Concise Report

Diffuse idiopathic skeletal hyperostosis may give the typical postural abnormalities of advanced ankylosing spondylitis

I. Olivieri, S. D’Angelo, M. S. Cutro, A. Padula, G. Peruz, M. Montaruli, E. Scarano1, V. Giasi, C. Palazzi2 and M. A. Khan3

Objectives. To describe a case-series of patients who presented with the typical postural abnormalities of long-standing advanced ankylosing spondylitis (AS) but were instead found to suffer from diffuse idiopathic skeletal hyperostosis (DISH).

Methods. We enrolled consecutive patients who showed postural abnormalities, which at first suggested to us the diagnosis of long-standing advanced AS, although the diagnostic process led us to the correct diagnosis of DISH. Each patient had a complete physical examination and radiographs of the spine and pelvis, and was investigated for HLA-B27 locus typing.

Results. From 15 June 1998 to 15 June 2006, 15 patients with DISH were seen who presented with the typical postural abnormalities of long-standing advanced AS. All patients were males with a median age of 69 yrs (range 51–91). All lacked HLA-B27 and denied personal or family history of spondyloarthritis. All measurements assessing cervical, thoracic and lumbar spinal movement were abnormal.

Conclusions. Patients suffering from DISH can occasionally have severe limitations of spinal mobility, along with postural abnormalities that resemble long-standing advanced AS. Thus, the differential diagnosis between DISH and advanced AS is not limited to the radiological findings and can also extend to the clinical findings in the two diseases, as is highlighted by our report.

KEY WORDS: Diffuse idiopathic skeletal hyperostosis, Ankylosing spondylitis, Differential diagnosis.

Ankylosing spondylitis (AS) and diffuse idiopathic skeletal hyperostosis (DISH) are two different diseases sharing the involvement of axial skeleton and peripheral entheses [1, 2]. Symptoms of AS start at a young age, usually in late adolescence and early adulthood, and consist of inflammatory spinal pain and stiffness, decreasing range of spinal motion, and after many years the illness can result in characteristic postural abnormalities (‘Bechterew stoop’). In contrast, DISH, also known as ankylosing hyperostosis, affects middle-aged and elderly persons and is often asymptomatic, or is associated with mild dorso-lumbar pain and/or some restriction of spinal mobility.

DISH has long been considered a radiographic entity with minor and non-significant clinical manifestations. Till some years ago, the differential diagnosis between AS and DISH was limited to the radiological aspects since both diseases produce bone proliferation (hyperostosis) in the spine and at extraspinal entheseal sites.

In 1998, we came across a patient (patient # 1) with postural abnormalities, which at first suggested to us the diagnosis of long-standing advanced AS, although further evaluation led us to the correct diagnosis of DIHS. This observation led us to describe a case-series of similar patients with the aim to better elucidation of this under-recognized aspect of the differential diagnosis between the two diseases.

Patients and methods

For the present study, we enrolled all consecutive patients seen for the first time in our three out patient clinics, who showed the postural abnormalities typical of long-standing advanced AS but subsequently were diagnosed to be suffering from DISH. Patients should have had forward stooping of the neck, high dorsal kyphosis, rounding of the shoulders, obliteration of the normal lumbar lordosis, wasting of the buttocks, flattening of the chest and ‘ballooning’ of the abdomen.

The study was approved by the local ethics committees and written consent was obtained from each enrolled patient according to the Declaration of Helsinki.

Each patient was questioned about clinical symptoms of spondyloarthritides (SpA) including inflammatory spinal pain, buttock pain, peripheral arthritis, peripheral enthesitis, dactylitis, chest wall pain, ocular symptoms of conjunctivitis and uveitis, cutaneous and gastrointestinal symptoms, and urogenital or enteric infections. Patients were also questioned about family history of SpA and HLA-B27 associated diseases.

Each patient had a complete physical examination, including peripheral joints, peripheral entheses and spine. Measures of spinal mobility included:

(i) Schober test: the examiner marked a point midway along the superior iliac spines. A second point was marked 10 cm vertically above when the patient was standing erect. Following maximal forward flexion of the spine, the aforementioned distance was reassessed and considered normal if the difference was >5 cm.

(ii) Finger-to-floor distance: distance between the tip of the middle finger and the floor following maximal lumbar lateral flexion with knees extended.

(iii) Lumbar lateral flexion: distance between the tip of ipsilateral middle finger and the floor following maximal lumbar lateral flexion, with both feet on the floor, knees extended and without rotation.

(iv) Chest expansion: the difference in centimetres between full expiration and full inspiration, measured at the 4th intercostals space.

(v) Occiput-to-wall distance: horizontal distance between occiput and wall while patient stands with heel and buttock against the wall.

(vi) Flexion and extension of the cervical spine: distance between the chin and the jugular notch of manubrium sterni in full flexion and in full extension, respectively.

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Each patient was also studied for complete blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), uric acid, glucose, urinalysis and tested for HLA-B27. Each patient had a pelvic radiograph (an anteroposterior view) and also radiographs of the spine. The diagnosis of DISH was made according to the criteria suggested by Resnick et al. [3, 4]:

(i) the presence of flowing ossification along the anterolateral aspect of at least four contiguous vertebral bodies;
(ii) the presence of relative preservation of the intervertebral disc height in the involved vertebral segment and the absence of radiographic changes of degenerative disc disease;
(iii) the absence of apophyseal joint bone ankylosis and sacroiliac joint sclerosis, erosion and fusion.

In order to obtain reference values for spinal mobility measures, a control group of age- and sex-matched healthy subjects was enrolled. In all control subjects radiographs of the spine and pelvis were performed.

Results

In the period extending from 15 June 1998 to 15 June 2006, 15 patients with the typical postural abnormalities of AS due to DISH were seen. All enrolled patients suffered from mild mechanical spinal pain. The reason for the consultation was long-standing limitation of spinal mobility.

All patients had four or more contiguous vertebrae bridged as a minimum criterion of DISH. No patient presented radiological evidence of sacroiliitis, squaring of vertebral bodies, syndesmophytes, spondylodiscitis, apophyseal joint sclerosis and ankylosis, or prominent reduction of intervertebral disc spaces. All patients showed ‘flowing mantles’ of ossification of the anterior longitudinal ligament (Figs 1 and 2).

All patients were males with a median age of 69 yrs (range 51–91). Acute phase reactants were normal in all patients except one. All lacked HLA-B27 and denied personal or family history of SpA.

The control subjects, all males with a median age of 69 yrs (range 47–84), did not show any DISH features but only signs of spondylosis on radiographs of the spine and pelvis.

In DISH patients, all measurements assessing cervical, thoracic and lumbar spinal movements were abnormal in comparison with control subjects (Mann–Whitney U-test, \( P < 0.05 \)). In particular, the median values of spinal measures, recorded in DISH patients and controls, respectively, were as follows: cervical flexion (jugular notch to chin distance) 7.0 cm (range 0.0–10.0) vs 3.0 cm (range 0.0–5.0); cervical extension (jugular notch to chin distance) 15.0 cm (range 13.0–22.0) vs 19.0 cm (range 16.0–24.0); occiput-to-wall distance 10.0 cm (range 3.0–19.0) vs 3.0 cm (range 0.0–9.5); Schober test 1.5 cm (range 0.5–5.0) vs 5.0 cm (range 4.0–5.5); right lateral flexion 58.0 cm (range 53.0–65.0) vs 52.0 cm (range 41.0–60.0); left lateral flexion 56.0 cm (range 52.0–65.0) vs 53.0 cm (range 37.0–58.0); chest expansion 2.0 cm (range 1.0–4.0) vs 4.5 cm (range 4.0–6.0); and finger-to-floor distance 32.0 cm (range 15.0–40.0) vs 12.0 cm (range 0.0–28.0).

With regard to comorbid conditions, five patients (# 3, 5, 9, 10, 12) had diabetes mellitus, five (# 6, 7, 8, 12, 15) had dyslipidaemia, one (# 4) had severe obesity (body mass index = 35), two (# 2, 15) had coronary artery disease and one (# 7) had multinodular goitre.

One patient (# 1) suffered from dysphagia caused by large anterior longitudinal ligament ossification encroaching the pharynx and the oesophagus.

Discussion

DISH has for a long time been considered a radiographic entity with less importance given to clinical signs and symptoms than other spinal diseases. In general this is true. However, patients with DISH may have spinal pain and marked decrease of spinal mobility. Occurrence in DISH patients of marked reduction in spinal mobility and greater physical disability when compared with healthy subjects has recently been emphasized by Mata et al. [5].

Spinal involvement of DISH is characterized by ‘flowing’ ossification of the anterior longitudinal ligament typically separated from the anterior aspect of the vertebral body by a thin radiolucent line [2]. The thoracic region is predominantly...
affected but any or all levels may be involved. To differentiate DISH from other spinal disorders including AS, spondylosis deformans and intervertebral osteochondrosis, Resnick et al. [3, 4] suggested the aforementioned criteria for the diagnosis of DISH. According to Arlet and Mazière [6], the involvement of three contiguous vertebral bodies at the lower thoracic level is sufficient for the diagnosis of DISH. Sacroiliac capsular bridging has been described in patients with DISH [7, 8]. This may give the false appearance of obliteration of the sacroiliac joint space that occurs in patients with AS on the pelvic X-ray anteroposterior view. In these cases, computed tomography shows intact sacroiliac joint space and presence of anterior capsular bridging due to capsular ossification.

On the contrary, axial involvement in AS is characterized by vertebral body squaring, Romanus lesion, syndesmophytes, spondylodiscitis, apophyseal joint sclerosis and ankylosis, and sacroilitis [9]. The radiological findings of axial involvement of AS and DISH are so different that in patients with the coexistence of the two diseases it is possible to identify the changes due to each disease at any level [10–12].

Clinical presentation of DISH patients with severe spinal limitation and postural abnormalities resembling long-standing advanced AS that we have highlighted in this report have not been previously reported to the best of our knowledge. All patients enrolled in the present study showed clinical and postural abnormalities that initially made us suspect the presence of AS. However, none of the reported patients had sacroiliac joint changes compatible with AS. This was the prime indication that AS was not the correct diagnosis; the presence of changes more characteristics of DISH (i.e. ossification at the anterior longitudinal ligament with a radiolucent line between the new bone and the adjacent vertebral body) and the absence of vertebral squaring provided strong supportive evidence.

In conclusion, our study suggests that patients suffering from DISH can have severe limitation of spine mobility and associated postural abnormalities typical of long-standing advanced AS. Our report suggests that the differential diagnosis between DISH and long-standing advanced AS is not limited to the radiological findings and can also extend to the clinical aspects.

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