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Imaging in dermatomyositis in adults and children

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Abstract

Dermatomyositis is a rare autoimmune disorder in which an abnormal immune reaction against vascular endothelial antigens and endomysium leads to obstructive inflammatory changes of blood vessels within muscles, skin and other tissues. The disease is characterized by involvement of muscles, and less frequently of other systems, including the gastrointestinal tract, heart and lungs. Dermatomyositis may be diagnosed based on a detailed patient history, through clinical examination, detection of characteristic physical findings and certain specialized tests. Additional imaging studies may be performed to aid in the diagnosis. These include magnetic resonance imaging and ultrasound of the affected muscles. Magnetic resonance imaging is the modality of choice in the diagnostic work-up and monitoring of dermatomyositis affecting muscles, fasciae, and the subcutis. It may recognize acute inflammatory edematous changes in the affected muscles as well fatty replacement and atrophy. The role of ultrasound to diagnose and follow up muscle echogenicity, vascularity, elasticity and volume during treatment has increased over the last years in both adults and children. Ultrasound is used to discriminate between high and low disease activity, may show features of subclinical disease and may be used to confirm remission.

Introduction

Dermatomyositis (DM) is a rare systemic autoimmune disorder in which an abnormal immune reaction against vascular endothelial antigens and endomysium appears to lead to obstructive inflammatory changes of blood vessels within muscles, skin and other tissues⁽¹⁾.

DM belongs to the group of idiopathic inflammatory myopathies (IIM) which according to the recent EULAR/ACR classification criteria, can be classified into six subgroups: polymyositis (PM), inclusion body myositis (IBM), dermatomyositis (DM), amyopathic dermatomyositis, juvenile dermatomyositis (jDM), and juvenile myositis other than jDM⁽²⁾.

DM, PM and jDM also belong to the group of rheumatic connective tissue diseases (Tab. 1). Each of them may

additionally coexist with another connective tissue disease, most commonly with scleroderma and systemic lupus erythematosus, and less commonly with rheumatoid arthritis or, in children, with juvenile idiopathic arthritis (JIA). In this situation, the term *overlap syndrome* is used^(3,4). In this paper, we review the available literature on the clinical presentation of adults and juveniles with dermatomyositis and describe imaging features of the disease in conventional radiography, ultrasound, magnetic resonance and computed tomography.

Epidemiology and clinical presentation

The annual incidence of DM is estimated between 1 and 10 new cases for every 1 million inhabitants, which leads

Tab. 1. Rheumatic entities belonging to the group of connective tissue diseases, including dermatomyositis

| Diffuse connective tissue diseases | |
|------------------------------------|----------------------------------|
| 1 | Rheumatoid arthritis |
| 2 | Juvenile idiopathic arthritis |
| 3 | Systemic lupus erythematosus |
| 4 | Antiphospholipid syndrome |
| 5 | Scleroderma |
| 6 | Dermatomyositis and polymyositis |
| 7 | Systemic vasculopathies |
| 8 | Sjögren's syndrome |
| 9 | Overlap syndrome |
| 10 | Others |

to a prevalence estimated between 1/50,000 and 1/10,000. It is more frequent in women than in men (sex ratio: 2F/1M). The disease may occur at any age, but has two peak ages of onset: approximately 40–60 years in adults and 5–15 years in children⁽⁵⁾.

Increased risk of malignancy has been noted in both genders, with a relative risk of 5.29 for males and 4.56 for females. This increased risk persists even after 5 years following the initial diagnosis of DM⁽⁶⁾. The subtypes of malignancies that are significantly associated with DM remain controversial. Lung, ovarian, pancreatic, gastric and colorectal malignancies have been described to be more frequently associated with DM, but heterogeneity may exist and could reflect differences in malignancy risk across different populations⁽⁶⁾.

In both adults and children with DM, early signs of the disease are characteristic skin lesions that may precede muscle weakness. However, in up to 40% of patients skin involvement may be the only presenting sign of DM (*dermatomyositis sine myositis*). Skin abnormalities include a distinctive rash that may be present on the upper eyelids, forehead, cheeks and bridge of the nose ("butterfly rash"/ heliotrope rash) or other regions, and Gottron's papules^(3,7,8). Apart from cutaneous involvement, the main clinical symptom is progressive proximal muscle weakness, with coexisting pain, tenderness, and stiffness, first related to muscle inflammatory edema, later to fatty atrophy and muscle fibrosis. Symmetrical involvement of large proximal muscles of the upper and lower extremities and relative sparing of the muscles of the body trunk are believed to be typical of DM in children and in adults^(3,9). Nevertheless, trunk muscles are also frequently affected in our practice. Systemic involvement can include cardiovascular, pulmonary (e.g. interstitial lung disease) or gastro-intestinal complications⁽³⁾.

The symptoms as well as physical and imaging findings associated with jDM are similar to those observed in adult-onset DM. However, there are some differences^(3,7,10). The

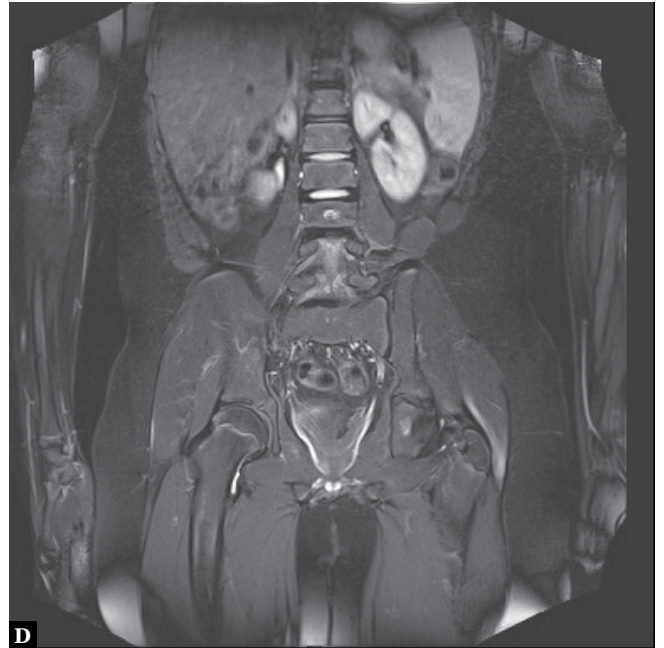
onset of DM in children is usually more sudden and more acute than in adults, and often involves skin manifestations followed by muscle weakness and inflammation. Vasculopathy is considered central to the pathogenesis of the jDM. The exact nature of vasculopathy is not yet understood, but it is a complex process with both inflammatory and non-inflammatory occlusive components. Development of vasculopathy is associated with severe extra-muscular manifestations of jDM, such as gastrointestinal and cardiac complications, interstitial lung disease, ulcerative skin disease or development of calcinosis, and portends a poor prognosis. In patients with GI vasculopathy, associated findings may include abdominal pain, constipation and tarry, black stools (melena) or vomiting of blood due to the development of bleeding peptic ulcers. Calcification of muscles, skin and subcutaneous tissue is more frequent and widespread in childhood DM as compared to adult forms; the deposits have high calcium content and tend to be firm and larger. Calcification and tendinopathic lesions may potentially lead to contractures and localized muscle atrophy. By contrast with adult-onset DM, the disease in children is not associated with malignancy or nail changes. Muscle weakness and impairment may progress to affect other areas, not only typical skeletal muscles, but also, for instance, GI tract muscles, the cardiovascular system



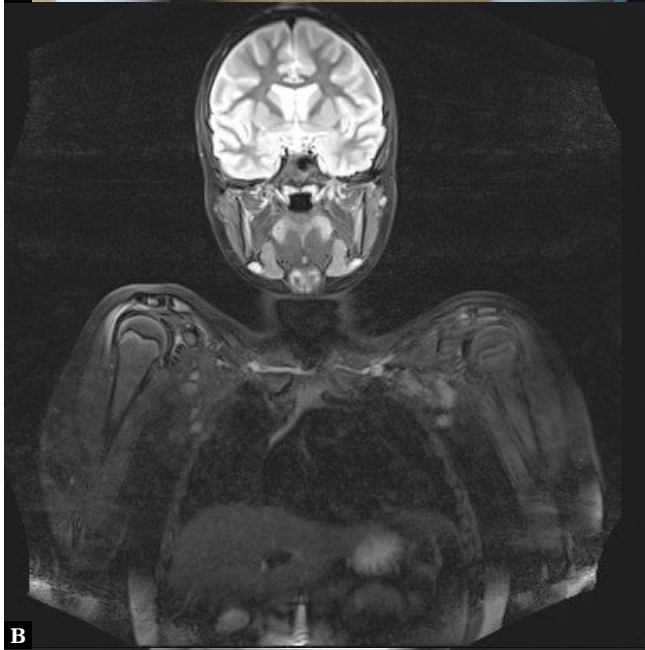
Fig. 1. AP radiographs of the knees and ankles in a 16-year-old boy with DM: diffuse bone loss and diffuse soft tissue calcification characteristic of DM



A



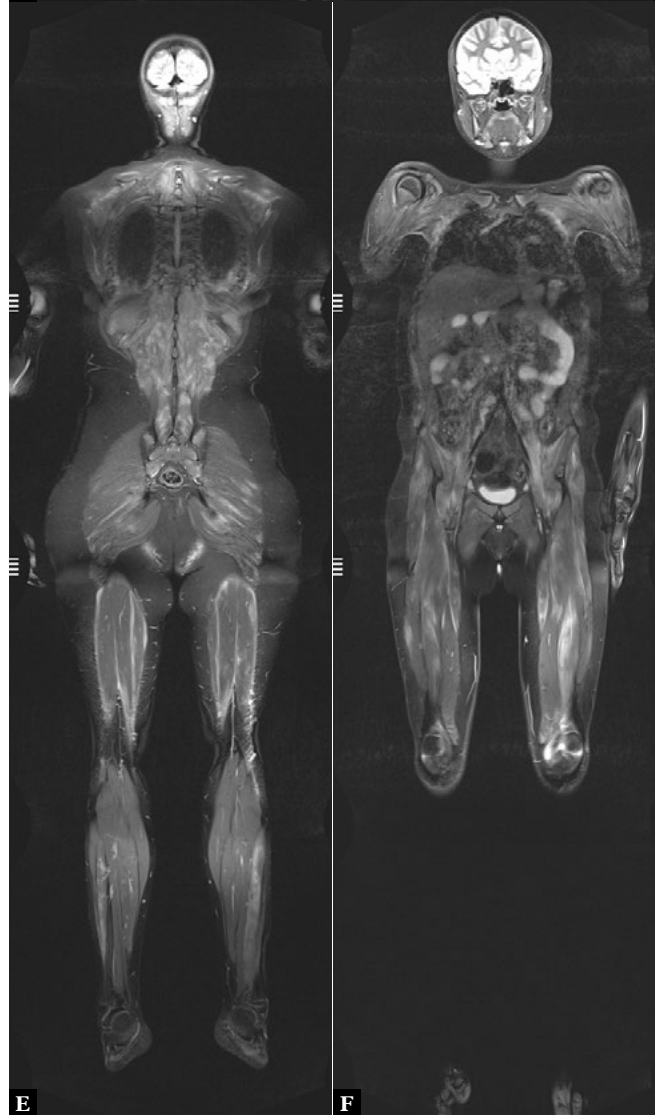
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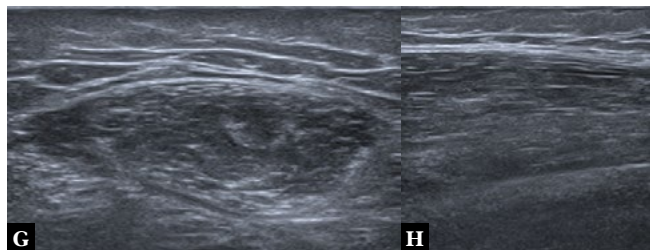


Fig. 2. A 12-year-old boy with DM: **A.** Gottron's papules (hyperkeratotic, erythematous, flat papules with central atrophy) on the dorsum of the metacarpophalangeal and interphalangeal joints; **B, C, D.** Initially, coronal T2-weighted TIRM WB-MRI shows characteristic diffuse, edema-like muscle involvement of several muscles, including tiny areas in the right shoulder, latissimus dorsi and greater trochanter; **E, F.** MRI performed 3 years later shows significant progression with the involvement of the neck muscles, whole limbs, back and pelvis; **G, H.** Ultrasound of the biceps in transverse (**G**) and longitudinal views (**H**) shows diffusely increased echogenicity and blurred fibrillar echotexture of the muscle, and thickened fascia (parents of the child did not administer the recommended medication)

(i.e. heart, blood vessels, and blood circulation), or the lungs. Children may also develop a tiptoe gait secondary to ankle stiffening.

Imaging findings

Radiographs

Radiographs in the acute phase may show discrete soft tissue thickening at the proximal parts of the limbs, increased radiodensity of the subcutaneous tissue and muscle, and poor delineation of the subcutis–muscle interface^(7–9). In the chronic phase, the most characteristic radiographic features are soft tissue calcifications, detected in approximately 25–50% of patients (Fig. 1). They may potentially lead to contractures and localized muscle atrophy. Bone loss of long bones and other complications, such as compression fractures of vertebral bodies or avascular necrosis secondary to steroid therapy, may also be diagnosed.

Ultrasonography

On ultrasound, calcifications may be seen in the locations mentioned above with posterior shadowing.

In the acute phase, some researchers noticed increased muscle thickness and decreased muscle echogenicity^(11–14). However, most published data show that increased echogenicity may be observed in both acute and chronic inflammatory changes in muscles, the latter resulting from fat replacement and fibrosis^(3,8,11,15–21) (Fig. 2). The similar echogenicity of muscle inflammatory edema and fatty involution may limit the ability of ultrasonography to differentiate

disease activity⁽¹⁾. Muscle atrophy, a late finding in the disease course, leads to muscle thinning^(11,21–23) (Fig. 3).

In adults with DM, it was found that echogenicity of muscles may normalize with treatment, paralleling improvement in clinical condition⁽¹⁹⁾. Habers *et al.* observed an increase in the echogenicity of muscles in the first 3 months after treatment initiation in jDM before it normalized, while muscle thickness decreased in the first month after treatment initiation⁽¹²⁾. Also Bhansing *et al.* found that muscle echogenicity could discriminate between high and low disease activity in jDM⁽¹³⁾.

Song *et al.* carried out a sonoelastographic study in 17 patients with inflammatory myopathies⁽²⁴⁾. They scored muscle elasticity using the strain technique and found that the affected muscles in adults and children were much stiffer than in healthy controls. Bachasson *et al.*⁽²⁵⁾ applied shear wave elastography and found lower muscle stiffness in patients with more severe muscle weakness. This may suggest the potential of shear wave elastography to differentiate between disease activity.

Meng *et al.*⁽¹⁸⁾ used power Doppler ultrasound to calculate vascularity scores in the affected muscles, showing higher vascularity in the shorter duration disease subgroups.

Muscle echogenicity may also be used to monitor treatment effects. In some studies, it remained high despite normalization of muscle enzymes, indicating subclinical disease⁽¹⁷⁾. In children with jDM, it was found that approximately 10 years after the onset of symptoms, most patients still had increased echogenicity in at least one muscle group even though they underwent treatment and were believed to be in remission⁽²⁶⁾.

Magnetic resonance imaging

Although MRI is not included as a diagnostic criterion of IIM, it is a reference imaging examination and offers the

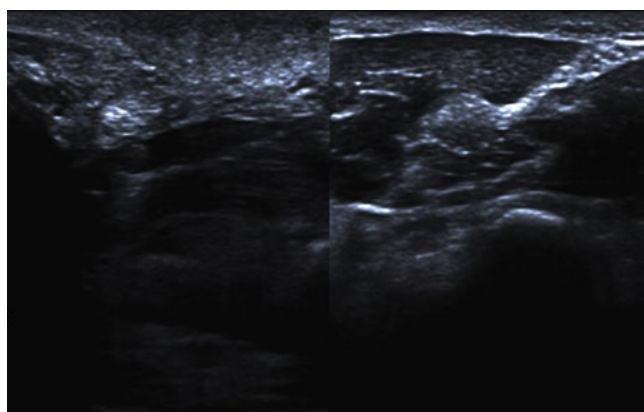


Fig. 3. Ultrasound of the thenar eminences. The left part of the image shows subcutaneous edema of the thenar eminence in the right hand with atrophy of the superficial head of the flexor pollicis brevis (arrow is pointing to the flexor pollicis longus tendon). The right part of the image presents the normal left hand

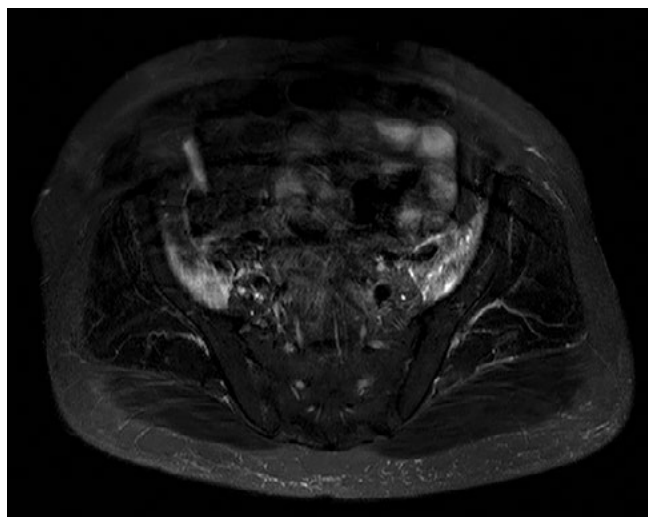


Fig. 4. Pelvic MRI shows bilateral active myositis of the iliopsoas muscle in an adult patient with known DM

best evaluation of soft tissue abnormalities, including muscles⁽⁸⁾. Coronal and axial short tau inversion recovery (STIR) or T2-weighted images, especially with fat suppression (FS), depict increased “edema-like” signal intensity in affected tissues, representing active myositis, fasciitis and subcutis inflammation^(3,8,27). Although these findings are non-specific, MR images are particularly useful in the detection of inflammatory changes within muscles, superficial and deep fascia, and subcutis, which correlate with disease activity (Fig. 4).

MRI is used to select an optimal site for biopsy for a histopathological examination, which is a diagnostic gold

standard in DM⁽⁹⁾. MRI can also detect clinically silent involvement of muscle groups or inflamed fasciae.

Whole-body MRI (WB-MRI), which provides a complete assessment of inflammation, is used for the monitoring of the disease course and for the evaluation of treatment effects, including side effects (avascular necrosis and fractures) (Fig. 3)^(3,7,9). This approach is especially valuable in detecting truncal/axial disease.

Successful treatment prevents muscle fatty atrophy, which is seen as a high signal on T1-weighted images, representing fatty degeneration in the chronic form of the disease, and is a conspicuous marker of damage^(3,7,28,29) (Fig. 5, Fig. 6).

Computed tomography

Computed tomography is less accurate in evaluation muscle edema/inflammation. It might be considered when the suspected etiology is infectious in order to rule out pyomyositis or abscess formation⁽³⁾. It is also more sensitive than MRI in calcinosis detection (Fig. 7).

Treatment and prognosis of dermatomyositis

Although there is no curative treatment for DM, long-term remissions are possible⁽³⁰⁾. The disease-related mortality is estimated at 10% in adult patients, with the cause of death mostly being associated cancer or pulmonary complications. Treatment-related complications can also occur, and muscle involvement can greatly impair patients' quality of life.

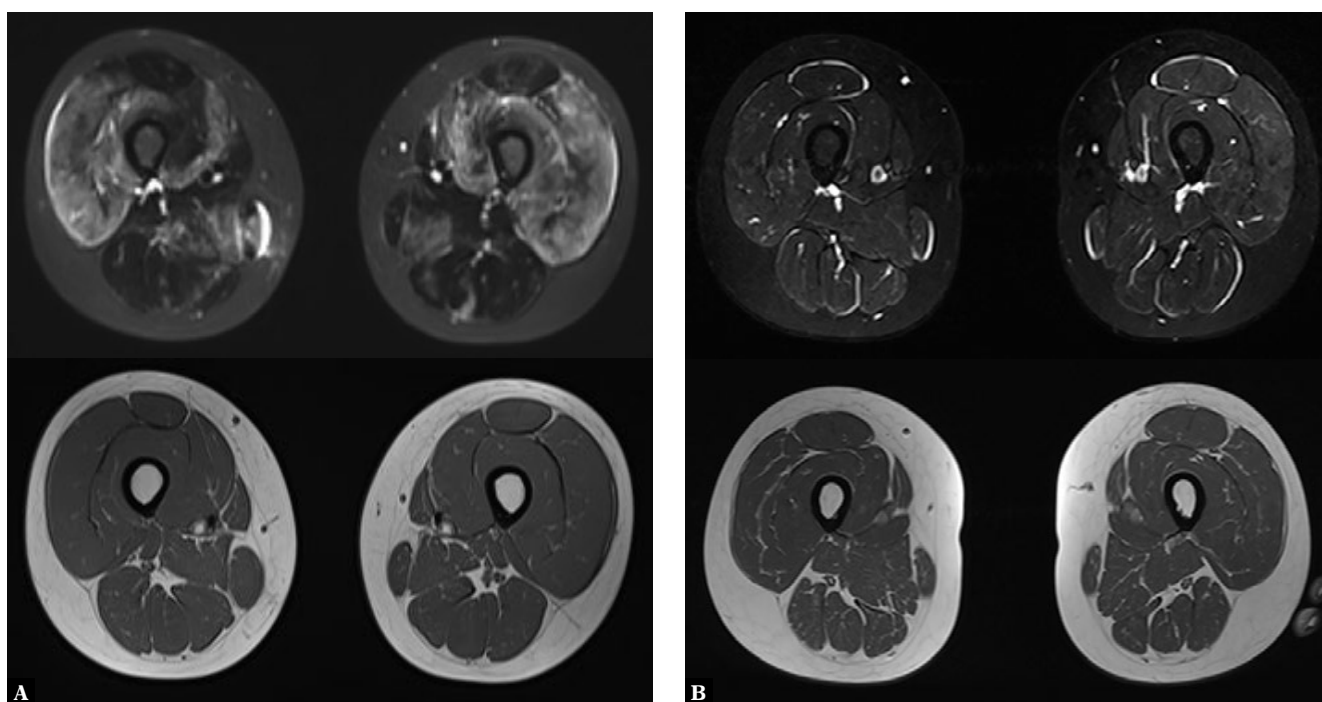


Fig. 5. T2-weighted FS MRI of the thighs shows evolution of muscle involvement in an adult patient with DM at the initial stage (A) and after 4 months of treatment with corticosteroids (B), with features of fatty involution despite treatment

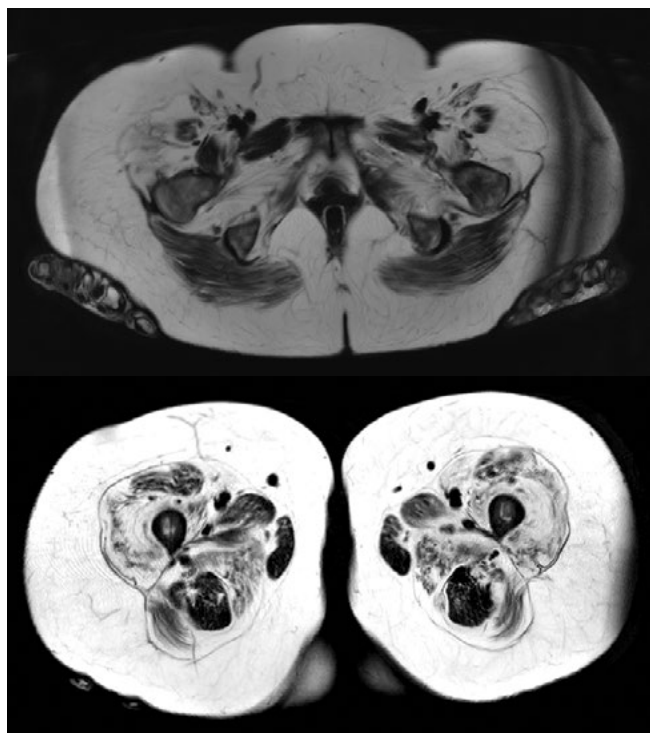


Fig. 6. T1-weighted MRI shows diffuse bilateral amyotrophy and fatty involvement of pelvic and thigh muscles, late-stage DM

In children, there are 3 types of the disease course:

1. Monocyclic, in which within 2 years after treatment initiation there is only one episode with subsequent remission and no relapses;
2. Polycyclic, in which long-term remission periods are interrupted by relapses;
3. With continuous chronic activity.

Treatment of DM is based on glucocorticosteroids (mainly prednisone) or corticosteroid-sparing agents, such as

methotrexate (MTX) or azathioprine. Since effects of MTX become visible after 3 months of therapy initiation, especially children with severe muscle weakness (some of them not being able to stand, sit, etc.) may initially require a high-dose prednisone therapy which starts acting immediately. A high-dose glucocorticoid therapy may, however, produce adverse effects, particularly after prolonged use. For that reason, as soon as clinical improvement is seen, corticosteroid doses may be gradually reduced to the lowest possible dose, sufficient to maintain normal enzyme levels and improve symptoms. Biologic therapies (rituximab and anti-TNF) are used rarely, only in difficult, refractory cases.

According to experts, children with DM may generally be able to discontinue prednisone after approximately two years, when an apparent cessation of symptoms (i.e. remission) is noted. An accurate method enabling disease monitoring and especially confirming remission is lacking.

Conclusion

Dermatomyositis is a rare, systemic disease, with a number of challenges, regarding both diagnosis and therapy. So far, MRI has been the modality of choice in the diagnostic work-up and monitoring of DM affecting muscles, fasciae, and the subcutis. The role of US as a diagnostic tool in the field of all IIMs has grown over the years to monitor changes in muscle echogenicity, elasticity and thickness during treatment. Continuous developments of MRI and ultrasound are expected to further improve patient management.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

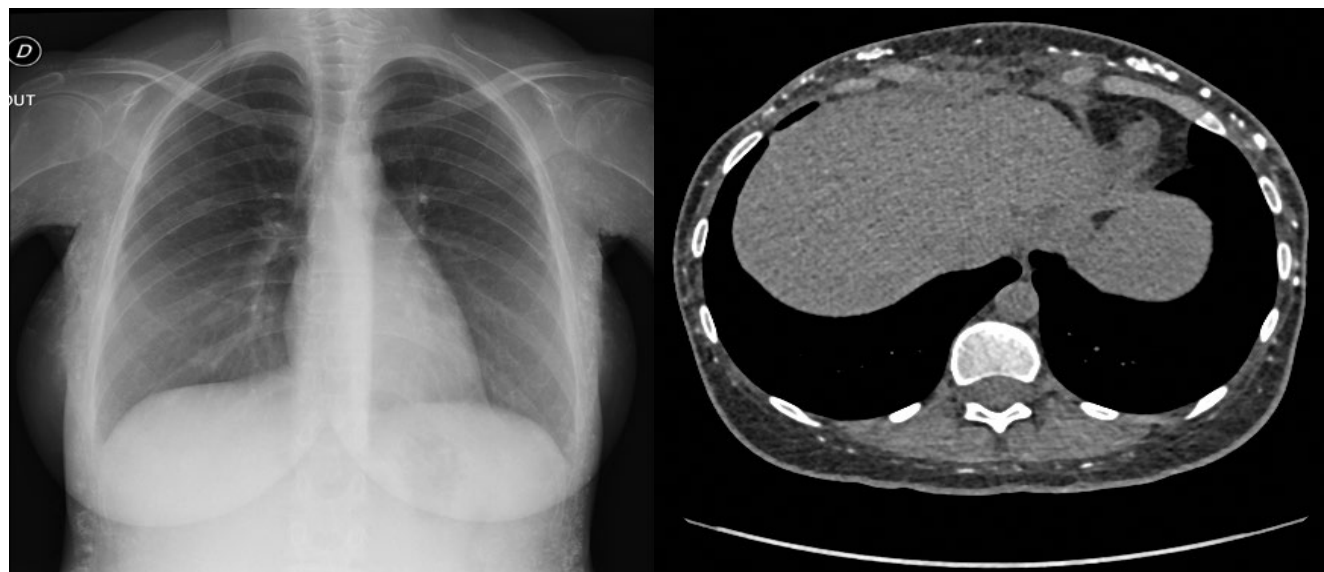


Fig. 7. Chest CT and chest X-ray show diffuse nodular calcifications of parietal soft tissues in a patient with known DM

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