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Letter to the Editor

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Letter to the Editor (Case report)

Whole-body MRI revealed generalized subcutaneous oedema in a patient with juvenile dermatomyositis

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Key message

- Whole-body MRI is useful for detecting generalized s.c. oedema, an indicator of disease severity.

Dear Editor, A 5-year-old girl developed difficulty in walking owing to muscle weakness and erythema of the face after coronavirus disease 2019. The patient had severe autism spectrum disorder, psychomotor retardation and an unbalanced diet. Erythematous eruptions were observed on the eyelids and midface, suggestive of heliotrope eruptions and malar rashes, respectively. No generalized oedema was observed upon physical examination. The following blood test results were obtained: creatinine kinase, 725 IU/l; aldolase, 17.3 mg/dl; C3, 76.9 mg/dl; C4, 38.9 mg/dl; and CH50, 44.1 U/ml. These cutaneous findings were suggestive of JDM. Short tau inversion recovery (STIR) MRI revealed high signal intensities for muscle and s.c. lesions in both thighs, consistent with myositis with s.c. oedema (Supplementary Fig. S1, available at *Rheumatology Advances in Practice* online). Whole-body MRI showed generalized myositis and s.c. oedema (Fig. 1). Chest radiography and CT revealed no abnormal findings. Myositis-specific antibodies were positive for antinuclear matrix protein 2 (NXP-2) autoantibodies. Pathological examination of the right thigh muscle showed mild to moderate variation in fibre size, clusters of necrotic/regenerated fibres, which are considered to reflect microinfarction, and expression of myxovirus resistance protein A, a diagnostic marker of DM (Supplementary Fig. S2A–C, available at *Rheumatology Advances in Practice* online) [1]. Pathological examination of the skin and s.c. tissue revealed no signs of inflammation suggestive of panniculitis, while lobular mucinosis was present in s.c. adipose tissue. The intercellular space of adipocytes was widened (Supplementary Fig. S2D, available at *Rheumatology Advances in Practice* online). Finally, the patient was diagnosed with JDM, with anti-NXP2 autoantibodies. CSs were initiated, but the patient showed poor clinical improvement. After IVIG administration, the clinical symptoms improved markedly.

Here, we describe a paediatric case of JDM with anti-NXP2 autoantibodies complicated by generalized s.c. oedema that could not be observed upon physical examination but was detected by whole-body MRI. Localized oedema is common in DM, whereas generalized oedema is rare. Generalized s.c. oedema has been observed in ∼6% of adult DM cases [2], but there are no reports in JDM. Generalized s.c. oedema can be an indicator of CS resistance and DM disease severity [2, 3]; however, it is rarely reported in JDM [4, 5]. In the Japanese JDM clinical practice guidelines, cases with generalized oedema are classified as severe or fulminant [6]. Paediatricians should consider that JDM with generalized oedema might be CS resistant and require additional treatment.

In recent years, whole-body MRI has been useful for the treatment of malignant tumours and rheumatic diseases. Moreover, it correlates well with muscle score and the childhood-myositis assessment scale [7]. In this case, whole-body MRI detected both generalized myositis and s.c. oedema. Therefore, whole-body MRI is useful in detecting generalized s.c. oedema that is not apparent upon physical examination.

Although the mechanism of generalized s.c. oedema remains unclear, some reports have suggested a pathological association between microinfarction [2] and microangiopathy owing to deposition of the membrane attack complex caused by complement activation on the vessel wall [4, 8]. Schildt

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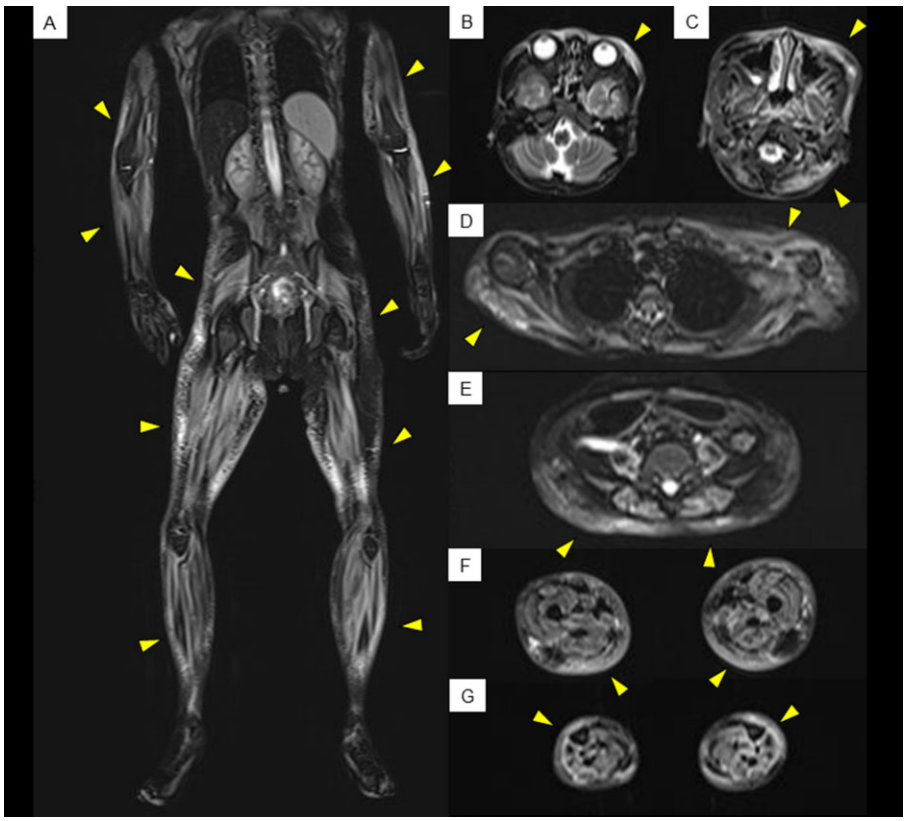

Figure 1. Whole-body MRI. Whole-body MRI on short T1 inversion recovery imaging shows generalized high-intensity signals in the generalized muscle and s.c. lesions (yellow arrow) suggestive of myositis and s.c. oedema. A, coronal view of the whole body. B and C, axial views of the head. D, axial view of the shoulder. E, axial view of the body trunk. F, axial view of the thigh. G, axial view of the lower leg

et al. [4] reported that s.c. oedema might be related to complement activation, because both cases in their study showed decreased C3 levels. Likewise, there is another case of generalized s.c. oedema in which the level of C3 decreased [3]. Here, our patient also showed a mild decrease in C3 levels, which might have been related to complement activation. In the future, it is desirable to investigate the pathology and treatment of generalized s.c. oedema.

Supplementary material

Supplementary material is available at *Rheumatology Advances in Practice* online.

Data availability

Data supporting the findings of this study are available from the corresponding author, Y.F., upon reasonable request.

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