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Editor's Message

Welcome to this issue of CHARM on inflammatory myositis. You may remember that one popular movie actress passed away earlier in 2017 because of this condition. In this issue, we are glad to have all-rounded contributions from rheumatologists, speech therapist, physiotherapist and nursing specialist. Dr Sung Chi Keung has comprehensively written on the diagnosis and the various types of inflammatory myositis. Dr Leung Moon Ho educates us on the risk of malignancy associated with inflammatory myositis. Mr Dennis Cheng Tze Ho, speech therapist, shares his professional knowledge in the evaluation and management of dysphagia presented by some sufferers. Dr Arnold Wong, physiotherapist, enlightens us the best exercise training for these patients. Ms Yip Ka Huen and Ms Siu Mei Yi described fall prevention and nursing care for these patients.

Happy reading!

Inflammatory myositis diagnosis

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Introduction

Inflammatory myopathy can be caused by drug, infection, toxic substance and rheumatic disease. Idiopathic inflammatory myopathy refers to polymyositis, dermatomyositis and inclusion body myositis. We now believe that autoimmunity is the underlying abnormality of polymyositis (PM)and dermatomyosits (DM). As a result, autoimmune inflammatory myopathy may be a better term to describe these two distinct diseases. What's more, muscle may not be the only organ involved and the degree of muscle involvement can vary a lot. Together with the identification of more and more myositis-specific antibodies (MSAs) and myositis-associated antibodies (MAAs), the diagnosis and classification of autoimmune inflammatory myopathy is changing.

Disease Spectrum Or Classification

In 1975, Bohan & Petter laid down the diagnostic criteria of PM and DM. The only distinction between these two conditions was the skin involvement in DM. Later it was found that the immunopathology of PM and DM was also different [1].

Apart from pure PM and DM, we can now recognize more clinical variations and relate these phenotypes to different MSAs and MAAs.

Inflammatory myositis diagnosis

Pure myositis

Pure or classical PM

It refers to a group of patients suffer from pure primary muscle inflammation. They present mainly with insidious onset of proximal weakness. Characteristic histopathology of the involved muscle is expected. The risk of interstitial lung disease (ILD) and cancer should be low. There is no known associated autoantibody.

Immune medicated nectrotizing myopathy IMNM

IMNM is a clinical entity similar to PM, but with a more severe muscle damage. The serum levels of muscle enzymes are very high. As the name suggests, biopsy show necrotic muscle fibres. MSA against single recognition particle (SRP) or 3-hydroxy-3-methylglutaryl co-enzyme A reductase (HMGCR) can be detected in ~60% of patient with IMNM [2]. The response to immunosuppression therapy is usually good.

Overlap myositis

Anti-synthetase syndrome

Apart from proximal weakness, this group of patients may also have raynaud's phenomenon, arthritis, and mechanics hands. Anti-synthetase antibodies are the MSAs for this clinical entity. Anti-histidyl-tRNA synthetase (Jo1) antibody is the most common type. Other anti-synthetase antibodies include anti-PL7, PL12, EJ,OJ, KS, Zo. ILD can be found frequently and presence of anti-synthetase antibodies carries a better prognosis [3].

Mixed connective tissue disease (MCTD)

Myositis may be a part of an overlap syndrome with systemic lupus erythematosus and scleroderma. Anti- U1 RNP can be found in this group of patient. Common presenting symptoms are raynaud's phenomenon and joint pain.

Dermatomyositis related diseases

Classical DM

A typical case of DM presents with insidious onset of symmetric proximal muscle weakness together with skin involvements like heliotrophic rash, periorbital edema, gottron papules at knuckles and extensor surfaces. Serum creatine kinase is high. Usually there is no other organ involvement and the risk of cancer association is low. This clinical phenotype is associated with anti-helicase (Mi2) antibody [4].

Amyopathic DM

In approximately 5-20% of patients with typical DM skin lesions, but there is no obvious weakness and no elevated muscle enzymes to suggest significant muscle involvement. In addition to the classical DM rashes, they may have ulcers, digital necrosis and mechanics hands. Arthralgia and arthritis are also common. Rapid and potentially fatal ILD is a serious complication, especially in our locality. Anti-MDA5 can be detected in 20-50% of Asian DM patients and is associated with relatively lower creatine kinase levels, a high frequency of ILD (90-95%), especially rapidly progressive ILD (50-80%), and a poor prognosis due to respiratory failure [5].

Cancer associated DM

Approximately 15% of of all DM cases are tumour-associated [1]. ILD appears to be protective against cancer. Anti-TIF1γis particularly related to the cancer development, negative predictive value can be up to 95% [6].

Anti-small ubiquitin-like modifier activating enzyme (SAE) associated DM

Patients with anti-SAE Ab present with classical DM rash. Muscle symptoms usually occur few months later and many of them have dysphagia. Associated ILD is uncommon and mild, but cancer may be found.

DIAGNOSTIC APPROACH

Symmetric proximal weakness is the classical symptom of autoimmune myositis. Those patients with have a waddling gait and physical examination will show a positive gower's sign. Presence of skin rash and associated symptoms are important in guiding further investigations for both diagnosis and complications screening.

Muscle weakness only

For those patients present with predominant muscle weakness, the next step is to confirm myositis is the underlying cause of the weakness with three laboratory examinations: serum muscle enzymes concentration, needle electromyography (EMG), and muscle biopsy. Active myositis will give a high serum creatine kinase. EMG shows myopathic changes like spontaneous activity with fibrillations, complex repetitive discharges, and positive sharp waves. Typical muscle biopsy of PM will reveal a multifocal lymphocytic infiltrates (CD8-positive cells) surround and invade healthy muscle fibres (expressing MHC class I antigens). Demonstration of inflammation distinguishes polymyositis from toxic or dystrophic myopathy. Lack of vacuolated muscle fibre rule out inclusion body myositis. Autoantibody screening will usually be negative in a pure care of PM.

If muscle biopsy show extensive necroses suggesting IMNM, then it is worthwhile to check anti-SRP or anti-HMBCR.

Inflammatory myositis diagnosis

Weakness with associated symptoms

If weakness is prominent and CK is high, then workup for myositis as described above is also applicable. When there is associated symptom, screening for anti-synthetase antibodies and MAAs (anti-Ro, La, RNP, PmScl, Ku) aids in classification. For those patients with raynaud's phenomenon, arthritis and mechanic hands, anti-synthetase antibodies will likely be found. If there are features of lupus or scleroderma, then positive anti-RNP suggest a diagnosis of MCTD. Patient should also be screened for ILD.

Skin involvement with or without weakness suggesting dermatomyositis

Typical rashes of DM are characteristics. Skin biopsy is usually not required and there is no pathognomonic histopathology. Usual findings are vacuolar degeneration of epidermal basal cells with epidermal atrophy and dermatitis with lymphocytes and macrophages. In doubtful cases, skin histopathology distinguishes DM from other papulosquamous diseases but not from cutaneous lupus. Ramified-giant capillary is a characteristic finding in naildfold capillaroscopy [7]. Microscopic features of muscle biopsy are perivascular, perifascicular or perimysial inflammation and later lead to perifasciculus atrophy. The primary target is the endothelium of the endomysial capillaries and the predominant lymphocytes are B cells and CD4 T cells.

Screening for ILD and cancer is recommended after a diagnosis of dermatomyositis.

Presence of MSSs further supports the diagnosis of DM and helps to predict the clinical course. Anti-Mi2 carries a better prognosis. The response to treatment is better and the risk of ILD and cancer is lower. On the other hand, patients in our locality with anti-MDA5 carry a high mortality due to a high chance of rapidly progressive pneumonitis. We have to look hard for cancer especially in those patients with anti-TIF1γ, anti-NXP2 or anti-SAE.

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Double Trouble – Malignancy is commoner in Inflammatory Myositis

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Idiopathic Inflammatory Myositis + Malignancy

This association was recognized more than a century ago. In particular, within idiopathic inflammatory myositis (IIM), patients with dermatomyositis (DM) have a higher chance of having malignancy than patients with polymyositis (PM). The association is evident in the following ways. Epidemiological studies showed that IIM patients have a higher chance of having malignancy diagnosed than comparable general population. Malignancy is likely to be diagnosed at a time close to the diagnosis of IIM. When malignancy is treated, IIM tends to be improving or even resolving. On the other hand, when IIM relapses, there is a chance malignancy is relapsing too.

Evidence for the double trouble

Epidemiological studies usually use the "Standardized Incidence Ratio" (SIR) technique to ascertain if there is a genuine relation between malignancy and IIM, and if so, how much the risk is. SIR is the ratio of observed number malignancy to the expected number of malignancy, adjusted in that population with respect to variables such as ethnicity, gender and age group, because the baseline malignancy rates could be different. It would be important to understand that, for an average Hong Kong citizen, the lifetime background (expected) risk of having malignancy diagnosed is more than one in three. In Western series, SIR figures ranged from 3 to 6. There is no formal population study in Hong Kong yet. A Taiwan study looked at their National Health Insurance Database from 1997 to 2007 that included 1,012 DM and 643 PM patients. Among DM patients, 9.4% of them developed malignancy (SIR = 5.1), whereas among PM patients, 4.4% developed malignancy (SIR = 2.2).

Within IIM, DM is commoner than PM in tropical or sub-tropical areas which receive more intense and longer duration of ultraviolet irradiation. Therefore in Hong Kong, we should be having more DM proportion among IIM patients, and as the risk of having malignancy is higher in DM compared to PM, we should be even more vigilant in excluding malignancy in our DM patients.

Double Trouble – Malignancy is commoner in Inflammatory Myositis

OK, more malignancy. Which types then?

The types of malignancies associated with IIM usually follow the local epidemiology of malignancies. In 2014, among 29,618 newly diagnosed malignancies in Hong Kong, the top three diagnoses were colorectum (18.8%), lung (15.8%) and breast (13.1%). Other malignancies are also commonly reported to be associated with IIM, e.g. cervical, ovaries, pancreas, bladder and stomach cancers. One important note is that nasopharyngeal carcinoma (NPC) is common in southern China and there are many reports to highlight the association of NPC and DM in Hong Kong and neighbouring regions. In a ten-year retrospective study of 246 DM patients in Guangzhou, China, 24.4% of them were diagnosed malignancies of the following types: NPC (35%), ovarian cancer (15%), lung (8%) and colon (8%).

When is malignancy usually diagnosed?

Malignancy can be diagnosed shortly before, at the same time with, or after the diagnosis of IIM. Overall, malignancy is most likely to be diagnosed at the same time with or within the first year of diagnosis, and the chance would be gradually coming down in the subsequent five years of follow-up. There is always a chance of late malignancy development, but it is necessary to take this in the light of the background cancer rate in the general population.

Which IIM patients are more likely to develop malignancy?

DM patients are more likely than PM patients. Other clinical features that may be associated with increasing risks are: older age of IIM onset, swallowing difficulty, skin necrosis, cutaneous leucocytoclastic vasculitis and those having muscle biopsy showing capillary damage. On the contrary, interstitial lung disease is negatively associated with malignancy. That is, IIM patients having co-existing interstitial lung disease have lower chance of being diagnosed malignancy. (However the prognoses of both rapidly progressive interstitial lung disease and malignancy in IIM are remarkably poor). Patients with certain myositis specific auto-antibodies are at higher risk of developing malignancy, for example anti-TIF1γ (transcription intermediary factor) and anti-NXP2 (nuclear matrix protein). Myositis specific auto-antibodies testing may be available in some special centres and some private laboratories.

How should we screen for malignancy in IIM patients?

IIM patients should be evaluated for the potential of underlying malignancy when they are newly diagnosed and when disease relapses after initial stabilization. The strategy should be symptoms and signs directed against common malignancies according to local epidemiology. Basic investigations would be useful, for example: complete blood picture, liver function tests (note that liver enzymes [like ALT and LDH] elevation might be indeed be muscle origin), urinalysis, stool occult blood and chest radiograph. Those screening strategies applicable to general population according to local guideline should be considered, for example age- and gender-specific colonoscopy, Pap smear and mammogram. In Hong Kong, it is prudent that these patients are examined by endoscopy by Ear, Nose, Throat surgeons to exclude NPC, which has the highest SIR of malignancy in many southern Chinese series. It is a common practice to offer PET/CT (positron emission tomography with computed tomography) to these patients as a rapid test for searching underlying malignancy. However PET/CT does have its false negative (not picking up cancer, like ovarian cancer) and false positive rates (inflammatory lesions with high uptake can mimic malignancy), and its role in IIM patients is yet to be fully defined.

Summary

IIM patients are at higher risk of developing malignancy, especially at the time of diagnosis and within the first year. In Hong Kong, the types of malignancies usually encountered are NPC, lung, colorectal and breast cancer. DM patients, older patients, patients with skin necrosis, with anti-TIF1δ and anti-NXP2 myositis specific auto-antibodies, and those without interstitial lung disease are at even higher risks. IIM patients should be screened as clinically directly and sometimes with PET/CT scan.

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Inflammatory Myositis and Dysphagia

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Dysphagia in Inflammatory Myositis

The swallowing process is divided into oral preparatory, oral, pharyngeal and esophageal phases. Patients with inflammatory myositis (IM), including inclusion body myositis (IBM), polymyositis (PM) and dermatomyositis (DM), often suffer from dysphagia at the oral and pharyngeal phases. Symptoms include choking, food getting stuck in the throat and multiple swallows [1]. The manifestation of severe dysphagia includes malnutrition, dehydration and aspiration pneumonia, resulting in possible need for non-oral feeding. A patient with significant risk of aspiration from oral feeding, as assessed and diagnosed by speech therapist, may be recommended to receive enteral nutrition by means of a naso-gastric (NG) tube or more permanently, a percutaneous endoscopic gastrostomy (PEG) tube. Figure 1 shows the major structures involved in swallowing.

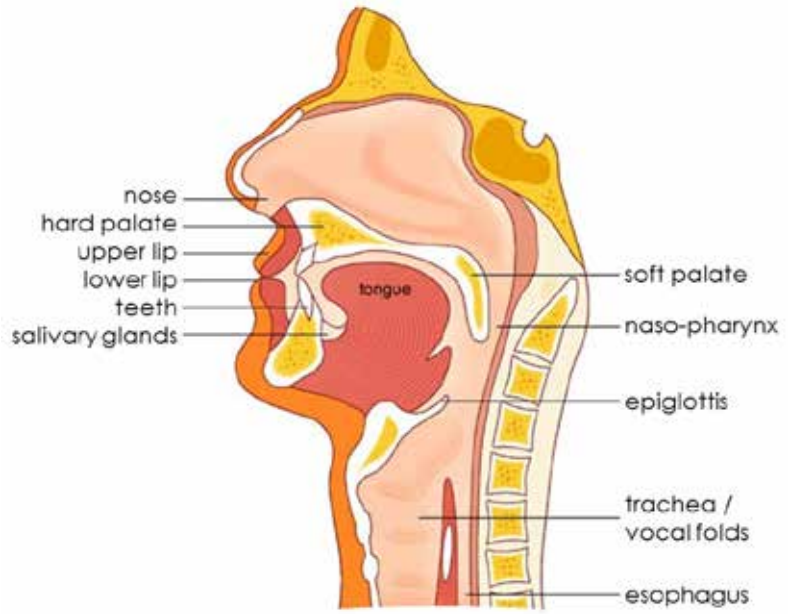

Fig 1a (left). Diagrammatic representation of swallowing anatomy

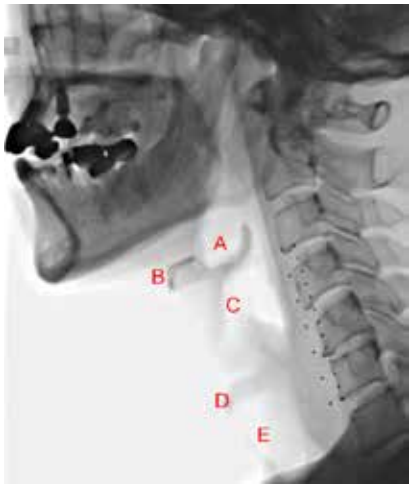

Fig 1b (right).
Videofluoroscopic image showing
A) vallecula; B) hyoid bone; C) laryngeal vestibule (entrance); D) vocal folds; and E) trachea

A retrospective review [2] of 783 patients diagnosed with IM during a 5-year period showed that dysphagia occurred in 62 of them. Among the 62 patients, 26 of them (42%) had IBM, which was considered to have the highest incidence of dysphagia among all types of IM. The mortality rate was high in patients who required percutaneous endoscopic gastrostomy. The main cause of death was respiratory failure due to aspiration pneumonia. In fact, swallowing prognosis in patients with IBM was worse than those with PM or DM, possibly due to its poor response to various forms of medical therapy [3].

Pathophysiology

Dysphagia in patients with IM was traditionally believed to mainly result from impaired opening of the cricopharyngeal (CP) sphincter [4] (also known as upper esophageal sphincter), causing failure of food to go down the esophagus. Recent studies [5,6] argued that the cause of dysphagia in the majority of IM patients was not CP dysfunction, but weakness of the pharyngeal and suprahyoid muscles. The most common videofluoroscopic abnormalities observed were pharyngeal pooling, tongue base weakness, laryngeal penetration, reduced pharyngeal contraction, CP dysfunction, and impaired laryngeal excursion (Figure 2).

Dysphagia is often under-reported by patients with IM. There was a discrepancy in patients' active report of dysphagic symptoms and the presence of swallowing impairments as evaluated by videofluoroscopic examination [1]. Symptoms related to impaired bolus propulsion and aspiration were frequently reported. Two questions were found to reliably predict impaired bolus propulsion: "does food get stuck in your throat?" and "do you have to swallow repeatedly in order to get rid of food?" This highlights the importance to screen for the existence of dysphagic symptoms so that appropriate referral can be timely made.

Inflammatory Myositis and Dysphagia

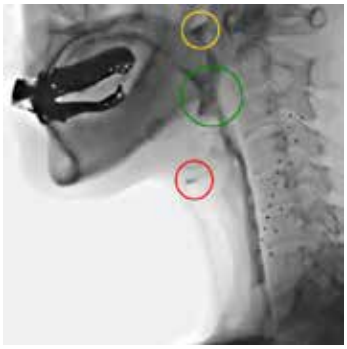

Fig 2a (left). Videofluoroscopic image showing nasal regurgitation (yellow), impaired pharyngeal contraction (green), and tracheal aspiration of food contrast (red)

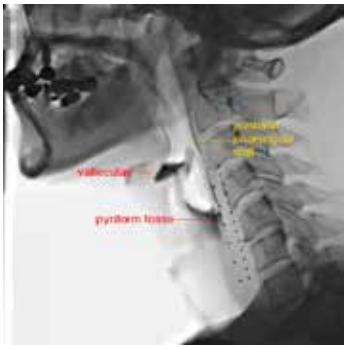

Fig 2b (right). Videofluoroscopic image showing post-swallow pharyngeal wall coating (yellow) and pharyngeal pooling (red) of food contrast

Diagnosing Dysphagia

When a patient with IM is referred to speech therapy, his/her oral motor and swallowing functions will be assessed. Apart from routine bedside swallowing evaluation, two common instrumental examinations may provide speech therapists with objective and detailed information regarding the patient's swallowing functions. They are regarded as the gold standard of instrumental swallowing evaluation.

1. Videofluoroscopic Study of Swallowing (VFSS) (also known as Modified Barium Swallow)

This is an examination using X-ray and a contrast agent (e.g. barium). The patient will be instructed to eat and drink different consistencies of food mixed with contrast. Most swallowing structures and events from the oral prep through the cervical portion of the esophageal phase can be visualized to discern the cause of swallowing problems (Figure 2).

2. Fiberoptic Endoscopic Evaluation of Swallowing (FEES)

This is an examination using endoscopy technique. A flexible laryngoscope will be inserted through the nose to visualize the pharyngeal phase of swallowing and related pharyngeal and laryngeal structures (Figure 3). The patient will be instructed to eat and drink different consistencies of food mixed with food colour for easy identification.

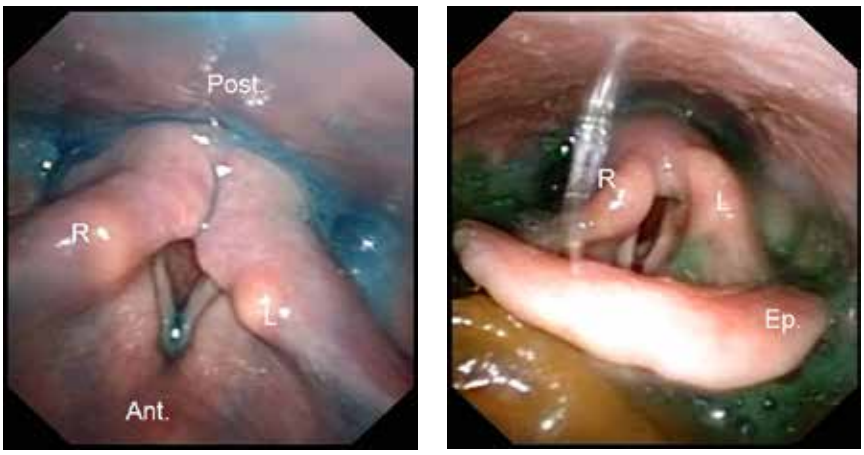

Figure 3. FEES images showing tracheal aspiration of blue liquid through the vocal folds (left) and significant pooling of yellow food in the vallecula and green liquid in the pyriform fossa (right) (FEES - Fiberoptic Endoscopic Evaluation of Swallowing; Ep. - Epiglottis)

Dysphagia Management

Speech therapists provide swallowing rehabilitation by including postural modification, compensatory strategy, diet modification and swallowing training. Table 1 summarizes different kinds of management with examples.

Inflammatory Myositis and Dysphagia

Management	Definition	Example
Postural modification	Modification of feeding posture to facilitate bolus flow and airway protection	Chin tuck Head tilt to stronger side
Compensatory strategy	A technique the patient learns to perform during swallowing to compensate for swallowing impairment	Effortful swallow Double or multiple swallows Supraglottic swallow
Diet modification	Modification of food and drink consistency to prevent aspiration and facilitate swallowing	Thickened fluids Pureed / Minced diet Powdered medications
Swallowing training	Treatment designed for strengthening swallowing muscles	Masako maneuver Mendelsohn maneuver Shaker exercise

Table 1. A summary of dysphagia management for patients with inflammatory myositis

In cases where cricopharyngeal dysfunction is confirmed, interventional procedure or surgery is considered:

1. **Cricopharyngeal myotomy [4,7]:**
   Surgical sectioning of the cricopharyngeus muscle to reduce spasm
2. **Pharyngoesophageal dilation [8]:**
   Fluoroscopy-guided balloon dilation of the CP sphincter to reduce stricture
3. **Botulinum toxin A (BTA) injection [9,10]:**
   Endoscopic injection of BTA into the cricopharyngeus muscle to decrease muscle tone

Conclusion

Dysphagia is a common symptom for patients with inflammatory myositis. The major goal of swallowing intervention is the prevention of aspiration and other dysphagia-associated complications. Timely referral to speech therapist and careful evaluation are important in identifying the etiology of each patient's swallowing problems, hence allowing for specific feeding recommendations as well as management plan.

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Physiotherapy management for Inflammatory myositis

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Introduction

Inflammatory myositis (or called idiopathic inflammatory myopathy, IIM) is an idiopathic inflammation of muscles that have different signs and symptoms. The estimated annual occurrence rate of IIM is 4 to 8 cases/10,000 individual years [1]. Patients with IIM usually experience proximal muscle weakness that may be associated with other organ problems or even malignancies [2]. Progressive IIM may lead to disability, morbidity, and potential death. Although the causes of IIM remain elusive, it is believed to be related to autoimmune problems that cause muscular inflammation. Depending on the clinical presentations, IIM can be classified into: (1) dermatomyositis, (2) necrotizing autoimmune myopathy, (3) anti-synthetase syndrome, (4) polymyositis, and (5) sporadic inclusion body myositis.

Various types of IIM

Dermatomyositis (DM) is characterized by proximal symmetric muscle weakness and associated skin rashes. The rashes may present with a V sign, shawl sign, Gottron papules, and heliotrope rashes over the eyelids with periorbital edema [1]. However, the clinical presentations of patients with DM are very diverse. Patients with amyopathic dermatomyositis only have skin rashes without muscle weakness, while patients with dermatomyositis sine dermatitis may display muscle weakness without skin rashes [2].

Necrotizing autoimmune myositis (NAM) accounts for 20% of IIM. NAM usually has an acute onset of significant proximal symmetric weakness at the early stage. Like DM, the serum creatine kinase (CK) level of patients with NAM usually elevates to more than 50 times of the normal upper limit, indicating muscle damages. Additionally, NAM is characterized by myositis-specific antibodies, 3-hydroxoy-3-methyglutaryl coenzyme A reductase antibodies and anti-signal recognition particles.

Anti-synthetase syndrome includes symmetric proximal muscle weakness and similar biopsy findings as DM. However, they also present with concomitant arthritis, fever, Raynaud's phenomenon, and "mechanic's hands". Approximately 70% of these patients develop interstitial lung disease. About three quarters of them have positive anti-Jo1 myositis-specific antibody in the biopsy finding.

Polymyositis (PM) comprises a wide range of IIM that cannot be classified into the other four subcategories or that have negative myositis-specific antibody. Clinically, these patients demonstrate subacute proximal symmetric muscle weakness without skin rashes. Patients usually have specific muscle histopathology, elevated serum CK levels to 50 times of the normal upper limit in subacute phase. T cell-induced muscle inflammation and degeneration may present in these patients.

Sporadic inclusion body myositis (sIBM) represents a type of muscle degenerative disease with a specific muscle weakness pattern affecting patients aged 50 years or older [2]. Men is more susceptible to sIBM. Clinically, these patients display asymmetric proximal and distal muscle weakness (e.g. finger flexors and quadriceps). Occasionally, patients may experience dysphagia, and weak facial and trunk muscles. While the progression of weakness is slow, it can eventually lead to loss of ambulatory ability. Given the insidious gradual onset of muscle weakness, sIBM remains to be unnoticed until the symptoms become severe. Serum CK may heighten up to 10 times of the normal upper limit [2]. sIBM may be a primary inflammatory or degenerative myopathy, or a cause/consequence of a neoplastic like process [3].

While there is no consensus on the pathophysiological causes of various types of IIM, immunosuppressive drugs, monoclonal antibody and steroid can be prescribed to control symptoms or progression. However, the long-term usage of high doses of immunosuppressants and corticosteroids may cause bone loss, muscular and metabolic injuries [4]. Additionally, the presence of pain, proximal muscle weakness, severe muscle fatigue, and non-suppurative inflammation of skeletal muscle in these patients may affect their activities of daily living and aerobic capacity (Table 1) [2], and heightens their risk of falls.

Physiotherapy management for Inflammatory myositis

Problems associated with idiopathic inflammatory myopathies	Positive effects of exercise
Progressive inflammation of muscles	Reduction of inflammation
Muscle weakness	Improved muscle strength
Muscle atrophy	Muscle hypertrophy
Muscle fatigue and muscle pain	Reduced fatigue and pain
Decreased muscle endurance	Enhanced muscle endurance
Decreased proportion of type 1 fiber	Restore type 1 fiber proportion
Decreased capillary density in muscles	Increased angiogenesis
Compromised quality of life	Improved quality of life

Table 1. Physical deterioration of patients with idiopathic inflammatory myopathies and potential positive impacts of exercise on these patients

Physiotherapy for IIM

Physiotherapy benefits patients with IIM by reducing their pain, maintaining muscle strength, and optimizing physical function and walking balance. Ice therapy and transcutaneous electrical nerve stimulation are prescribed to reduce pain, whereas exercise therapy can improve quality of life (Table 1) [5]. Notably, physical exercises can increase muscle strength and mass, reduce muscle damage, lower serum levels of aldolase and CK, improve blood lipid profile, normalize blood pressure and insulin sensitivity, optimize body composition, and enhance neuromuscular adaptations/controls (Please refer to Pedersen and Saltin for detailed reviews [6]).

In the past, active exercise training was thought to be inappropriate for people with acute IIM because activity might aggravate inflammation of affected muscles [6]. Therefore, active rehabilitation was recommended to commence during inactive phases of disease with or without the supplementation of medication. A randomized controlled trial showed that a 6-week of 30 minutes of aerobic training on a cyclo ergometer thrice a week at 60% of individual's maximal heart rate significantly decreased pain and muscle impairment, and improved muscle function, VO2max, and quality of life [6]. Likewise, a 12-week exercise program involving 15-minute of flexibility and isometric strengthening exercises of upper and lower limbs for 5 days a week demonstrated significant improvement in pain and muscle function of patients with PM/DM. Importantly, these studies did not display signs of muscle inflammation on magnetic resonance imaging, muscle biopsy, or serum CK or aldolase levels.

Recently, more evidence substantiates the benefits of exercises in patients with active muscle inflammation. Hicks et al. found that exercise therapy in patients with PM/DM during the active phase of the disease significantly increased the isometric peak power after a 4-week isometric training at 60% of maximal voluntary contraction. Others showed that a 12-week home exercise program for patients with active PM/DM (i.e. 15 minutes of strength exercises followed by 15 minutes of walking) conducted at 5 times a week significantly reduced fatigue scale index by 12 to 16% and significantly improved SF-36 scores by 25 to 45%. Similarly, Varju et al. revealed that 3 weeks of submaximal endurance resistive and flexibility exercises significantly improved 40% of muscle strength in several upper and lower limb muscle groups in patients with acute exacerbation of PM/DM.

The benefit of long-term exercise training is supported by research. It was noted that a 6-month exercise training significantly improved VO2max, isokinetic strength and functional ability of patients with chronic IIM as compared to non-exercised controls [5]. While the controls showed significant increase in serum CK levels, nothing was observed in the exercised group.

Physiotherapy management for Inflammatory myositis

Conclusions

While structured exercise training has proven to attenuate pain, and improve muscle strength and function of patients with IIM without increasing serum CK levels, there is no consensus on the most effective exercise modality for these patients. Given the heterogeneity of IIM, there is no single effective physiotherapy intervention for these patients. Future studies should investigate psychosocial/physiological benefits of various exercise approaches and the underlying exercise-related adaptations in patients with IIM. Further, the effects of other physiotherapy treatments (e.g. functional electrical stimulation) in patients with IIM should also be investigated.

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Nursing care for patients suffering from inflammatory myositis

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Introduction

Inflammatory myopathy affects all ages and is often difficult to identify an exact cause for the condition. Autoimmune disorder is one of the main causes, leading to inflammation and destruction of large muscle groups. Polymyositis and dermatomyositis are the two commonest conditions of inflammatory myositis, which occur in about one person per 100,000. Twice as many women get these inflammatory diseases compared to men. Patients with inflammatory myopathies show gradual loss of muscle strength, but less commonly report pain in the affected muscles. They may have weakness in the large muscles around the neck, shoulders and hips. Some of them also have breathing problems, or choking while eating. Myopathies are chronic diseases. Treatment regimen mainly involves the use of corticosteroids and immunosuppressive drugs, as well as physical therapy for rehabilitation. Most cases of inflammatory myositis will respond to therapy. It is important to practice good health measures to achieve good treatment outcome.

1. Fall prevention and muscle training

Fall can be a painful experience for patients with myopathies. It may take long period to recover from the injuries sustained. After recovery, some people remain fearful and lack confidence in their abilities to mobilize independently. Physical therapy for muscle and balance reinforcement is an essential treatment component in helping patients with inflammatory myopathies. Fine motor task and resistance exercise training can improve muscle strength and balance. Range-of-motion exercise is helpful to maintain flexibility of joints. In addition, learning proper use of assistive devices, such as frame or walking stick, aid mobility and balance.

Next, it is important to maintain a safe environment for mobilization. Patients and care-givers should identify and raise awareness of environmental hazards, to evaluate the home environment conditions continuously. Adequate lighting, especially night time is essential. Floor should always be kept dry and flat, be clear of clutter to prevent fall accident. Safety measures such as handrails can be installed in bathroom, retain bed /chairs at a proper height to facilitate safe transfer when the patient rises to a standing position. Personal and situational causes such as poor-fitting shoes, or long pants can impede ambulation and increase risk of fall. Patients are advised to wear well-fitting slippers/shoes with non-slip soles and low heels when ambulating. They are also encouraged to avoid prolonged sitting and standing, maintain regular limb and walking exercise for better muscle strength.

2. Prevention on aspiration

Aspiration of food or fluid occurs with decreased strength of muscles involved in mastication. It can be a fatal complication for patients with inflammatory myopathies. After diagnosis, patients are referred to speech therapist to evaluate the strength of mastication muscles and develop specific maneuver to deal with impaired swallowing. To enable safe swallowing, patients are instructed to moisten dry foods, avoid mixing foods of different texture or putting too much food/fluid into the mouth at a time. In certain circumstances, they may need to eat soft or semi-solid foods, or puree as recommended by speech therapist.

All patients and care-givers need to be aware of symptoms of swallowing problems, including coughing, choking, spitting of food, drooling, pocketing of food, wet-sounding voice, double swallowing or major delay in swallowing, etc. If any of these occurs, the patient is directed to stop eating and remove all food from the oral cavity. Some measures can be adopted to reduce aspiration risk. Patients are encouraged in sitting-up position with the head flexed slightly forward during meal, and stay at an upright position for 30 to 45 minutes after meal. Environmental stimuli (e.g. TV, radio) can be eliminated to facilitate better concentration on food ingestion. Talking and laughing is avoided while eating. Patients need sufficient time to masticate and swallow. Meals should not be hurried. Patients should chew entirely, eat gently, and swallow frequently. Lastly, patients should be assessed for nutritional status regularly by recording a food dairy and checking for body weight. Any sign of malnutrition warrants to initiate consultation to a dietician for calorie count and food preferences.

3. Safe precautions on long-term steroids

Corticosteroids are key anti-inflammatory agent in controlling myopathy inflammation. Despite its potent therapeutic effect, long-term corticosteroid therapy may bring about serious adverse effects in multiple body systems. Patient education should be centered around the rationale of drug therapy, the expected adverse effects and reporting to the healthcare provider. Patient are encouraged to carry a medical identification (e.g. card or bracelet) at all times with information regarding medical diagnosis, drug therapy and name of the physician-in-charge.

Patients and care-givers should be alerted for infective complications and impaired wound healing from corticosteroid and immunosuppressive use. Corticosteroids induce hyperglycemia by inhibiting insulin secretion from the pancreas and stimulating gluconeogenesis through synthesis of carbohydrates from lipid and protein sources. Furthermore, corticosteroids reduce gastric mucus production and predispose patients to peptic ulcers. Besides, patient should be cautious about drug interactions. Combination of corticosteroids and non-steroidal anti-inflammatory drugs increases the risk for gastritis and peptic ulcer disease. Administration of corticosteroids with some diuretics can cause hypokalemia.

To reduce undesirable adverse outcomes on corticosteroid therapy, some health practices are recommended. Firstly, patients are advised to avoid aspirin, alcohol and caffeine which may contribute to steroid-induced gastric ulcers. Secondly, a high protein, calcium, and vitamin D diet is prudent to reduce risk of corticosteroid-induced osteoporosis. Next, to lower susceptibility for infection, patients are prompted to wash hands often and monitor for effective personal hygiene. They are also urged to avoid crowded areas and should limit their contacts to people with acute and serious infections. Long-term corticosteroid therapy increases protein breakdown, negative protein balance and can lead to further muscle wasting and demineralization of the bones, posing high risk for injury. Fall risk prevention techniques should be reinforced. Patients are taught to inspect skin daily for cuts and wounds, and to cover any injuries with a sterile dressing. They should notify healthcare providers of non-healing wound or wound infection. Changes in body appearance, such as moon face, weight gain, muscle wasting and increased fat deposition in the trunk, are common features of long-term corticosteroid use. Most importantly, patients should avoid discontinuation of long-term corticosteroid without medical instruction. Sudden discontinuation can lead to adrenal crisis and possible death. Feelings of helplessness or loss of control often reflect their psychological disturbance. Nurses should be active listeners and make appropriate referrals, such as counselling services and/or support groups for patients and their caregivers.

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HKARF/HKSR Scholarship Awardee 2016

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I am extremely grateful to the HKSR/HKARF for their prestigious scholarship, which allowed me to undergo overseas training in London (2016-2017) for Allergy and Clinical Immunology.

Under the guidance of the world renowned experts and colleagues, I quickly felt competent to see patients independently as an honorary specialist registrar. I had the opportunity to work together with a multitude of different specialists and healthcare professionals, and was greatly impressed by the holistic and multi-disciplinary teamwork available. There is much that we can learn from the differences with our local health care systems and patient management. My supervisors were also kind enough to involve me in various academic projects and research. This has led to multiple publications – both accepted by major peer-reviewed journals and awarded for best oral / poster abstract at major conferences.

Neither the spectrum of patients nor the expertise/facilities to deal with such a specialized field are available in Hong Kong. Without this experience, I would not be able to complete my specialist training in Immunology & Allergy. Again, I am thankful to the HKSR and HKARF for this invaluable opportunity – I am confident that the knowledge, skills and experiences I acquired in this past half a year will surely have a long-lasting impact on my lifelong career. With this experience as my groundwork, I hope to better develop our local allergy/immunology services to better serve our patients in Hong Kong. I look forward to working closely with the HKSR/HKARF towards this endeavor in the near future!

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