Cervical Cord Injury in an Elderly Man with a Fused Spine - a Case Report

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ABSTRACT
We report a case of an elderly man presenting with co-existing diffuse idiopathic skeletal hyperostosis (DISH) and ossified posterior longitudinal ligament (OPLL) resulting in central cord syndrome. Only three such cases have been reported co-existing with DISH. The patient recovered most of his neurological deficit through conservative management. A discussion on the radiological features of DISH co-existing with OPLL and how these differ from ankylosing spondylitis (AS) follows.

Keywords: Forestier's disease, OPLL, enthesopathy, spinal cord compression

INTRODUCTION
Cervical spine injuries leading to neurological sequelae as a result of low energy falls are fairly common occurrences in the geriatric age group. These patients have pre-existing degenerative cervical spine changes that predispose them to impingement of the cervical cord. This is the result of compression of the spinal cord by lax infolding and hypertrophied ligamentum flavum, prolapsed intervertebral discs and posterior vertebral body bone spurs or osteophytes. The most common presentation is incomplete cord injury with central cord syndrome, being the most common. A part from cervical spondylosis, other neck conditions that pose a higher risk for cervical cord injuries include ossification of posterior longitudinal ligaments, ossified yellow ligaments, fractures in a cervical spine affected by ankylosing spondylitis and diffuse idiopathic skeletal hyperostosis (DISH) or Forestier’s disease. We report a case of central cord syndrome occurring in an elderly gentleman who has pre-existing double pathology in his fused cervical spine.

CASE REPORT
A 72-year-old Chinese male with hypertension on medication and a history of neck injury several years before was admitted through the Accident & Emergency Department. He had fallen in a coffee shop half an hour earlier and had loss of consciousness of 10 minutes’ duration. He complained of numbness of his whole body from the neck down, and weakness of all four extremities.

On examination, he was alert, Glasgow Coma Scale 15. He had diaphragmatic breathing. There was no cranial nerve palsy. Palpation of his neck revealed mild tenderness over the posterior aspect of the upper cervical levels. Neurological examination revealed Grade 3 (MRC) muscle power in the lower limbs and Grade 4 in the upper limbs. Reflexes were normal in the upper limbs, but he was hyperreflexic in the lower limbs. Plantar responses were down going. He had diminished sensation to pin prick and light touch from the neck down. He had a normal anal tone and...
sphincter reflex. There were no other significant findings. X-rays of his cervical spine showed bony ankylosis from C1 to C7 (Fig. 1a). There was also wavy ossification of the posterior longitudinal ligament extending from C2 to C4. No fractures were noted. His thoracic and lumbosacral spine X-rays also showed ossification of his anterior longitudinal ligament (Fig. 2). His sacroiliac joints appeared normal (Fig. 3).

The patient’s neck was protected with a hard collar. He was given oxygen and catheterised. The full blood count, urea and electrolytes were normal. Intravenous methyl prednisolone was started immediately in accordance to the Second National Acute Spinal Cord injury study protocol(1).

In view of the neurological findings, an MRI was done and this revealed C3/C4 disc prolapse associated with bony bar compressing the cord giving rise to signal changes (Fig. 1b). A diagnosis of central cord syndrome from a soft disc prolapse in an ankylosed spine was made. Six hours after his injury, neurological review showed that he had regained full motor power in both his lower limbs. There was residual weakness of right upper limb grip, and the numbness persisted. By Day 10 post-injury, the patient was able to stand with assistance. Three weeks after his injury, the patient was able to ambulate with a walking frame and was transferred to a community hospital for further rehabilitation. The HLA-B27 was negative and erythrocyte sedimentation rate (ESR) normal. The patient was discharged after three weeks of inpatient rehabilitation. Three months after his injury, he was walking independently with a quad stick. Motor power was full in all four extremities, but there was still hyper-reflexia. His sensory deficit to pin prick had recovered but light touch remained diminished.

DISCUSSION
Diffuse idiopathic skeletal hyperostosis (DISH or Forestier’s disease) is a fairly common spine condition which presents more commonly in elderly men. It has an estimated prevalence rate of up to 10% (2). This condition can affect any part of the spine. The thoracic spine is most commonly involved followed by the cervical spine. DISH is often discovered incidentally, and symptoms if present are usually mild. DISH is diagnosed when non-marginal syndesmophytes are present on X-rays at three or more successive vertebral levels. DISH may be associated with extraspinal ossification in large joints and increased heterotopic ossification following joint replacement surgery.

Radiographically, the features in the spine may superficially resemble ankylosing spondylitis (AS), as both appear “bamboo-like”. However there are significant differences. DISH shows typical thick,
wavy ossification of the anterior longitudinal ligament with relative preservation of disc spaces. The ossification is seen only in the antero-lateral aspect of the affected spine. In AS, there is squaring of the vertebrae as seen on lateral radiograph (Fig. 4). Ossification occurs in both anterior and posterior longitudinal ligaments as well as the annulus fibrosis creating marginal osteophytes. The most pathognomonic radiographic change would be sacroiliac joint obliteration. Our patient’s sacroiliac joints were normal. However, the
concomitant ossification of the posterior longitudinal ligament in a patient with DISH may lead physicians to diagnose ankylosing spondylitis. Clinical presentations of DISH and AS differ in terms of age presentation, although both have a predilection for males. AS patients tend to present at a younger age group, typically 15 - 30 years, complaining of insidious onset of pain and stiffness, culminating in gradual loss of spinal mobility. AS affects the hip joints in up to 50% of cases. Rib articulations are also not spared and this gives rise to limited chest expansion on inspiration. The hallmark of AS is symptomatic sacroiliitis with its attendant radiographic changes. These changes usually occur on the iliac side, with patchy periarticular osteopaenia. This is followed by loss of definition with superficial erosions and then subchondral sclerosis with an apparent widening of the joint. The final radiographic stage is eventual fusion of the joint and obliteration of the joint space. Acute spinal pain and neurological deficits usually only occur in a fractured AS spine. The HLA B27 test is useful only in aiding diagnosis of AS and is positive in 90% of cases. However, HLA B27 is present in 7% of the local population who do not have clinical features of AS (4). A proportion of these subjects may develop AS or other HLA B27 related conditions like Reiter’s syndrome or Behcet’s syndrome.

Treatment for AS is largely conservative. Surgical options include total hip replacements for severely affected hips, surgical decompression and fusion for spinal fractures with neurological deficits and corrective osteotomies for realignment of severely deformed spines. DISH, on the other hand, is largely asymptomatic and detected as an incidental radiological finding. It is an idiopathic condition which affects those over 60 years of age. There have been reported cases of ossification of spinal ligaments associated with long-term therapy with retinoic acid (5,6). Patients with DISH may present as stiffness. Large osteophytes may develop, resulting in compressive effects on structures anterior to the affected spine. These include dysphagia secondary to oesophageal compression (7), chronic obstructive pneumonia due to compression of a bronchus (8) and even inferior vena cava compression (9). While several articles have reported DISH presenting with compressive symptoms, the authors are aware of only one reported case of DISH causing cervical cord compression (10). There are three reported cases of DISH (11,12) co-existing with ossified posterior longitudinal ligament (OPLL) giving rise to neurological sequelae as a result of minor trauma to the neck. OPLL alone, giving rise to cervical myelopathy and cervical cord injuries is fairly common and has been extensively reported (13,14).

Table I summarises the comparative features of DISH, AS and OPLL.

<table>
<thead>
<tr>
<th></th>
<th>DISH</th>
<th>AS</th>
<th>OPLL</th>
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<tbody>
<tr>
<td><strong>Aetiology</strong></td>
<td>Unknown</td>
<td>HLA-B27 related inflammatory response</td>
<td>Largely idiopathic, long term therapy with retinoic acid</td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td>Predominantly male</td>
<td>90% male</td>
<td>Equal</td>
</tr>
<tr>
<td><strong>Age of onset</strong></td>
<td>Over 60</td>
<td>20 - 30</td>
<td>Over 60</td>
</tr>
<tr>
<td><strong>Clinical presentation</strong></td>
<td>Incidental finding; largely asymptomatic</td>
<td>Insidious onset of back ache and stiffness</td>
<td>Cervical myelopathic symptoms</td>
</tr>
<tr>
<td><strong>Compressive symptoms</strong></td>
<td>Dysphagia, if present</td>
<td>None</td>
<td>Cervical myelopathy</td>
</tr>
<tr>
<td><strong>Affected sites</strong></td>
<td>Thoracic &gt; cervical sacroiliac joints spared</td>
<td>Whole spine, especially Sacroiliac joints</td>
<td>Cervical</td>
</tr>
<tr>
<td><strong>Physical findings</strong></td>
<td>Stiffness in affected spine Trauma induced neurological deficit</td>
<td>Stiffness, decreased chest expansion, positive “pump handle” test, Neurological deficits following fracture</td>
<td>Rarely causes stiffness, neurological findings of myelopathy, most commonly central cord syndrome</td>
</tr>
<tr>
<td><strong>X-ray features</strong></td>
<td>Antero-lateral ossification, non marginal syndesmophytes</td>
<td>Squaring of vertebrae, ossification of both anterior and posterior longitudinal ligaments, symmetrical ligaments, symmetrical sacroiliitis</td>
<td>Ossification of only posterior longitudinal ligament</td>
</tr>
<tr>
<td><strong>HLA B27 marker</strong></td>
<td>Absent</td>
<td>Positive in 90%</td>
<td>Absent</td>
</tr>
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</table>

**CONCLUSION**

The distinction between AS and DISH may only be an academic exercise in our patient as the management would have been along similar lines, which is conservative and expectant. This is because 75% of patients with central cord syndrome recover useful neurological function. In order to make the distinction between AS and DISH with co-existing OPLL, X-rays
of the sacroiliac joints may provide the only clue, coupled with the clinical picture of presentation. Limitations in chest expansion on inspiration and HLA B27 markers are helpful indicators. Fracture in an AS afflicted cervical spine will require bony stabilisation in addition to surgical decompression (although in our patient, conservative management was chosen).

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REFERENCES