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# Coexistence of diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis

O.L. RILLO, E.J. SCHEINES, C. MORENO, J.C. BARREIRA, A.A. PORRINI, J.A. MALDONADO COCCO

Department of Rheumatology, National Institute of Rehabilitation, Buenos Aires, Argentina.

SUMMARY *To the best of our knowledge, only two patients with concurrent diffuse idiopathic skeletal hyperostosis (DISH) and ankylosing spondylitis (AS) have been reported so far. Here we present 3 patients in whom clinical and radiological findings indicative of DISH and AS coexisted. Two of these cases exhibited HLA B27. Although the presence of sacroiliitis would appear to exclude DISH, calcification and ossification of the anterior common vertebral ligament (ACVL) confirmed diagnosis of the latter disease.*

Key words: Ankylosing Spondylitis, Diffuse Idiopathic Skeletal Hyperostosis, Sacroiliitis, Computarized Tomography.

## INTRODUCTION

Forestier and Rotes Querol described in patients older than 50 years a disease featuring vertebral rigidity, invariably dorsal, often lumbar and mostly painless, accompanied by extensive calcification and ossification of the anterior common vertebral ligament (ACVL), of the fibrous ring belonging to the intervertebral disc and of the paravertebral connective tissue (1). Besides spinal involvement, Resnick et al reported peripheral hyperostotic lesions, which led them to term the entity diffuse idiopathic skeletal hyperostosis (DISH) (2). Ankylosing spondylitis (AS), a chronic inflammatory articular disease, mainly involves the vertebral column and its ligamentous component, with or without systemic manifestations, and occurs mostly in the young adult, particularly males. Radiologically, sacroiliitis is the conclusive diagnostic finding. Exceptionally, to the best of our knowledge, the coexistence of both pathologies has so far been reported in the English literature in only two isolated cases (3,4).

The purpose of this work is to describe 3 patients who fulfilled AS diagnostic criteria and also presented radiological findings indicative of DISH.

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Correspondence to: Dr. J. A. MALDONADO COCCO, Sección Reumatologia, Instituto Nacional de Rehabilitación, Echeverría 955, (1428) Buenos Aires, Argentina.

## CASE REPORTS

### Patient 1

This 55-year-old male first presented, at age 29, with progressive pain and rigidity in the dorso-lumbar region which improved with activity. Gradually kyphosis and limited movement in the cervical spine and both hips developed. There was no history of urethritis, diarrhoea, rash, conjunctivitis or iritis, and family background was unremarkable. The patient is a professional driver and with some adaptations in his vehicule is still able to work.

At physical examination, kyphotic posture was obvious and walking was normal, while the cervical and dorsal spine were globally limited; he had a 2 cm respiratory excursion,

a 2 cm Schobert test and a rectified lumbar column. At maximal flexion, fingers-to-floor distance was 44 cm. Sacroiliac articulations were painless and hips limited for internal rotation. The remainder of the osteo-articulomuscular findings proved normal. At functional respiratory examination, there was severe restrictive ventilatory insufficiency. Laboratory findings disclosed 47% hematocrit; 14.2 g% hemoglobin; 15 mm/h erythrosedimentation; 151 mg% glucemia (normal value up to 110 mg%); 224 mg% cholesterol and 60 mg% HDL cholesterol; 390 mg% triglycerides (normal value up to 160 mg%); negative latex and Rose Ragan tests. At radiology, the cervical spine showed ACVL calcification and ossification from $C_3$ to $C_6$, with preserved intervertebral discs (Fig. 1). In the dorsal region, there was right ACVL anterolateral calcification and ossification from $D_1$ to $D_{12}$, with spared intervertebral discs and radiolucence between ACVL and the anterior borders of the vertebral bodies in the lower portion. In the lumbar column, calcification and ossification was observed in ACVL from $L_3$ to $L_5$. Radiological examination of sacroiliac articulations disclosed grade IV sacroiliitis on the right and grade I on the left. Besides, there was hyperostosis in humeral diaphyses, iliac crests, trochanters, patellae, anterior tibial tuberosity and Achillean calcaneum region. Sacroiliac CT scanning showed, on the right, irregularities along articular borders with partial ankylosis in the lower third; on the left, there was evident sclerosis along articular borders.

**Patient 2**

On a 58-year-old male, this case started at the age of 25 with lumbar pain unrelieved by rest, which spread to the cervicodorsal region, with progressively kyphotic posture and

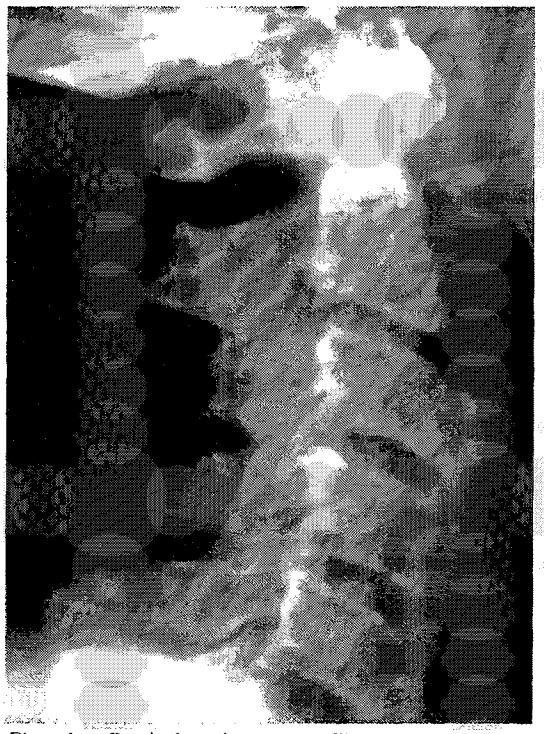

*Fig. 1:* Cervical column (profile view): ACVL calcification and ossification from $C_3$ to $C_6$. Spared intervertebral discs.

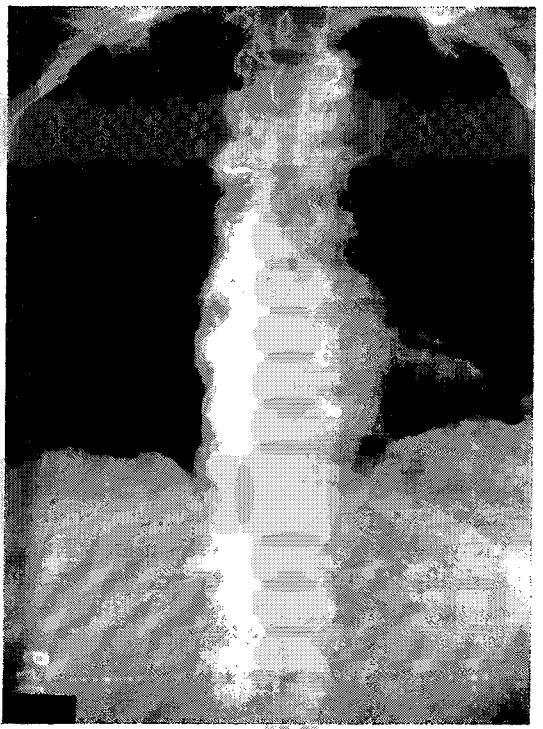

*Fig. 2:* Dorsal spine (front view): extensive ACVL calcification and ossification.

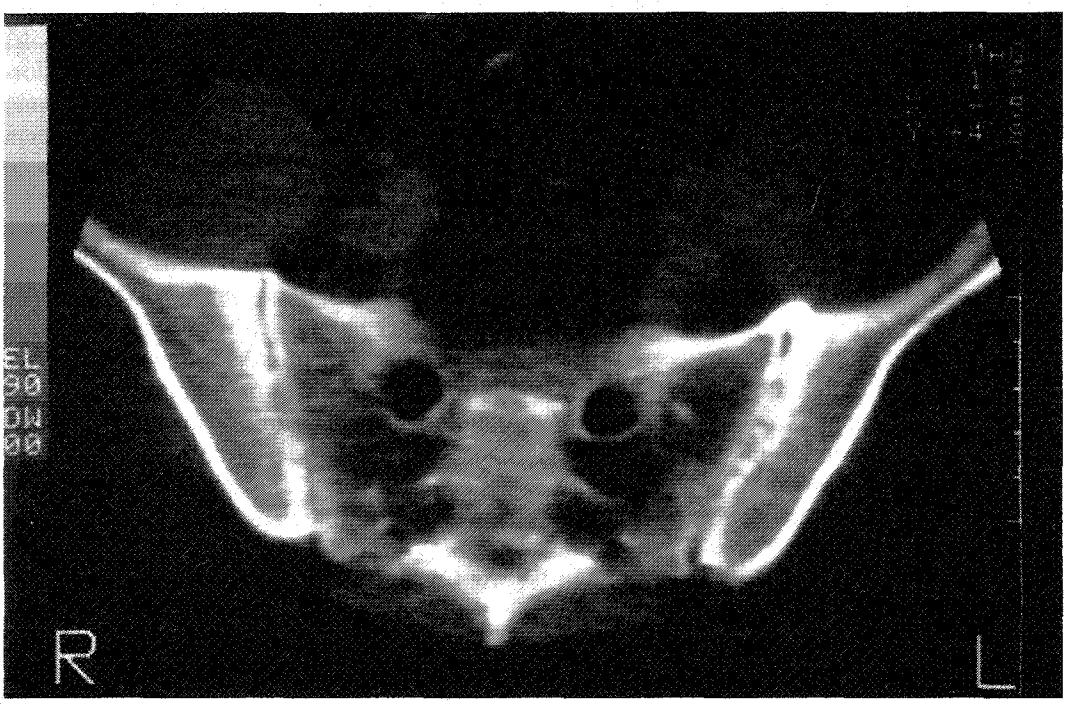


*Fig. 3:* CT scan of sacroiliac articulations: bilateral sclerosis and erosions. Ankylosis in the lower third of the right articulation.

marked movement limitation in these segments. There was no personal history of urethritis, diarrhoea, rash, conjunctivitis or iritis; neither was family history contributory. To date, the patient presents moderate limitations for daily activities. Physical examination showed a kyphotic posture with normal gait. Cervical, dorsal and lumbar columns were slightly painful and markedly impaired as to mobility. Thoracic excursion was 2 cm, with a rectified lumbar column and a 2 cm Schobert test. Fingers-to-floor distance in maximal flexion was 40 cm. Sacroiliac joints were painless and hips normal. Remaining osteo-articulo-muscular tests proved normal. Laboratory data were as follows: 45% hematocrit; 15 g% hemoglobin; 24 mm/h erythrosedimentation; 79% glucemia; negative latex and Rose Ragan tests; and positive HLA B27.

Vertebral column radiological examination disclosed uninterrupted ACVL calcification and ossification from $C_3$ down to $L_4$ (Fig. 2), together with grade III sacroiliitis on the right side and grade I on the left. Hyperostosis was observed in iliac crests, trochanters, ischia and Achillean calcaneum region. At CT scanning, sacroiliac articulations showed bilateral sclerosis and erosions with bone ankylosis in the lower half of the right articulation (Fig. 3).

**Patient 3**

This 61-year-old male was referred due to recurrent acute anterior uveitis, with a history of lumbalgia from age 51, accompanied by axial morning stiffness which improved with exercise and which failed to interfere with everyday activity. He had no history of urethritis, diarrhoea or rash, and no relevant family background. At physical examination, there was global impairment in lumbar mobility and in thoracic excursion, with loss of lumbar curvature. Fingers-to-floor distance at max-

imal flexion was 20 cm, with a 3 cm Schobert test and a 2 cm thoracic excursion. Laboratory data were as follows: 45% hematocrit; 14 g% hemoglobin; 10 mm/h erythrosedimentation; 95 mg% glucemia; 230 mg% cholesterol and 62 mg% HDL cholesterol; and 110 mg% triglycerides. Latex and Rose Ragan tests, as well as HLA B27, were all negative. Spinal radiology disclosed extensive ACVL calcification from $D_5$ to $D_{12}$ and in segments $L_1$, $L_2$ and $L_4$, with spared intervertebral discs and radiolucence between the vertebral body and ligamentous calcification in lower dorsal segments.

At X-ray studies in Fergusson position, bilateral grade III sacroiliitis was observed. Other findings included bilateral hyperostosis in ischia, pubis, iliac crests and plantar fascia insertion, as well as in Achilles tendon. Sacroiliac CT scanning showed articular border irregularities and partial ankylosis in the lower sector of both articulations.

## DISCUSSION

As a rule, DISH patients are mostly males over 50 years of age who show tendinous and ligamentous calcification and ossification. In roughly 40% the disease is associated to non-insulin dependent diabetes mellitus (5), as happened in one of our cases.

To establish a firm diagnosis of DISH three basic radiological criteria have been proposed by several authors (6-8): 1) anterolateral ligamentous calcification and ossification of at least 4 adjacent vertebral bodies, in the absence of significant radiological alterations of axial degeneration; 2) absence of ankylosis in interapophyseal articulations; and 3) absence of sclerosis, erosions or ankylosis in sacroiliac articulations.

Criteria 2) and 3) are useful to distinguish AS from DISH. AS is a chronic inflammatory disease that habitually presents in the young adult, courses with negative rheumatoid factors and is associated to HLA B27 positivity in over 90% of patients (9). Briefly, its clinical and radiological criteria (10) are given below: a) lumbar spine mobility limitation in extension and anterolateral flexion; b) history or presence of pain at the dorsolumbar junction or in the lumbar column; c) thoracic expansion limitation, lower than 2.5 cm; and d) unilateral or bilateral sacroiliac sclerosis, erosions and/or ankylosis.

Among DISH patients, radiological alterations in sacroiliac articulations are found in the upper third and may present as ankylosis; on the other hand, the lower third of this articulation, which possesses a synovial membrane, is usually normal or merely displays spatial narrowing and uniform cortical sclerosis as a result of degenerative changes (11). On occasion, ossification anterior and posterior to the sacroiliac articular capsule is observed (11,12). All such findings may be confused with sacroiliitis typical of AS.

Given their anatomical complexity, conventional radiology of sacroiliac articulations is not always sufficient for the differential diagnosis of these two entities. CT scanning provides images free of bone and soft tissue superposition, offering an ideal method to evaluate this articulation (13). In our 3 patients there was CT evidence of sacroiliac alterations compatible with AS, such as articular interlinear irregularity, iliac sclerosis, erosions and partial ankylosis in the lower third, findings already described by other authors (14,15). However, in DISH cases no subchondral bone erosions or intra-articular bone ankylosis in the lower third can be observed (11,16).

ACVL calcification in DISH is radiologically similar to that seen in spondylitis associated to psoriasis and Reiter's syndrome. In these diseases, syndesmophytes may induce reactive changes in the ACVL and adjacent paravertebral tissue leading to gross calcification and ossification in such structures (17,18). None of our 3 patients had a history of psoriasis or any symptoms or signs of Reiter's syndrome. Cases reported herein showed ACVL calcification typical of DISH. Patient 3 failed to present the B27 histocompatibility antigen which occurs in roughly

10% of AS cases. Although the late onset of AS symptoms was noteworthy, the presence of bilateral grade III radiological sacroiliitis, confirmed at CT scanning and recurrent acute anterior uveitis, grounded the diagnosis of this inflammatory disease.

Sacroiliac articulation CT has thus become a most useful complementary examination for the differential diagnosis of DISH and AS.

The existence of these 3 cases, plus the other 2 previously reported (3,4), shows that AS and DISH may infrequently course concurrently, posing on occasion troublesome differential diagnosis due to clinico-pathological overlapping. An alternative explanation is that some AS patients undergo atypical radiological evolution of the disease with gross ACVL calcification and ossification capable of mimicking DISH.

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