Cervical spinal calcinosis in a patient with systemic sclerosis


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KEYWORDS
Systemic sclerosis; Calcinosis; Spine

A 67-year-old female patient was diagnosed with systemic sclerosis due to the existence of sclerodermiform skin, sclerodactyly, telangiectasia, calcinosis of the hands, Raynaud’s phenomenon, haemorrhagic ulcerated oesophagitis with antral gastritis, pulmonary fibrosis with severe restrictive lung disease, PAH of 60mmHg with signs of right ventricular insufficiency, mitral and aortic insufficiency and dilated cardiomyopathy. Results of autoimmunity tests were positive for antinuclear antibodies (1/500), anti-SSA and anti-Jo1 antibodies, and negative for antinative DNA and anticientromere antibodies. Renal function was normal, as were calcium and phosphate levels. There was microcytic anaemia of inflammatory origin. She was treated with bosentan, a calcium channel blocker, and 10 mg per day of prednisolone.

Five years after the diagnosis of systemic sclerosis, severe upper neck pain occurred radiating into the occipital region, associated with an inflammatory reaction (CRP 100 mg/l, ESR 105 mm in the first hour).

A cervical CT-scan showed a lesion with calcified density extensively developed and affecting the left atlanto-axial joint (Fig. 1). This lesion was exophytic, irregular and heterogeneous, resulting in destruction of the left cortical bone of the atlas. Cervical MRI found a large, lobulated, left paravertebral, circumferential mass, 2 cm thick, surrounding the C1–C2 joint, which was hypointense on T1- and T2-weighted and fat suppressed MR images. It did not enhance with contrast and was eroding the peripheral part of the left lateral mass of the atlas. This calcification was crossed by the vertebral artery, the canal of which was still intact (Fig. 2). Bone scintigraphy showed that the lesion had increased tracer uptake, unlike the scintigraphy performed 6 years earlier. The same was true for the left wrist. Hand and wrist radiographs confirmed the existence of extensive subcutaneous calcinosis (Fig. 3).

After suggesting the diagnosis of calcinosis and in order to exclude other conditions, a radiologically-guided needle biopsy with lavage was performed, with bacteriological and
histological examination. This found fibrinoid necrosis without any suspect cells suggesting malignancy, subjacent infection or micro-crystalline arthropathy (such as hydroxyapatite rheumatism).

Treatment was initiated with colchicine, resulting in regression of the cervical pain and inflammatory syndrome.

**Discussion**

Spinal calcifications are rare in scleroderma, particularly in the cervical spine.

Patients classically present with more or less intense local pain associated with stiffness [1—3]. The development time for scleroderma before the appearance of spinal calcification is variable. Very rarely it can be the first sign that leads to systemic sclerosis being suspected [4].

Spinal locations are most frequently described in diffuse cutaneous systemic sclerosis, but they can be encountered in a limited CREST form [5].

Standard radiographs particularly of the upper cervical region can be falsely normal at the onset of symptoms [2]. A CT-scan is the most appropriate examination for diagnosis and for studying spinal calcinosis [6], which presents as a large homogeneous, lobulated mass with calcified opacity, sometimes extending over several vertebral segments and developing at the expense of the vertebra [1,2,5]. A fluid level may possibly be seen within the mass [4]. Some cases involve osteolysis [1,5,7] which can be the source of structural and instability disorders. Finally, small calcifications can be found in adjacent soft tissue [5,7]. MRI is of use in evaluating neurological disorders related to invasion of the spinal canal by the calcinosis [6].

The histology of such lesions varies, with deposits either of hydroxyapatite [1] or amorphous calcified material with fibrotic tissue [4] or even osteonecrosis with calcified debris [7] or fibrinoid necrosis, as in our case. Biopsies are not necessarily helpful.

Spinal locations can be complicated by neurological involvement. Spinal cord compression due to prominent spinal and para-spinal calcification protruding into the spinal canal has been described several times [2,8—10]. In some cases, it is the very consequences of destructive calcification with osteolysis which are the cause of neurological disorders such as upper spinal cord compression due to instability [10]. As far as the cervical vasculature is concerned, there

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**Figure 1.** Cervical CT-scan, axial and coronal slices. Burgeoning, heterogeneous mass, with calcified opacity, that has developed at the expense of the left atlanto-axial joint resulting in osteolysis of C1 (a—b).

**Figure 2.** Cervical MRI. Paravertebral mass, eroding the left lateral mass of C1, hypointense with T1-weighting (axial, a) and T2-weighted fat sat (coronal, b).
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is sometimes displacement of the vascular routes within the calcified mass, which can be complicated by extrinsic compression and thus have neurological repercussions [2].

Conclusion

Cases of spinal calcinosis are rare in diffuse cutaneous systemic sclerosis. These masses usually appear as extensive, voluminous, multilobed, calcified lesions which are sometimes destructive and can mimic a neoplastic or infectious lesion. They are rarely isolated, and periarticular or peripheral soft tissue calcification is often present. They are responsible for spinal pain or stiffness, and sometimes for radiculalgia or even compression of the spinal cord or the arteries feeding the brain, in the case of a cervical location. These masses may evolve in certain cases into osteolysis, which can be responsible for disrupting spinal structure, causing instability and secondary neurological disorders. Surgical intervention requires complex techniques both for excising such masses and for the reconstruction that needs to follow.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References