

Spinal cord compression by ectopic calcinosis in scleroderma

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ABSTRACT

Systemic sclerosis (SS) is a chronic, multisystemic disease, characterized by inflammation associated with fibrosis. Calcinosis is one of the manifestations of this disorder, observed in 10 to 20% of cases. It is usually located on the extensor surface of the phalanges, peri-articular tissue and near the bone prominences. There are only a few cases reported of SS with vertebral column involvement and spinal cord compression by calcinosis causing serious neurological complications. We describe a fatal case of SS who showed tetraplegia secondary to ectopic calcinosis in the cervical medulla and present a review of the literature on the subject.

Introduction

Systemic sclerosis (SS) is a chronic disease, of unknown aetiology, characterized by inflammation associated with fibrosis. It involves the skin, synovium, muscles, blood vessels and internal organs, notably the gastro-intestinal tract, heart, lungs and kidneys.

Calcinosis is one manifestation reported in 10 to 20% of cases of PSS (1). It occurs predominantly in females and it is usually located on the extensor surfaces of the phalanges, elbows and knees. Very rarely calcinosis may be located in the spinal medulla, causing serious neurological complications. We describe a fatal case of diffuse SS who showed tetraplegia secondary to ectopic calcinosis in the cervical medulla and present a review of similar cases previously published.

Case report

A 51-year-old Brazilian black female was diagnosed with systemic sclerosis 22 years ago. She had a diffuse form of the disease characterised by "sclerodermic facies"; Raynaud phenomenon; cutaneous thickening of the sites proximal to the elbows and knees, the chest and abdomen; finger pad scarring; telangiectasias on the face and palm; symmetrical polyarthralgia and dysphagia for solids. Latterly, she presented calcinosis on the hands and re-absorption of the distal phalanges, detected by radiography; and delay of oesophagus em-

ptying on the roentgenologic barium examination. Chest radiograph revealed calcified nodules resulting from pulmonary tuberculosis, which was previously treated, and gross calcifications in shoulders and para-vertebral areas.

In January 2002, she presented pain and progressive weakness in her upper limbs, which confined her to bed. She was taking D-penicillamine 250 mg/day. Cardiovascular, respiratory and abdominal exams showed no abnormalities. The skin was diffusely thickened (modified Rodnan skin score = 34), with telangiectasias on face and palm regions and with finger flexion contractures. Laboratory tests such as haemoglobin level, leukocytes and platelets count, ESR, urine analysis as well as biochemical profile were within the normal range. The antinuclear antibody (ANA) test by IFI in HEp-2 was positive, presenting a speckled pattern, title of 1:160. Anti DNAtopoisomerase I antibody was positive, ENA and anti centromere were negatives. The roentgenologic barium examination of the oesophagus revealed a reduction in peristalsis. The echocardiography and a high resolution CT of the torax were normal. The variables assessed to define organ system involvement are shown in Table I (2). A CT scan of the cervical spine showed calcification of the posterior longitudinal ligament from C4 to T1, with reduction of the vertebral canal diameter and probable medullar compression (Fig. 1). Conservative treatment was chosen with analgesics, NSAIDS and muscle relaxants. Low dose warfarin was used for calcinosis. She was discharged with partial improvement of the painful symptoms, although with persistent weakness in her limbs.

In April 2002, the weakness and paresthesia became worse and a new CT scan of the cervical and thoracic spine was undertaken, which confirmed a reduction of the neural canal diameter and medullar compression. In June 2002, a posterior cervical-thoracic laminectomy was performed. Post-operatively, her condition progressively deteriorated and she died 3 months after the surgical procedure, due to spinal compression.

Table I. Variables assessed to define organ system involvement.

Skin	Modified Rodnan Score 34
Vessels	Raynaud and finger pad scarring were present
Joints	Finger flexion contractures on the hands
Tendons	No tendon friction rubs
Muscles	Proximal weakness with normal serum CPK
Gastrointestinal tract	Dysphagia; roentgenologic barium examination with reduction in peristalsis
Lung	Normal high resolution CT
Heart	Normal ecocardiography
Kidney	Arterial pressure, serum creatinine and urinalysis were normal

Discussion

Neurological manifestations are described in 0.8 to 18.5% of SS patients (3) namely abnormalities in cranial nerves, particularly in the trigeminal nerve; peripheral neuropathy; vasculitis of the central nervous system (CNS) and autonomic dysfunction. Frequently these neurological findings are secondary to compressive phenomena or are associated with Sjögren's syndrome. The primary involvement of the CNS in SS is rare and some authors have suggested that an abnormal production of collagen in the neurological tissue associated with microvascular disease can be involved in its pathogenesis (4). Additionally, calcinosis has also been reported as a rare cause for neurological signs and symptoms seen in SS. Generally, the type of calcinosis associated with connective tissue diseases is dystrophic (5), characterized by the presence of crystals of hydroxyapatite

and it is believed that a microvascular insufficiency and local tissue damage are the main factors involved in its pathogenesis (6). However, although this type of crystal may be responsible for the majority of the PSS cases with medullar compression, there are reports of para and intra-spinal calcifications secondary to chondrocalcinosis, in which the type of crystal is calcium pyrophosphate (7-10).

In the present case, the finding of calcifications in shoulders and para-vertebral areas on thorax x-ray, led us to believe in the possibility of extension of the calcification to the medullar canal as a cause for the neurological manifestations, which was confirmed by CT of the cervical spine. Unfortunately, we could not identify the type of crystal involved in the calcinosis of our patient, as the primary objective of the laminectomy was to decompress the medulla and consequently alleviate the symp-

toms, and a tissue sample for examination was not available.

Reviewing the literature we observed that only 11 other cases of SS with para and/or intra-vertebral calcification have been reported. In the majority of these cases the calcinosis was located in the cervical spine (5, 6, 11-14), although there are two reports of calcinosis in the thoracic section (15,16) and one in the lumbar section (6). In half of the reported patients a concomitant peripheral calcinosis was also described. The age of these patients varied from 45 to 78 years and the time of diagnosis varies from 4 to 27 years after the diagnosis of SS. In all cases there were clinical signs of medullar compression and they were submitted to surgical procedure with an improvement of the compressive and painful symptoms. Additionally, there were three cases of SS with intra-cerebral calcification and consequent cognitive and motor manifestations, which were considered as primary CNS features, after vascular aetiology was discarded (3, 17).

We emphasize here the importance of the investigation of calcinosis by imaging methods in SS patients complaining of cervicgia or dorsalgia, in view of the possibility of having compressive medullar lesions. The poor response to the clinical treatment as well as the high risk of the surgical procedure are responsible for the reserved prognosis.

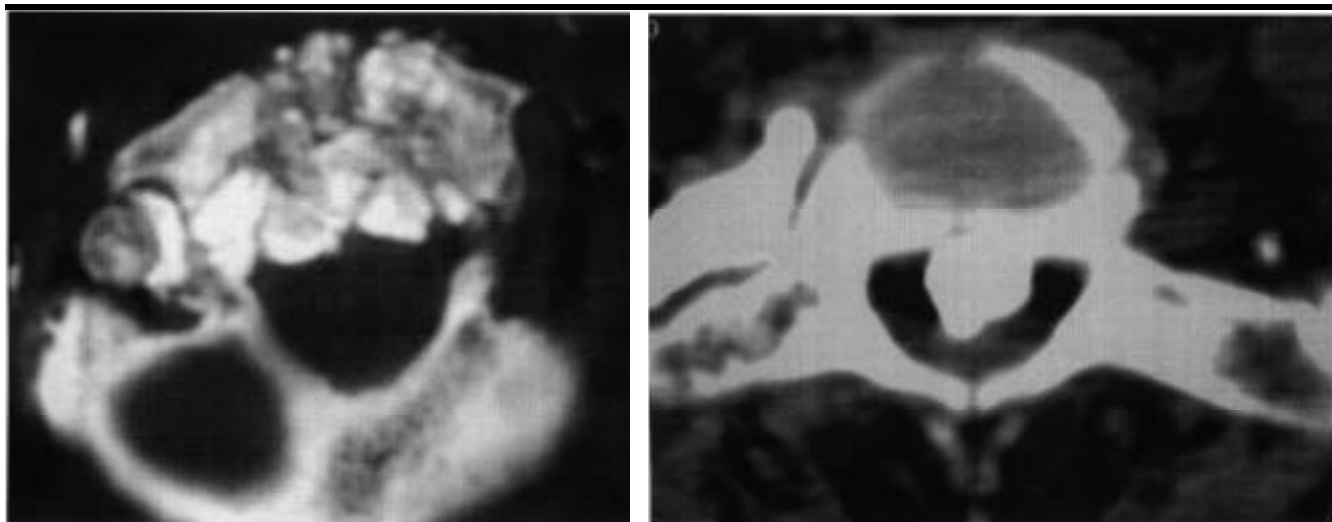


Fig. 1. Posterior calcification in the vertebrae bodies and posterior longitudinal ligament between C4 and T1, with reduction of the neural canal diameter and medullar compression.

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