Active subcutaneous calcinosis demonstrated by fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography in a case of limited cutaneous systemic sclerosis

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ABSTRACT

Systemic sclerosis (SSc) is a rheumatic autoimmune disease of unknown origin causing fibrosis of the skin and the internal organs. The limited cutaneous variant is the most common subtype of SSc, and it is predominantly characterized by skin and soft-tissues involvement. A 72-year-old woman, who had been diagnosed with the limited cutaneous form of SSc 16 years before, underwent fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography (PET/CT) examination due to unexplained weight loss and recent onset of fatigue and joint pain. PET/CT images showed widespread soft-tissue calcinosis characterized by elevated glucose uptake.

Keywords: Positron emission tomography/computed tomography, soft-tissue calcinosis, systemic sclerosis

We report the case of a 72-year-old woman, who at the age of 56 had been diagnosed with the limited cutaneous variant of systemic sclerosis (SSc) on the basis of Raynaud phenomenon and symmetrical skin thickening of the fingers, both of which had regressed spontaneously over the following 10 years. She presented to our institution with unexplained weight loss and recent onset of fatigue and joint pain. At admission, laboratory tests were notable for the elevation of serum C-reactive protein levels (2.11 mg/dL) and erythrocyte sedimentation rate (57 mm/h). The patient was referred for a fluorine-18 fluorodeoxyglucose (F-18 FDG) positron emission tomography/computed tomography (PET/CT) scan to evaluate for possible recurrence of SSc. Images demonstrated extensive areas of irregular tracer uptake around the shoulders and the hips, corresponding to extensive calcifications within muscles and periarticular soft tissues [Figure 1a-c, e and f]. In addition, focal areas of tracer deposition were identified in smaller calcifications of the soft-tissues of the thighs [Figure 1d and g]. These findings were suggestive for diffuse active subcutaneous calcinosis, and thus, the patient was started on steroid and immunosuppressant therapy, achieving rapid regression of symptoms and normalization of inflammatory markers.

SSc is a chronic systemic autoimmune disease characterized by diffuse micro-vascular damage and pathologic deposition of collagen in the skin, and potentially, every organ of the body.[1] Limited cutaneous SSc (LcSSc) is the most common form of SSc, accounting for about 60–70% of cases, and it was previously identified by the acronym CREST in reference to its most typical clinical manifestations (calcinosis, Raynaud's phenomenon, esophagitis, sclerodactyly, and telangiectasias).[2-4]

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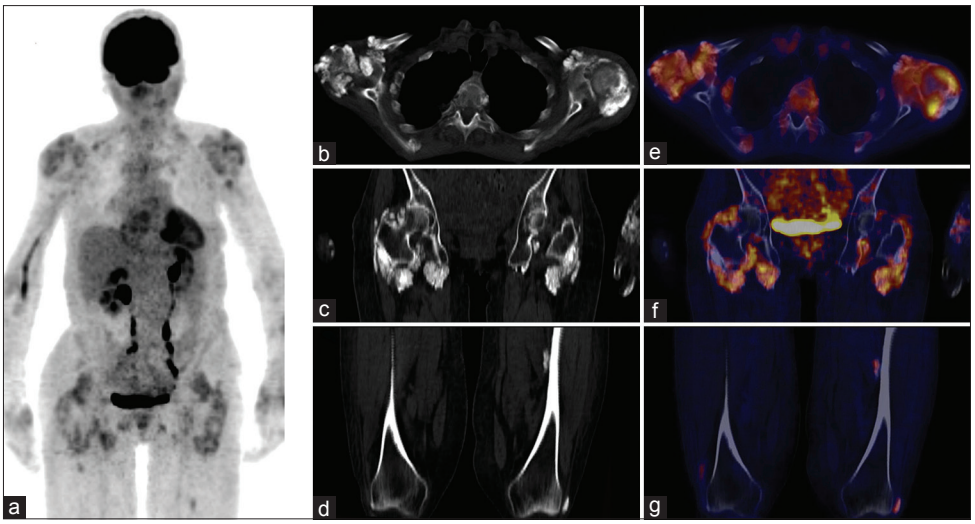

Figure 1: Whole-body fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography maximum intensity projection image (a) revealing bilateral inhomogeneous fluorodeoxyglucose uptake around the shoulders and the hips and smaller areas of focal tracer uptake in the thighs. Computed tomography images showed widespread calcifications in the soft-tissues and muscles around the shoulders (b) and the hips (c) and smaller calcium concretions in soft-tissue of the thighs (d). Corresponding fused positron emission tomography/computed tomography images (e-g) demonstrated areas of increased fluorine-18 fluorodeoxyglucose deposition at the sites of calcifications

Subcutaneous calcinosis is the pathological deposition of insoluble calcium salts in muscles and soft-tissues in the absence of serum calcium and phosphate abnormalities. It is a frequent occurrence in many different rheumatic disorders, and especially in LcSSc, in which it is reportedly present in up to 25% of cases.[5] Although the exact pathogenesis of calcinosis in LcSSc is unknown, altered calcium metabolism due to chronic tissue hypoxia and increased inflammatory cells activity appears to play a pivotal role in the development of calcifications.[6]

Calcinosis represents a great clinical challenge in LcSSc as calcium deposits are often painful and may become infected and cause skin ulcerations.[7] At present, clinical evaluation is the main approach for monitoring soft-tissue manifestations of SSc, and there was no clear consensus on the additional employment of imaging techniques.[8]

It is a well-known fact that F-18 FDG does accumulate in inflammatory lesions. The higher tracer uptake in the areas of inflammation is explained by the increased number of glucose transporter proteins expressed on the cell membrane of activated macrophages, which rely on glucose as their main energy source.[9] Indeed, F-18 FDG PET/CT has been successfully applied in the evaluation of a wide variety of chronic and acute inflammatory disorders.[10] However, there are not many studies exploring the potential use of F-18 FDG PET/CT in the management of SSc. The present report shows that F-18 FDG PET/CT may be able to provide a whole-body assessment of the extent and the inflammatory activity of subcutaneous calcinosis in LcSSc.

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Conflicts of interest

There are no conflicts of interest.

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