

Review

Multimodal imaging for the clinical assessment of dermatomyositis and polymyositis: A systematic review

Yanlong Jia ^{a,b}, Hongan Tian ^a, Jing Deng ^{a,b}, Kaihu Yu ^{a,b,*}

^a Department of Medical Imaging, Clinical Medical College, Hubei University of Science and Technology, Xianning 437100, Province Hubei, China

^b Department of Radiology, Xianning Central Hospital, The First Affiliated Hospital of Hubei University of Science and Technology, 228 Jingui Road, Xianning 437100, Province Hubei, China

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Abstract

Dermatomyositis (DM) and polymyositis (PM) are two common autoimmune myopathies. In the past, the diagnosis criteria of DM/PM depended primarily on clinical features, blood enzyme levels, electromyogram and muscle biopsies. However, there still are some imperfections of the diagnosis criteria of DM/PM. Multimodal imaging as multidisciplinary approach has been proven to be valuable in evaluating patients with DM/PM. It is can not only identify the lesions distribution, evaluate muscle atrophy and fatty replacement, but also provide more information of muscle biochemical changes, myofibrillar structure and blood supply. In recent years, multimodal imaging has provided new insights into pathogenesis, diagnosis, therapeutic evaluation and progression of DM/PM.

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1. Introduction

The idiopathic inflammatory myopathy (IIM) is a group of chronic systemic connective tissue diseases, which mainly include dermatomyositis (DM), polymyositis (PM) and inclusion body myositis (IBM). Dermatomyositis and polymyositis are more common than IBM in the world. The incidence and prevalence of DM and PM are 1.4 and 5.8, 3.8 and 9.7 cases among 100,000 people in the United States, respectively [1]. Although the etiology and pathogenesis are still unclear, most of scholars believe that a combination of genetic and environmental factors determines susceptibility to DM and PM [2,3].

Because of nonunique and more overlaps between the clinical features of DM and PM, it is not uncommon to make a misdiagnose between PM and DM. As the most important method, muscle biopsy can make the accurate diagnosis and distinguish the different subtypes of myositis. Some studies have demonstrated that [4] the pathogenesis of DM and PM are different. DM exhibits mononuclear cell infiltration consisting of β cells and CD4⁺ T cells in the perimysial and perivascular areas, which supports a humoral-mediated pathogenesis mechanism. While PM is characterized as invasion of non-necrotic muscle fiber cells by the mononuclear macrophages cells and cytotoxic CD8⁺ T cells, which suggest a cell-mediated cytotoxic mechanism.

Additionally, most of the previous studies [1–6] have widely reported myopathy-related malignancy in Western and Asian populations. The prevalence of malignance is higher in DM than in PM patients, with a ratio of 1.36:1. The female patients with PM or DM is more prone to malignant transformation with a mean age of 45.5 ± 5.1 years. Extramuscular organs or systems may be involved, such as

* Corresponding author. Department of Radiology, Xianning Central Hospital, The First Affiliated Hospital of Hubei University of Science and Technology, 228 Jingui Road, Xianning 437100, Province Hubei, China.

E-mail address: yukaih2003@aliyun.com (K. Yu).

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nasopharyngeal carcinoma, ovarian cancer, lung cancer, interstitial lung disease (ILD), or diseases of gastrointestinal tract and cardiac system etc, among which the lung cancer in western countries and nasopharyngeal carcinoma in Asia and northern Africa are the most common myopathy-related malignancies [5,7].

In 1975, Bohan and Peter [8] first proposed the DM and PM diagnostic criteria which are widely used nowadays: (1) characteristic skin findings (erythematous rash), (2) symmetric proximal muscle weakness, (3) elevated muscle enzyme levels within the blood, such as creatine kinase (CK), lactic dehydrogenase (LDH), aspartate amino transferase (AST), Alanine transaminase (ALT) and myositis specific autoantibodies may be detected, (4) electromyographic (EMG) abnormalities, and (5) evidence of muscle edema, degeneration, inflammation cell infiltration and fibrosis necrosis on muscle biopsy. (Last four of the 5 criteria must be present for a definite diagnosis of PM).

Although the diagnosis criteria of DM and PM widely used today, there are still some imperfections: (1) muscle strength testing may have a strong subjective component, (2) serum levels of muscle enzymes are not elevated in all patients, and may be normal or only slightly higher in very early or chronic stage; other factors such as exercise, myocardial infarction, hepatitis, and drug effect can affect the concentration of serum muscle enzymes [9], (3) EMG and biopsy are invasive procedures and not suitable for repeated application. Because of these limitations, it was desirable to find more adequate techniques to recognize the onset, progression or improvement of myositis before and during therapy.

Recently, multimodality imaging including both structural and functional imaging has gradually been used for evaluating the myositis and may play an important role in investigating pathogenesis and diagnosis of myositis. Here, we provide an up-to-date review regarding the application of multiple imaging in DM and PM, and discuss the pros and cons of these methods in this field.

2. Traditional imaging techniques

2.1. X-ray and Computed Tomography (CT)

It makes up to 20% of myositis patients are associated with ILD which may lead to complications such as pulmonary hypertension or cor pulmonale and may be fatal under rapidly progressive condition. As the earlier imaging tools, X-ray and Computed Tomography (CT) have been used in the diagnosis of the myositis [4]. Chest X-ray allows detection of pulmonary changes prior to appearance of clinical symptoms. High-resolution CT is sensitive to the specific interstitial pneumonitis appearances of ground-glass opacities and honeycombing [10]. Additionally, perfusion CT is useful for evaluating skeletal muscle microcirculation perfusion, and CT-controlled targeted needle biopsy is also useful for sampling [11]. However, the lower resolution of soft tissue, the risk of contrast agent-related adverse events and radiation effect limit wide clinical application of CT in myositis.

2.2. Ultrasound

Compared with CT, ultrasound has the advantages of popularity, convenience, high spatial resolution and real-time imaging without exposure to ionizing radiation and has been widely used in clinical practice [12,13]. As we all know, ultrasound can not only appreciate the appearance of muscle structure (muscle, fascia, adjacent fat), but also guide the muscle biopsy. Recent ultrasound studies demonstrated that skeletal muscle perfusion by utilizing contrast agents was useful in evaluating myositis activity and response to treatment [12,14]. Power Doppler ultrasonography (PDUS) is useful for the detection of DM-related fasciitis, especially in the early stage [13]. Additionally, ultrasonic elastography, as a new technology, can offer a good functional evaluation by measuring the stiffness/elasticity of the muscles (Figs. 1 and 2) [15]. The elasticity of DM and PM usually decrease, which may attribute to the muscle fibrosis and atrophy. However, the sensitivity and specificity of contrast-enhanced ultrasound in diagnosis of myositis are lower than that of MRI, because of the false positive and negative predictive values of contrast-enhanced ultrasound are higher than that of MRI [14].

2.3. Conventional MRI (T1WI, T2WI and STIR)

Magnetic resonance imaging with the advantages of high resolution/high contrast, noninvasion, multiplanar imaging, high sensitivity and specificity, was widely used in the diagnosis of inflammatory myopathy including disease activity evaluation, clinical efficacy evaluation, and guiding rational treatment. Edema exists in early stage of myositis, while fatty infiltration/transformation or muscle atrophy occurs in later stage of myositis. Based on the presence or not of edema, affected muscles can be distinguished from non-affected muscles using MRI [12]. Usually, the areas with inflammatory edema exhibit hyperintensity, whereas the less affected or non-affected muscles present low signal intensity on T2WI (Fig. 3).

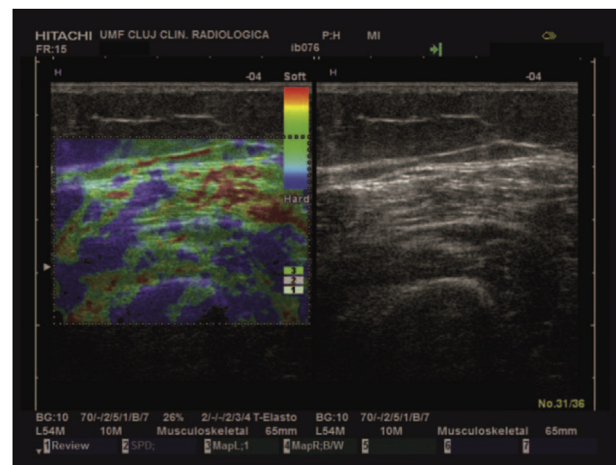


Fig. 1. Polymyositis: F, 55 years – elastography at the medium third of the thigh (alteration of normal striated appearance, with important reduced elasticity in the intern part of the muscle) [15].

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