Cervical and lumbar paraspinal calcinosis in systemic sclerosis

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SUMMARY

Calcinosis is a well-recognized manifestation of systemic sclerosis. Paraspinal or intraspinal calcinosis is rare, with reports of calcinosis involving the cervical, thoracic and lumbar separately, but not together. We now report a case of limited cutaneous scleroderma with extensive paraspinal calcinosis of the cervical and lumbar spine.

Key words: *Scleroderma; cervical calcinosis; lumbar calcinosis.*

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INTRODUCTION

Calcinosis is a well-recognized manifestation of systemic sclerosis, occurring in up to 25% of patients (1). It is most common in the hands, but can also occur around the elbows, shoulders or hips. Paraspinal or intraspinal calcinosis is rare, with reports of calcinosis involving the cervical, thoracic or lumbar spine separately (2-5), but not together. Here we report a patient with limited cutaneous scleroderma and with extensive paraspinal calcinosis involving the cervical and lumbar spine.

CASE REPORT

The patient, a 79-year old Chinese lady, was first seen in our clinic in February 2015, with two years history of progressive breathlessness. Her daughter reported intermittent 'red discolouration of the fingers', but the rest of the systemic enquiry was unremarkable. In particular, there was no history of other cardiorespiratory symptoms, dysphagia or altered bowel habits. Her other medical problems included hypertension, dyslipidaemia and osteoarthritis in her knees. Her regular medications included amlodipine, atenolol and lovastatin. She had never smoked, and been teetotal all her life.

On examination, her vital signs were satisfactory, with blood pressure of 140/80 mm Hg. The skin was taut over her face, with evidence of pinched nose and microstomia. There was evidence of sclerodactyly in her fingers, but no skin thickening in her proximal upper limbs, lower limbs or trunk. There were a few telangiectasia in her fingers, but no evidence of digital infarcts or pitting scars. The rest of the examination was unremarkable.

Her full blood count, liver function and renal function tests were satisfactory. Urinalysis showed evidence of microscopic haematuria and proteinuria, with albumin: creatinine ratio of 71.6 mg/mmol. Her serum anti-nuclear antibody was positive in a titre of 1/640, with a speckled pattern on immunofluorescence. Serum anti-double stranded DNA, antibodies to extractable nuclear antigens and anti-ScL-70 were negative. Testing for anti-centromere antibody would have required the specimen to be sent to the Mayo clinic. As this would have incurred a substantial cost, the family was not keen for the test to be done. Her pulmonary function test showed a restrictive pattern, and a high resolution computed tomography scan of her lungs showed subpleural cystic changes and possible traction bronchiectasis in the bases of her lungs, in keeping with usual interstitial pneumonia. Her two-dimensional transthoracic echocardiography revealed evidence of severe

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pulmonary hypertension, with pulmonary artery systolic pressure of 70 mm Hg and right ventricular overload. It was felt that a right heart catheterization was not worth pursuing because of her advanced scleroderma and its limited impact on her overall management.

Over the next three months, she became increasingly frail and dependent on her daughter for her activities of daily living. She was subsequently commenced on long-term home oxygen therapy because of worsening lung function. A swallowing assessment, with video fluoroscopy, was requested, as her daughter reported intermittent episodes of choking. The video fluoroscopy reported moderate oropharyngeal dysphagia but made no specific mention of the oesophageal motility. An incidental finding on the video fluoroscopy was the presence of a sclerotic mass at the level of C3-C5, but this was not compressing the oesophagus and was not thought to be the cause of her dysphagia.

Computed tomography scans of the neck, abdomen and pelvis showed evidence of exophytic and calcific bony masses behind the vertebrae at C3-5 and L3-5 levels (Figures 1 and 2). There was also evidence of periarticular soft tissue calcification, adjacent to the left hip joint and the right shoulder. Serum calcium (2.30 mmol/L), phosphate (1.17 mmol/L) and alkaline phosphatase (81 units/L) were all within the normal range.

The paraspinal as well as the periarticular calcinosis were presumed to be related to her systemic sclerosis. Surgical intervention was not considered, as she was asymptomatic from the paraspinal calcinosis, and there was no evidence of cord compression. Because of her poor mobility, declining functional status and severe breathlessness, she was referred to a home hospice team on discharge. Her prognosis was deemed reserved because of her severe pulmonary hypertension.

DISCUSSION AND CONCLUSIONS

There are reports of spinal calcinosis complicated by cord compression, radicular involvement and spinal stenosis (6-8), so physicians should consider spinal calcinosis in the differential diagnosis for scleroderma patients with weakness or radicular pain in the limb. Plain radiographs and computed tomography scans may reveal the calcinosis, but magnetic resonance imaging is preferable in patients with neurological symptoms (3). There are case reports and case series suggesting the usefulness of several medications such as calcium chan-

Figure 1 - Cervical paraspinal calcinosis.

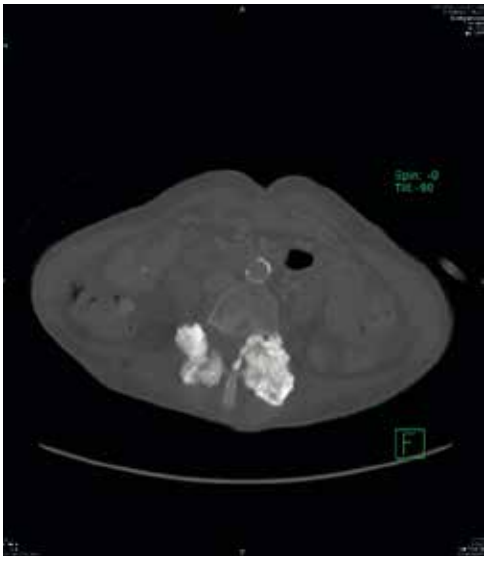

Figure 2 - Lumbar paraspinal calcinosis.

nel blockers, warfarin, bisphosphonates, probenecid, minocycline and rituximab (9-11), but for those with neurological involvement, surgical management may be indicated (12).

Conflicts of interest: Neither of the authors has any conflicts of interest to declare.

REFERENCES

1. Avouac J, Guerini H, Wipff J, et al. Radiological hand involvement in systemic sclerosis. Ann Rheum Dis. 2006; 65: 1088-92.
2. Ogawa T, Ogura T, Ogawa K, et al. Paraspinal and intraspinal calcinosis: frequent complications in patients with systemic sclerosis. Ann Rheum Dis. 2009; 68: 1655-6.
3. Bisson-Vaivre A, Somon T, Alcaix D, et al. Cervical spinal calcinosis in a patient with systemic sclerosis. Diagn Interv Imaging. 2013; 94: 645-7.
4. Ogawa T, Ogura T, Hayashi N, Hirata A. Tumoral calcinosis of thoracic spine associated with systemic sclerosis. J Rheumatol. 2009; 36: 2552-3.
5. Weerakoon A, Sharp D, Chapman J, Clunie G. Lumbar canal spinal stenosis due to axial skeletal calcinosis and heterotopic ossification in limited cutaneous systemic sclerosis: successful spinal decompression. Rheumatology (Oxford). 2011; 50: 2144-6.
6. Ward M, Cure J, Schabel S. Symptomatic spinal calcinosis in systemic sclerosis (scleroderma). Arthritis Rheum. 1997; 40: 1892-5.
7. Petrocelli AR, Bassett LW, Mirra J, et al. Scleroderma: dystrophic calcification with spinal cord compression. J Rheumatol. 1988; 15: 1733-5.
8. Lima IV, Galrao LA, Maia TS, Santiago MB. Spinal cord compression by ectopic calcinosis in scleroderma. Clin Exp Rheumatol. 2005; 23: 704-6.
9. Reiter N, El-Shabrawi L, Leinweber B, et al. Calcinosis cutis: part II. Treatment options. J Am Acad Dermatol. 2011; 65: 15-22, quiz 23-24.
10. Daoussis D, Antonopoulos I, Liossis SN, et al. Treatment of systemic sclerosis-associated calcinosis: a case report of rituximab-induced regression of CREST-related calcinosis and review of the literature. Semin Arthritis Rheum. 2012; 41: 822-9.
11. de Paula DR, Klem FB, Lorencetti PG, et al. Rituximab-induced regression of CREST-related calcinosis. Clin Rheumatol. 2013; 32: 281-3.
12. Smucker JD, Heller JG, Bohlman HH, Whitesides TE Jr. Surgical treatment of destructive calcific lesions of the cervical spine in scleroderma: case series and review of the literature. Spine. 2006; 31: 2002-8.