CASE REPORT

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

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SUMMARY
Both diffuse idiopathic skeletal hyperostosis (DISH) and ankylosing spondylitis (AS) demonstrate bone proliferation in the spine and extraspinal entheseal 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.

INTRODUCTION
Both diffuse idiopathic skeletal hyperostosis (DISH) and ankylosing spondylitis (AS) demonstrate bone proliferation in the spine and extraspinal entheseal 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 36.5 kg/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. Lumbar 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 entheseal 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

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

<table>
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<tr>
<th>Diffuse idiopathic skeletal hyperostosis (DISH)</th>
<th>Ankylosing spondylitis (AS)</th>
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<td>&quot;Flowing mantles&quot; 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.</td>
<td>Syndesmophytes are thinner and oriented vertically. Bony proliferations at entheses are more muted. Iliolumbar ligaments are not affected. Sacroiliitis and facet joint ankylosis.</td>
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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.

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. 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.

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CONFLICT OF INTEREST STATEMENT
The authors have no conflicts of interest to declare.

REFERENCES