Sporadic inclusion body myositis: a rare hazardous entity with important imaging findings

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ACTA REUMATOL PORT. 2020;45:147-149

Sporadic inclusion body myositis (IBM) is a rare acquired muscle disease that predominantly affects individuals older than 45 years of age[1].

IBM pathogenesis is unknown although it has been classified together with polymyositis, dermatomyositis and immune-mediated necrotizing myopathies as an idiopathic inflammatory myopathy (IIM).

However, IBM has a distinct clinical course being characterized by a slowly progressive weakness, usually asymmetric, involving primarily the finger flexors and quadriceps muscles accompanied by inflammatory and degenerative changes and resistance to immunosuppressive therapy[2].

Imaging assessment is important not only for diagnosis but also for follow-up and treatment response evaluation[3].

Magnetic resonance imaging (MRI) can assist in confirming the diagnosis and phenotyping the disease. Muscle biopsy site should be based on areas of muscle involvement and disease activity identified on MRI pointed to avert the high false-negative rate of blind muscle biopsies.

Continued follow-up using MRI can add important information about the activity of muscle disease and detect cases where sustained immunosuppressive therapy is no longer justified due to complete fatty replacement of muscle (Figures 1, 2, 3, 4)[3].

On T_1-weighted images muscle atrophy and fatty replacement can be accurately assessed, making this imaging modality a reliable method to quantify the degree of muscle atrophy. T_2-weighted images with fat suppression or short tau inversion recovery (STIR) sequences demonstrate muscle edema corresponding with disease activity and treatment response (Figures 2, 5)[1,4].

IBM has some distinctively image features regarding muscle involvement, it has been reported that fatty replacement occurs preferentially in the quadriceps femoris muscles with relative sparing of the rectus femoris (Figure 3)[2].

Despite being well described, the pattern of fatty infiltration does not grant MRI the power for an established differential diagnosis between the different IIM therefore MRI features are not currently included in IBM diagnostic criteria[2].

IBM is diagnosed on the basis of clinical presentation, elevated serum skeletal muscle enzymes, findings on electromyography and muscle biopsy[5].

Genetic testing is also part of the diagnostic procedure as these patients are sometimes misdiagnosed as having an acquired myositis. Genetic related muscle dystrophies should also be considered in the differential diagnosis with either an autosomal dominant or recessive patterns of inheritance. The patient presented tested negative for myotonic dystrophy type 2 (the most common form).

There is no effective treatment for IBM with most patients being resistant to treatment, however the new

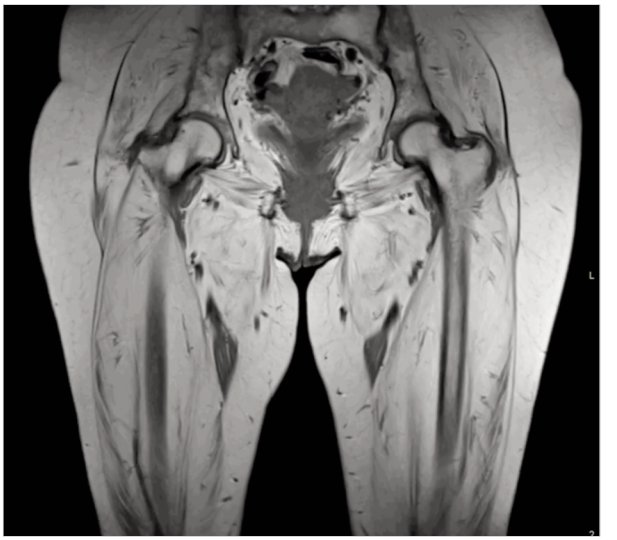

FIGURE 1. Coronal T1-weighted image showing diffuse advanced fatty infiltration of both thigh and pelvic muscles bilaterally and near complete replacement by fat of the quadriceps muscles bilaterally

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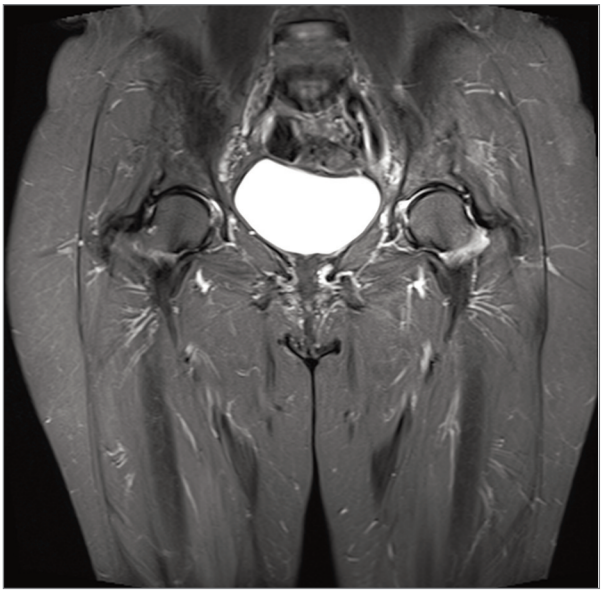

FIGURE 2. Coronal STIR image showing absence of significant edema at the level of the thigh.

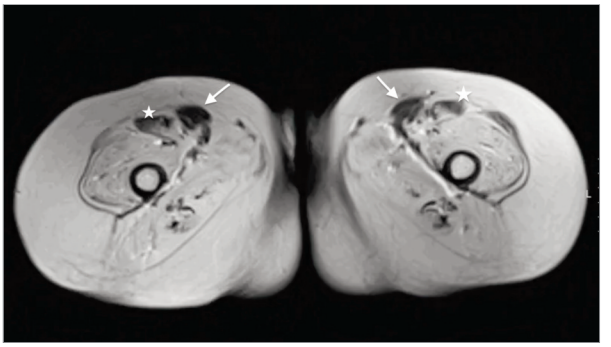

FIGURE 3. Axial T1-weighted image showing near complete replacement by fat of the quadriceps muscle bilaterally with a relative spare of the rectus femoris muscle (stars) and the sartorius muscle (arrows)

pathophysiological insights have led to the search of alternative therapeutic approaches expected to be effective in the future[1].

In conclusion, MRI can accurately document the magnitude and nature of muscle abnormalities with high signal intensity seen in the active phase and refractory treated patients on STIR images. The last few years have witnessed important progress in the understanding of IBM. These advances may eventually lead to improved diagnosis and the discovery of effective pharmacotherapy for this progressive, highly morbid entity with poor treatment response[1].

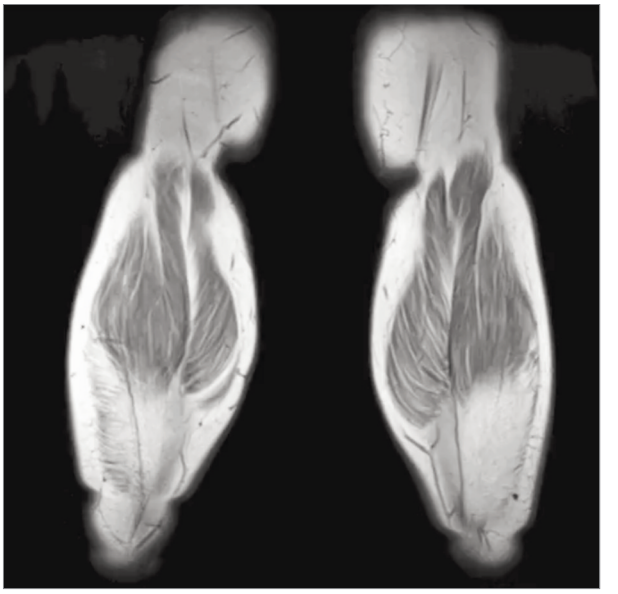

FIGURE 4. Coronal T1-weighted image of the posterior legs showing diffuse moderate fatty infiltration of both leg muscles.

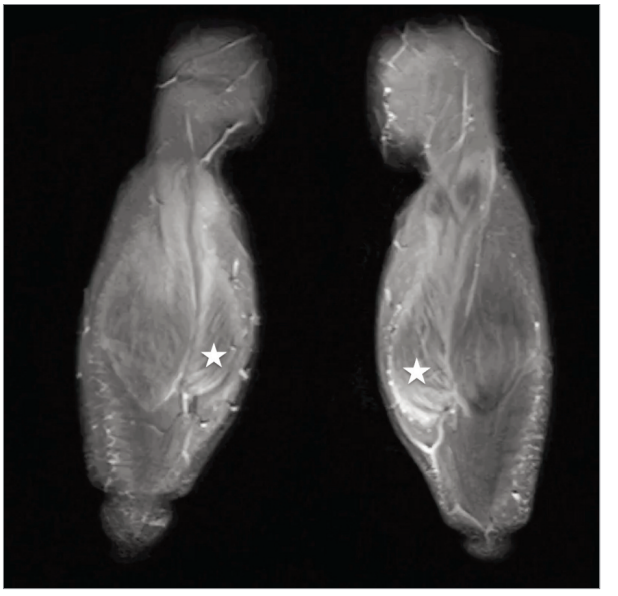

FIGURE 5. Coronal STIR at the same level as Fig. 3 showing active disease with muscle edema involving preferentially the medial gastrocnemius in both legs (star)

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