Diffuse idiopathic skeletal hyperostosis (DISH) – A case report

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Abstract
Diffuse idiopathic skeletal hyperostosis is a non inflammatory systemic disease affecting axial skeleton which results in ossification and calcification of spinal ligaments and entheses. The pathophysiology of the disease is poorly understood. The closest differential diagnosis for DISH remains ankylosing spondylitis. Further research is necessary to treat the disease and to improve the prognosis and quality of life of patients affected with DISH. This article reports a patient with DISH and its management in a tertiary care hospital.

Keywords: Forestier’s disease, Diffuse idiopathic skeletal hyperostosis, Calcification, Ossification, Anterior longitudinal ligament, Candle wax, Ankylosing spondylitis, Syndesmophytes.

Introduction
Diffuse idiopathic skeletal hyperostosis is a benign, slowly progressive condition affecting axial skeleton which results in ossification and calcification of spinal ligaments and entheses.¹ The pathophysiology of the DISH still remains unclear. The multifactorial web of causation led to further investigation of the factor linkage to the outcome of the disease and its prognosis. Here we have reported a case of DISH with its clinical features, diagnosis, management and future prospects of the disease.

Case Report
Here we report a case of 58 years male patient came to JJM Medical College with a chief complaints of lower back pain from past 3 months and radiating to whole of the left lower limb, weakness of left lower limb and tingling & numbness over bilateral lower limbs from past 1 month. The patient has normal bowel and bladder habits. The patient is a non diabetic and a normotensive individual. Systemic examination is normal.

On examination, the patient walked with antalgic gait. Tenderness is present over L3 – L4, L4 – L5 and L5 – S1 spinous process. Paraspinal muscle spasm is present over lumbar aspect of both sides. There is no kyphoscoliosis and no exaggerated lumbar lordosis. Movements over LS spine shows 60° flexion, 20° extension, 30° right lateral bending and 20° left lateral bending movements noted which are associated with pain.

Neurological examination
a) Higher mental function – Normal
b) Cranial nerve examination – Normal
c) Motor system

d) Sensory system
   Touch, temperature, pressure, vibration, 2 point discrimination, stereognosis over lumbo-sacral region – Normal

Table 1

<table>
<thead>
<tr>
<th>Components</th>
<th>Right</th>
<th>Left</th>
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<tr>
<td>Bulk</td>
<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Tone</td>
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Table 2

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<thead>
<tr>
<th>Tests</th>
<th>Right</th>
<th>Left</th>
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<tbody>
<tr>
<td>EHL and FHL</td>
<td>5/5</td>
<td>3/5</td>
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<tr>
<td>SLRT Active</td>
<td>90</td>
<td>5</td>
</tr>
<tr>
<td>Passive</td>
<td>90</td>
<td>75</td>
</tr>
<tr>
<td>FABER test</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>Lassegue’s test</td>
<td>Positive</td>
<td>Positive</td>
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Investigations
a) Hemogram
   i. Hb – 14.3 gm/dL
   ii. Total count – 6000 cells/mm³
   iii. RBC – 4.3 million/mm³
   iv. Platelets – 2.3 lakh cells/mm³
   v. ESR – 7 mm/hour
   vi. CRP – 6 mg/L

b) Peripheral smear – Hypochromic microcytic anemia

c) Renal function tests – Urea 31 mg/dL and creatinine 1.6 mg/dL

d) Random blood glucose – 78 mg/dL

e) HIV and HbsAg – Non reactive

f) HLA B27 – Negative
g) RA factor – 12 IU/mL
h) Anti CCP antibodies – 9 u/mL

i) Radiography (Fig. 1, 2 & 3)

Table 3

<table>
<thead>
<tr>
<th>X ray of the part</th>
<th>Radiological findings</th>
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<tbody>
<tr>
<td>Right knee – AP and lateral</td>
<td>Multiple osseous projections noted in patella and proximal tibia and calcified patellar tendon noted</td>
</tr>
<tr>
<td>Left knee – AP and lateral</td>
<td>Bony outgrowth noted in infra patellar region with periosteal reaction noted in fibula</td>
</tr>
<tr>
<td>Right hand – AP and oblique</td>
<td>Periosteal reaction noted in the lateral aspect of 1st metacarpal</td>
</tr>
<tr>
<td>Cervical spine – Lateral</td>
<td>Marginal osteophytes noted in C5 and C6 levels</td>
</tr>
<tr>
<td>Lumbosacral spine – AP and Lateral</td>
<td>Syndesmophytes noted in L1 – L2, L2 – L3, L3 – L4 vertebral levels</td>
</tr>
<tr>
<td>Pelvis with both hips – AP</td>
<td>Calcification of sacrotuberosous ligament and mild whiskering in the ischial tuberosities</td>
</tr>
</tbody>
</table>

and proximal tibia and calcification of right patellar tendon

j) MRI spine

i. Anterior and lateral bridging osteophytes noted at multiple levels in cervico-thoraco-lumbar vertebrae

ii. Flowing calcification and hypertrophy noted over anterior longitudinal ligament

iii. Ligamentum flavum and facet hypertrophy noted at multiple levels in cervico-thoraco-lumbar vertebrae

iv. Disc bulge noted at C5 – C6, C6 – C7, L4 – L5 and L5 – S1 vertebral level with ligamentum flavum and facet joint arthropathy indenting the thecal sac

v. Cord edema noted at T10 – T11 and T11 – T12 vertebral level

Fig. 1: X ray of LS spine lateral view showing syndesmophytes in L1 – L2, L2 – L3, L3 – L4 vertebral levels with preservation of disc height

Fig. 2: X ray pelvis with both hips AP view showing calcification of sacrotuberosous ligament and mild whiskering in the ischial tuberosities

Fig. 3: X ray right knee in AP and lateral view showing multiple osseous projections noted in patella

Fig. 4: MRI of T2 sagittal section of cervico thoracic spine showing anterior bridging osteophytes in multiple adjacent cervico thoracic vertebra and calcification of anterior longitudinal ligament
Discussion

In 1950, Jacques Forestier and Jaume Rotes-Querol coined Forestier’s disease and termed it under “senile ankylosing vertebral hyperostosis”. In 1976, Resnick and Niwayama coined the term “diffuse idiopathic skeletal hyperostosis” (DISH), which is a diffuse, systemic and non-inflammatory disease characterized by ossification of entheses and calcification of ligaments. Diffuse idiopathic skeletal hyperostosis is also called as Forestier’s disease, senile ankylosing spondylosis or ankylosing hyperostosis. DISH has a slight predilection over males and incidence increases with age of 40 years. DISH occurs more commonly in thoracic vertebrae followed by cervical and lumbar vertebrae. The etio-pathogenesis of the disease remains controversial. Multifactorial web of causation of DISH is proposed in the form of mechanical factors, genetic factors, environmental exposures, drugs and metabolic conditions. The main pathophysiology underlying the DISH is the abnormal growth and function of the osteoblasts in osteoligamentary binding and the pathological calcification along the anterior longitudinal ligament.

Diagnostic criteria for DISH
1. Flowing calcification along anterolateral aspect of atleast 4 successive vertebrae
2. Preservation of disc height in the involved vertebral segment
3. Relative absence of significant degenerative changes
4. Absence of facet joint ankylosis and sacroiliac joint involvement

The patient with DISH presents with chronic low back pain, stiffness of lower back which is worse in the morning, pain and stiffness over neck, dysphagia, stridor and hoarseness of voice. On physical examination, there will be restriction of movements over spine. Skeletal survey and MRI of spine stood the diagnostic of choice for diagnosing DISH.

Here our patient presented with lower back pain from past 3 months and radiating to whole of the left lower limb, weakness of left lower limb and tingling & numbness over bilateral lower limbs from past 1 month. He is a non-diabetic and normotensive. Laboratory investigations showed a non-inflammatory results. The radiological analysis showed the presence of syndesmophytes in consecutive 4 lumbar vertebrae. MRI spine revealed anterior and lateral bridging osteophytes noted at multiple levels in cervico-thoraco-lumbar vertebrae and flowing calcification and hypertrophy noted over anterior longitudinal ligament of spine.

The closest differential diagnosis for DISH is ankylosing spondylitis. There are subtle differences between both the clinical entities which are discussed below.

Table 4

<table>
<thead>
<tr>
<th>Features</th>
<th>DISH</th>
<th>Ankylosing spondylitis</th>
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<tbody>
<tr>
<td>Affection</td>
<td>Thoracic vertebra</td>
<td>Lumbosacral vertebra</td>
</tr>
<tr>
<td>Laterality</td>
<td>Unilateral</td>
<td>Bilateral</td>
</tr>
<tr>
<td>Age group</td>
<td>Elderly</td>
<td>Younger</td>
</tr>
<tr>
<td>Genetics</td>
<td>No association with HLA-B27</td>
<td>Strong association with HLA-B27</td>
</tr>
<tr>
<td>Radiograph</td>
<td>Candle wax dripping appearance</td>
<td>Bamboo spine appearance</td>
</tr>
<tr>
<td>Disc space</td>
<td>Affected disc space preserved</td>
<td>Affected disc space reduced</td>
</tr>
<tr>
<td>Degenerative changes</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Sacroiliac joint involvement</td>
<td>Absent</td>
<td>Present</td>
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</table>
The management of disease remains symptomatic and empirical. There is no proposed definitive management protocol for DISH. Non operative management includes analgesics, antioxidants and physical therapy in the form of short wave diathermy, interferential therapy and intermittent pelvic traction. Surgical management is advised for the patients present with unstable vertebral fractures.

Recent studies and research on DISH states that DISH is associated with obesity, diabetes mellitus, hypertension, syndrome X and hyperuricemia. These patients have a higher risk for development of stroke and coronary vascular diseases.5 Hence our patient has been explained about the future prospect of the condition DISH and advised for screening of diabetes, hypertension and syndrome X in a regular basis to prevent stroke and coronary vascular complications in a long term. Reassurance has been given for the patient about the course of the disease and to improve the quality of life.

**Conclusion**

A complete evaluation of low back ache is needed to arrive at the diagnosis of diffuse idiopathic skeletal hyperostosis which should be further differentiated from ankylosing spondylitis. Early diagnosis and treatment should be strictly instituted which can prevent future complications such as metabolic syndrome, stroke and cardiovascular complications. Future research on metabolic targets of DISH has to be performed which will provide the exact pathophysiology for the development of disease.7

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**References**