PET/CT scan revealing active idiopathic inflammatory myopathy with normal serum sarcoplasmic enzyme levels

Sirs,

We describe a 60-year-old woman who was admitted to our Department because of shoulder and pelvic girdle muscles weakness and dysphagia. Two years before, she was diagnosed with polymyositis (PM) on the basis of proximal muscle weakness. electromyographic myopathic changes and endomysial inflammatory cell infiltrates surrounding and invading muscle fibres expressing MHC-I-antigen (1, 2). Serum sarcoplasmic enzyme levels were within normal limits, as observed in less than 10% of patients affected with idiopathic inflammatory myopathies (IIM) (2, 3). She was given prednisone 50 mg/day with scheduled tapering and azathioprine 2 mg/kg/day. One year later, because of increased liver function tests and poor clinical response, treatment was switched to mycophenolate mofetil (MMF) 2 g/day. Two months before admission she spontaneously discontinued MMF because of acute-onset dysphagia.

At admission she was receiving prednisone 12.5 mg/day and lansoprazole 30 mg/day. Clinical examination showed atrophy of

shoulder and pelvic girdle muscles and weakness more pronounced in the proximal muscles of the neck and upper limb, dropped head and ulcerated calcinosis of the upper back. Serum sarcoplasmic enzyme levels were within normal limits. In order to identify hyper-metabolic lesions related to malignancy and active myositis, after instructing patient to minimise muscle activity for 48 hours, a whole-body PET/CT scan (Discovery ST8, GE healthcare) with 18F-fluorodeoxyglucose (FDG) was performed. Quantitative FDG uptake was examined by an experienced nuclear medicine physician using Xeleris software. Maximum standardised uptake value (SU-Vmax) and mean SUV were calculated by creating an elliptic region of interest with a fixed area of 0.5 mm^2 at the region with the highest FDG uptake in each muscle, excluding the region obviously influenced by FDG uptake in other anatomical structures. FDG-PET/CT scan (Fig. 1) revealed hypermetabolic foci in the posterior wall of distal oesophagus (SUVmax 5.4) and in upper back calcinosis; moreover, an asymmetric uptake at level of the deltoid (mean SUV : right = 5.9; left = 2.5) and pectoralis (mean SUV: right = 3.7; left = 1.4) muscle and a symmetric uptake at level of psoas (mean SUV: right = 1.2; left = 1.2) and biceps femoris (mean SUV : right = 0.7; left = 0.8)

muscle were detected. An oesophagogastroduodenoscopy revealed severe oesophageal ulceration with stricture, bleeding and pseudomembranes due to Candida albicans, while biopsies excluded malignancy demonstrating that the stenosis was secondary to peptic esophagitis likely worsened by MMF therapy. She denied use of statin or other myotoxic drugs and no causes other than active IIM were found in order to explain muscle weakness and increased muscular uptake on PET/CT. Accordingly, MMF was replaced with Methotrexate up to 20 mg weekly plus folic acid 5 mg. Six months later she presented in our outpatient clinic reporting improved muscle strength which was objectively confirmed on clinical examination and, therefore, steroid tapering was scheduled.

Several approaches assist in evaluating and monitoring the extent of disease activity in IIM including key clinical features, laboratory tests, immunological markers, muscle T1 and short τ inversion recovery MRI sequences (4). Recently, few authors have reported the use of FDG-PET/CT in patients with IIM (5-8). FDG-PET/CT was reported as effective in screening for malignancy (5), which are associated with IIM in 17– 40% of cases within 2 years after diagnosis (9), and also sensitive and specific in distinguishing patients with active myositis from

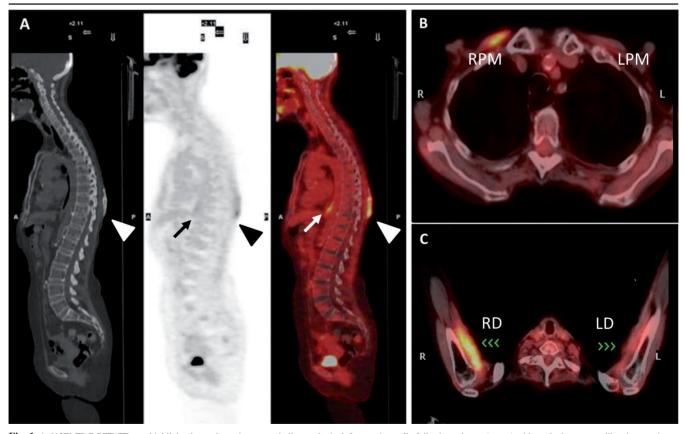


Fig. 1. A. [18F]-FDG-PET/CT scan highlighted a moderate hypermetabolic area in the left posterior wall of distal esophagus (arrow) with marked upstream dilatation, moderate and irregular uptake in the upper back calcinosis (arrow head) extending up to the cutaneous planes from D1 to D8. B. Diffuse and moderate tracer uptake in the right pectoralis major (RPM) muscle. C. Diffuse and moderate tracer uptake in the deltoid muscles bilaterally, more pronounced in the right (RD) than in the left (LD).

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unaffected controls (6-8). Nevertheless, high cost and exposure to ionising radiation recommend to restrict the use of FDG-PET/ CT to a subset of patients. Our case report suggests that FDG-PET/CT may be useful in patients with clinical suspicion of malignancy and active IIM but with normal muscle enzyme levels.

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