

Atypical case of diffuse idiopathic skeletal hyperostosis mimicking ankylosing spondylitis in a young woman with polycystic ovary syndrome

Ong Swee Gaik, MRCP, Ding Hui Jen, MRCP

Rheumatology Unit, Department of Medicine, Kuala Lumpur Hospital, Ministry of Health Malaysia, Kuala Lumpur, Malaysia

SUMMARY

Both diffuse idiopathic skeletal hyperostosis and ankylosing spondylitis present with similar clinical manifestations of restricted spinal mobility and postural abnormalities, and radiographic resemblances including axial spine involvement and enthesopathy. Nonetheless, they are two entirely different diseases. We report an unusual case of DISH in a young woman whose diagnosis was established based on radiologic features. This case report aims to highlight the under-recognised radiologic aspects of the differential diagnosis between DISH and AS in order to avoid an inaccurate diagnosis.

INTRODUCTION

Both diffuse idiopathic skeletal hyperostosis (DISH) and ankylosing spondylitis (AS) demonstrate bone proliferation in the spine and extraspinal enthesal sites. DISH typically affects men and present in the fifth or sixth decade of life, while symptoms of AS start at a young age, typically in late adolescence and early adulthood. Both can present with postural abnormalities, spine stiffness and restricted spinal mobility. Symptoms of DISH are generally mild despite its remarkable radiographic manifestations. On the contrary, patients with AS may present with inflammatory back pain which is debilitating. Recognition of characteristic radiologic differences between these two entities is imperative to avoid diagnostic confusion and inappropriate therapies.

CASE REPORT

A 41-year-old woman was referred to the rheumatologist at Hospital Kuala Lumpur for management of AS in view of limited neck movement and presence of syndesmophytes on cervical spine radiograph. She had hypertension at the age of 35 years old, was obese with a body mass index of 36.5kg/m², had impaired glucose tolerance and polycystic ovary syndrome (PCOS). She gave a 6-year history of progressive neck stiffness with limited neck movements. She denied early morning stiffness of the spine, inflammatory spinal pain or buttock pain; and had not required analgesics. Clinical examination revealed a stooped posture with severely restricted mobility of the entire spine. Cervical spine movement in all planes was negligible, modified Schober's test was 3cm (normal: >5cm) and chest expansion was 1cm (normal: >5cm). Lateral flexion of the lumbar spine revealed

a finger-to-floor distance of 2cm (normal: >10cm). Tragus-to-wall distance was increased at 23cm (normal: <10cm). Peripheral arthritis was absent.

Cervical spine radiograph (Figure 1a) showed bridging syndesmophytes from C2 to C5 vertebrae and marked ossification of the posterior longitudinal ligament. There was "flowing mantle" of ossification anterior to the vertebral bodies extending inferiorly from C5 vertebra. Interestingly, there was no facet joint ankylosis. Lumbosacral spine radiograph showed irregularly pointed bony excrescences at the superior and inferior margins of the L4 vertebra which were linked by new ossification along the vertebral body (Figure 1b). Radiolucency was visible between the new ossification and subjacent vertebral body. Pelvic radiograph (Figure 1c) showed ossifications at both iliolumbar ligaments, prominent osseous proliferation and bony excrescences at the iliac crests, ischial tuberosities, greater trochanters and lesser trochanters. As expected, the sacroiliac joints appeared intact on pelvic radiograph. Given the differential diagnosis of AS, further evaluation of sacroiliac joints using magnetic resonance imaging (MRI) was undertaken. As expected, MRI did not reveal any abnormality in the sacroiliac joints. In conclusion, these imaging findings were consistent with DISH, rather than AS. In addition, the diagnosis of DISH was supported by a negative HLA-B27 test.

DISCUSSION

It is not uncommon for a patient suffering from DISH to be misdiagnosed as AS in view of similar clinical manifestations and radiologic findings. This case highlights the under-recognised radiologic aspects of the differential diagnosis between DISH and AS in order to avoid an inaccurate diagnosis. DISH and AS both demonstrate bone proliferation in the spine and extraspinal enthesal sites. However, the manner and characteristics of bone proliferation are not alike.

Resnick and Niwayama¹ had reported radiographic spinal features in DISH which included flowing calcification and ossification along the anterolateral aspect of vertebral bodies extending over at least four contiguous vertebrae, which may occur in any one or more segments of the spine. This finding had been included in the classification criteria of DISH. The middle and lower segments of the thoracic spine are

This article was accepted: 4 July 2019

Corresponding Author: Dr. Ong Swee Gaik

Email: ongsweeg@gmail.com

Table I: Radiographic features differentiating Diffuse idiopathic skeletal hyperostosis (DISH) from Ankylosing spondylitis (AS) in our patient

| Diffuse idiopathic skeletal hyperostosis (DISH) | Ankylosing spondylitis (AS) |
|--|---|
| "Flowing mantles" of ossification anterior to vertebral bodies. Large, coarse bony proliferations at the entheses, namely iliac crests, ischial tuberosities, lateral acetabulum, and greater and lesser trochanters. Ossification of iliolumbar ligaments. No sacroiliitis or facet joint ankylosis. | Syndesmophytes are thinner and oriented vertically. Bony proliferations at entheses are more muted. Iliolumbar ligaments are not affected. Sacroiliitis and facet joint ankylosis. |



Fig. 1: A) Lateral view of the cervical spine radiograph showing bridging syndesmophytes (bold arrow) from C2 to C5 vertebrae together with ossification of the posterior longitudinal ligament (arrowhead). "Flowing mantle" of ossification (arrow) anterior to the vertebral bodies extend inferiorly from C5 vertebra. B) Lateral view of the lumbosacral spine radiograph showing irregular pointed bony excrescences at the superior and inferior margins of L4 vertebra which were linked by new ossification along the vertebral body. A linear area of radiolucency (arrowhead) was visible between the new ossification and subjacent vertebral body. C) Anteroposterior view of the pelvic radiograph depicting ossification at both iliolumbar ligaments (arrow). Large bony excrescences are noted on the iliac crests (arrowhead), ischial tuberosities, and the greater and lesser trochanters. The sacroiliac joints appear normal.

reportedly most commonly involved. Subjacent radiolucency and pointed bony excrescences are other imaging features described in DISH.

Even though ossification of the posterior longitudinal ligament (OPLL) had reportedly been associated with DISH,² we would like to emphasise that this finding is not unique to DISH. OPLL had been observed as an isolated finding in certain individuals, as well as in patients with AS and other spondyloarthropathies.³

In AS, the most distinctive and early finding occurs in the sacroiliac joints. The well-known pre-radiographic phase of AS where radiography has limited capacity in detecting early sacroiliitis has led to the utilisation of MRI to identify early cartilage abnormalities and bone marrow oedema in early sacroiliitis.⁴ The presence of intact sacroiliac joint space in our patient's MRI clearly refuted the diagnosis of AS. In addition, calcification and ossification in the iliolumbar ligaments in her pelvic radiograph were congruent with features described in DISH.

Enthesopathies, which often are ossified, can occur in both DISH and AS. The sites of enthesal ossification together with its coarse nature by way of large, symmetrical and exuberant excrescences as observed in our patient's pelvic radiograph,

are highly characteristic of DISH. On the contrary, ossified enthesopathies in AS are more muted.

Table I depicts the radiographic features differentiating DISH from AS as observed in our patient.

We believe the presence of syndesmophytes in the cervical radiograph of our patient may have contributed to the misdiagnosis of AS. Syndesmophyte has been described as the hallmark of spinal disease in AS. Nevertheless, syndesmophyte in AS hardly ever occurs in the absence of significant sacroiliitis.⁵ Since the MRI of our patient did not demonstrate sacroiliitis and her other radiographic features are inconsistent with AS, it is possible that syndesmophyte can occur in DISH albeit it is uncommon and under-recognised.

Other factors that contributed to an increased risk of misdiagnosis of AS includes her gender and the presentation of disease at a younger age, both of which are clearly atypical of DISH. Nonetheless, she had metabolic derangements such as hypertension, obesity and impaired glucose tolerance, associations that are well-known to DISH. Apart from DISH, our patient also had concomitant PCOS. It is highly possible that PCOS may have been a chance occurrence. Indeed, PCOS has been linked to hyperinsulinaemia, insulin

resistance and obesity. The exact mechanisms resulting in new bone formation in DISH remain unclear. It has been postulated that certain metabolic factors such as insulin-like growth factor 1 and insulin, may stimulate bone formation. To the best of our knowledge, the occurrence of DISH in patients with PCOS has not been reported.

CONCLUSION

The cognisance and recognition of characteristic radiographic differences between DISH and AS are crucial to avoid misdiagnosis and inappropriate therapies.

FUNDING

This work received no specific grant from any agency.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

REFERENCES

1. Resnick D, Niwayama G. Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* 1976; 119(3): 559-68.
2. Resnick D, Guerra J Jr, Robinson CA, Vint VC. Association of diffuse idiopathic skeletal hyperostosis (DISH) and calcification and ossification of the posterior longitudinal ligament. *Am J Roentgenol* 1978, 131(6): 1049-53.
3. Ramos-Remus C, Russell AS, Gomez-Vargas A, Hernandez-Chavez A, Maksymowych WP, Gamez-Nava JJ, et al. Ossification of the posterior longitudinal ligament in three geographically and genetically different populations of ankylosing spondylitis and other spondyloarthropathies. *Ann Rheum Dis* 1998; 57(7): 429-33.
4. Braun J, Bollow M, Eggens U, König H, Distler A, Sieper J. Use of dynamic resonance imaging with fast imaging in the detection of early and advanced spondylitis in spondyloarthropathy patients. *Arthritis Rheum* 1994; 37(7): 1039-45.
5. Østergaard M, Lambert RGW. Imaging in ankylosing spondylitis. *Ther Adv Musculoskel Dis* 2012; 4(4): 301-11.