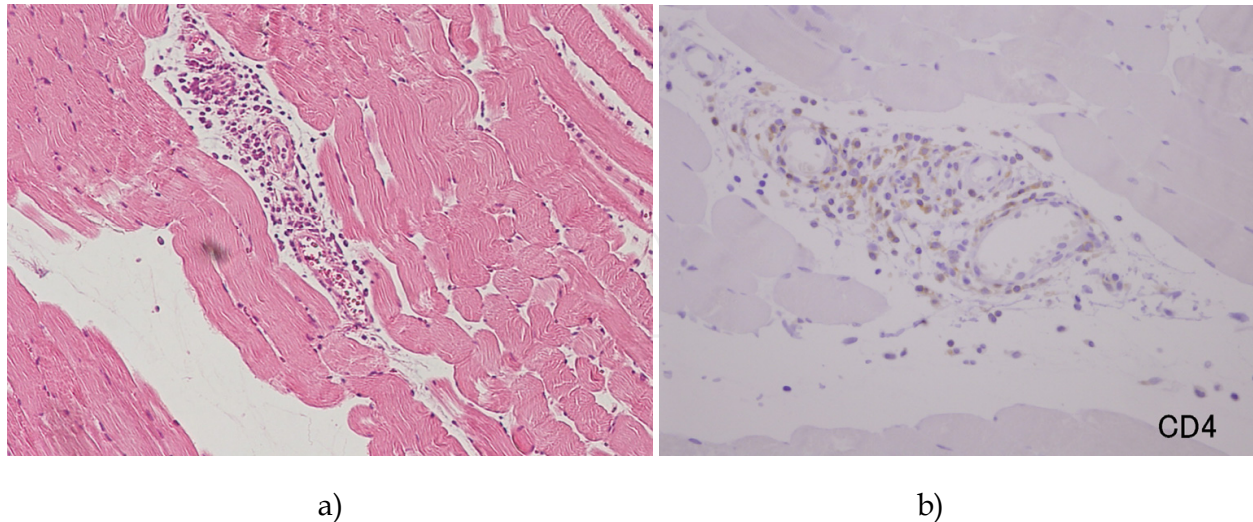


Fig. 4. a) A biopsy specimen of muscle tissue from the left biceps muscle of patient with dermatomyositis (DM) (Case 3). Lymphoid cell infiltration around the vessels was found in the interstitial tissue (HE, 200x). b) Immunohistology of CD4<sup>+</sup>T cells in the interstitial perivascular infiltration of the biceps muscle (Avidin-biotin stain, 20x). The infiltrated cells are CD4<sup>+</sup>>CD8<sup>+</sup>>CD56<sup>+</sup> mononuclear cells.

A 66 years old man (Case 8) was referred to our clinic for sudden episode of helio-trope-like colored erythematous eruption on the sun-exposed areas including the face, V-neck area, upper back and upper extremities associated with “myopathy” exhibiting muscle weakness and myalgia (Fig. 5). The patient had heavily smoked cigarettes a pack or more a day. He was initially suspected to have photosensitive dermatitis due to some photosensitizer and/ or DM and the examinations including skin and muscle biopsy from left-upper arm were performed. Laboratory examinations revealed WBC 11,070 /  $\mu$ l, RBC 327  $\times$  10<sup>4</sup> /  $\mu$ l, CRP 14.28 mg/dL (WNL: 0.30), WNL of serum transaminases (AST and ALT), high levels of CK 1,706 U/L (WNL: 259-2.5), ALD 8.2 IU/L (WNL: 5.6-2.5), myoglobin-U 29.7 ng/mg (WNL: 10~0) and KL-6 1876 U/mL (WNL: 499-0). The titers of auto-Abs showed WNL as to anti-DNA, anti-Jo-1, anti-RNP, Sm, and anti-SSA and anti-SSB Abs except for ANA 20x (speckled type). Although CEA, CA15-3, AFP-L3, -L2, -L2, 3 and CA602 were negative, we suspected the patient might have an association with lung carcinoma after the chest X-ray and CT examination. The skin histology of the helio-trope-like erythema revealed as SLE-like findings exhibiting liquefaction degeneration of the D-E junction and edema of the upper dermis with a little lymphoid cell infiltration (Fig. 6) and immunohistologically, IgG, IgM and complement C3 were linearly deposited at the D-E junction. The muscle biopsy from the biceps of the left arm exhibiting myalgia shows a tiny interstitial perivascular infiltration between the muscle bundles, suggesting “myositis”, although obvious muscular-degeneration was not found. The symptoms of the skin rash and myalgia of the extremities were improved temporary after treatment with oral PSL and NSAIDs. However, the patient died by lung cancer 2 years after his first visit to our clinic.





Fig. 5. A 66 years old patient with DM (Case 8) associated with lung cancer. Helio-trope-like erythema can be seen on the face and upper breast (so-called sun exposed areas).

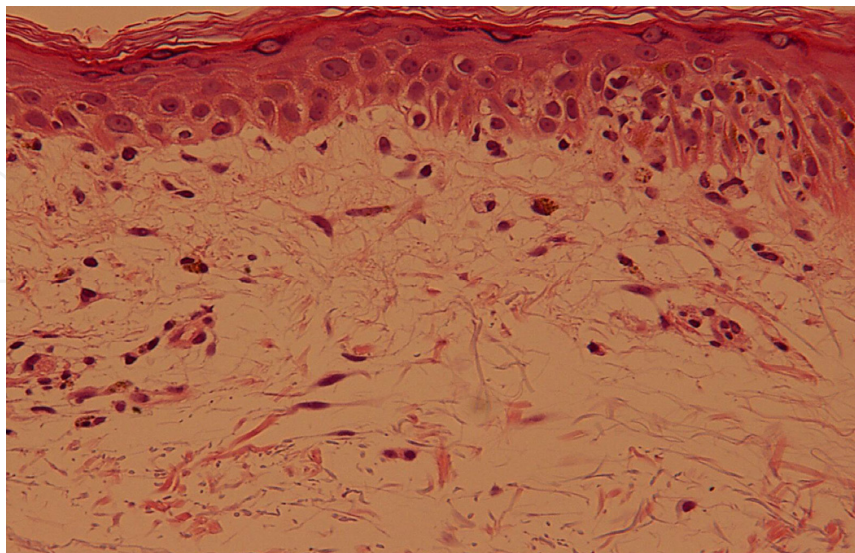


Fig. 6. A biopsy specimen of the helio-trope-like erythema from the upper chest. Liquefaction degeneration can be seen at the D-E junction and edema and a few lymphoid cell infiltration are present in the upper dermis (HE, 200x).

### 3. Discussion

It is rare to see the cases of pSjS with myalgia in Japan, but about 30 % of pSjS patients are reported to be associated with muscle involvement known as fibromyalgia in US and European countries (5, 6). The main cutaneous involvement is purpura, annular shaped erythema and/or macules, erythematous papules and ischemic ulcers due to microvasculitis in pSjS (7, 8). In this patient (Case 1 in Table 2), the cutaneous eruptions including the swollen and helio-trope-like colored erythema on the eye-lids, thin-reddish macules on the limbs, purpuric spots and Gottron's papule-like eruption on the dorsa of the fingers were recognized and quickly disappeared after administration with low dose of PSL. Although the clinical signs-like DM did not reappear by the treatment, the continuous treatment with PSL seems to be still needed. As reported that this disorder is a bothersome and slowly progressive disease (10-12), we should follow the clinical course whether the patient might develop lymphoproliferative disorders in a near future because the enlargement of the lymphnode and hepatosplenomegaly was noted initially. Regarding Case 2, the clinical symptoms and laboratory examination were suggestive of DM associated with SjS without the skin manifestations. We considered him as having pSjS associated with myopathy because dry-eye symptoms, positive anti-SSA Ab and no cutaneous symptom of DM were noted. Although these 2 cases of pSjS were associated with myalgia, the cause of their myopathy is not clear because no inflammation was found in the biopsy specimens from their biceps muscle. However, it is reported that the myopathy might be due to small-vessel injury by auto-Abs or circulating immune complexes because electrondence deposits were noted (13). As to the infiltrated cells around the vessels in the cutaneous lesion, the CD4<sup>+</sup> T cells were dominant as similar to the findings in myositis of patients with DM.

DM is an idiopathic inflammatory disorder characterized by inflammatory myopathy, indicating myositis, and skin manifestations and it can be associated with the secondary SjS sometimes. The etiology is still unknown and the prevalence is estimated as 1-10 cases per million population, but in children 3.2 cases per million which are distributed in the whole world (3). The clinical types of DM are classified in Table 3 and the internal malignancies are frequently associated in the adult type of DM. The risk is reported to be a 6.5-fold higher than ordinary persons after 45 years of age (14). Regarding myopathy in DM patients it might be characterized by inflammatory myopathy progressing to myositis and degeneration of muscle fibers, and the helio-trope-like eruptions on the sun-exposed areas showed SLE-like changes histologically. On the other hand, there are the cases associated without myogenic symptoms in DM, which is called as "amyopathic DM" (DM sine myositis). However, these cases should be considered as "pre-myopathic DM" because they might be rather early diagnosed (15). Though the direct cause of myopathy is still unknown, there are some pathogeneses reported, such as the presences of myositis-specific auto-Abs (15,16) and inflammatory cytokines from T cells including interleukin (IL)-17 and IL-23 in early stage of patients with DM (18). The study regarding Th1/Th2 balance showed that Th2 cell predominance was suggested in patients with active stage of DM (19). In plasma of patients with DM and/or polymyositis, microparticles derived from CD14<sup>+</sup> mononuclear cells, CD3<sup>+</sup> T cells and CD19<sup>+</sup> B cells were found to be elevated by electron microscopic examination, which suggests these diseases were immunological disorders (20). It is reported that CD19<sup>+</sup>CD23<sup>+</sup> cells are increased and that CD4<sup>+</sup>CD45RO<sup>+</sup> cells are decreased in the peripheral blood of patients with DM, suggesting reduction of regulatory T cells (21). It

was also suggested that CD4<sup>+</sup>CD25<sup>+</sup> cells, forkhead/winged helix transcription factor (Fox P3)<sup>+</sup>, transforming growth factor<sup>+</sup> and IL-10<sup>+</sup> cells were reduced in peripheral blood of patients with DM (21). Actually, CD4<sup>+</sup>CD8<sup>+</sup> cells were significantly distributed around vessels in the interstitial tissues of the muscle bundles in our patients with DM. These cells might be CD4<sup>+</sup>CD 28<sup>+</sup> (null) cells and CD8<sup>+</sup>CD28<sup>+</sup> (null) cell infiltration, as reported, and it is of interest that circulating CD4<sup>+</sup>CD28<sup>+</sup> cells and CD8<sup>+</sup>CD28<sup>+</sup> cells were significantly increased in seropositive individuals, responded to human cytomegalovirus antigen stimulation and correlated with disease duration (22).

As to treatment for myopathy of patients with pSjS and DM, adequate doses of NSAIDs and/or oral steroids were mainly used in corresponding to their clinical severities and these were considered to be effective. However, in addition to these drugs, the combination with immunosuppressive agents such as azathioprine, cyclosporine, mycophenolate or methotrexate should be used for the autoimmune diseases, if they were not clinically controlled. The biological agent, rituximab, and tacrolimus may offer additional benefit to some patients and emerging agents against T cells, B cells, transmigration or transduction molecules may be discussed as New treatments (23).

#### 4. Conclusion

Myopathies are neuromuscular disorders exhibiting myalgia and muscular weakness due to dysfunction of muscle fibers which are frequently seen in autoimmune diseases. We here discussed about the non-inflammatory myopathy seen in patients with pSjS and the inflammatory myopathy, that is myositis, found in patients with DM is suggested to be Immunological dysfunction in pathogenesis. Although the mechanism of the myopathy is still unclear, it might be due to inflammatory cytokines released from CD4<sup>+</sup>CD 28<sup>+</sup> (null) cells and CD8<sup>+</sup>CD28<sup>+</sup> (null) cells infiltrated around vessels in the muscles of patients with MD.

#### 5. Abbreviations

Ab, antibody; ALD, aldolase; ANA, anti-nuclear antibody; anti-SSA Ab, anti- Sjögren's syndrome A Ab; anti-SSB Ab, anti- Sjögren's syndrome B Ab; CK, creatine kinase; D-E junction, dermo-epidermal junction; DM, dermatomyositis; IL, interleukin; NSAID, non-steroidal anti-inflammatory drug; pSjS, primary Sjögren's syndrome; PSL, prednisolone; VAS, visual analog scale; WNL, within normal limit

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