

## 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.<sup>1</sup> The pathophysiology of the DISH still remains unclear. The multifactorial web of causation of DISH has been proposed. This 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

- Higher mental function – Normal
- Cranial nerve examination – Normal
- Motor system

**Table 1**

Components	Right	Left
Bulk	Normal	Normal
Tone	Normal	Normal

Power	Hip	5/5	4/5
	Knee	5/5	4/5
	Ankle	5/5	4/5
Reflexes	Abdominal	Normal	Normal
	Knee jerk	Normal	Decreased
	Ankle jerk	Normal	Decreased
	Plantar	Flexor	Flexor

- Sensory system  
Touch, temperature, pressure, vibration, 2 point discrimination, stereognosis over lumbo-sacral region – Normal
- Autonomic system  
Bowel and bladder – Normal and regular
- Cerebellar examination – normal
- Special tests

**Table 2**

Tests	Right	Left	
EHL and FHL	5/5	3/5	
SLRT	Active	90°	5°
	Passive	90°	75°
FABER test	Positive	Positive	
Lassegue's test	Positive	Positive	

#### Investigations

- Hemogram
  - Hb – 14.3 gm/dL
  - Total count – 6000 cells/mm<sup>3</sup>
  - RBC – 4.3 million/mm<sup>3</sup>
  - Platelets – 2.3 lakh cells/mm<sup>3</sup>
  - ESR – 7 mm/hour
  - CRP – 6 mg/L
- Peripheral smear – Hypochromic microcytic anemia
- Renal function tests – Urea 31 mg/dL and creatinine 1.6 mg/dL
- Random blood glucose – 78 mg/dL
- HIV and HbsAg – Non reactive
- HLA B27 – Negative
- RA factor – 12 IU/mL

h) Anti CCP antibodies – 9 u/mL

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

**Table 3**

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



**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 sacrotuberous 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

**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. 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



**Fig. 5: MRI T2 sagittal and T2 axial images of spine showing ligamentum hypertrophy causing spinal canal stenosis and compression of traversing nerve roots**

This patient was treated conservatively with bilateral above knee skin traction. The patient was treated with indomethacin tablets 75 mg twice daily for 2 weeks, pregabalin – nortriptyline combination tablet once daily at night for 2 months and supplemented with anti-oxidant medications. The patient was treated with physical therapy in the form of hot fomentation, intermittent pelvic traction, short wave diathermy and interferential therapy twice daily for 2 weeks. The patient has been explained about the natural course, outcome and prognosis of the disease. Reassurance was given and the patient is discharged with the advice to perform exercises daily which will improve the quality of life.

### Discussion

In 1950, Jacques Forestier and Jaume Rotes-Querol coined Forestier's disease and termed it under "senile ankylosing vertebral hyperostosis".<sup>2</sup> 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.<sup>3</sup>

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.<sup>4</sup> 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.<sup>6</sup>

**Table 4**

Features	DISH	Ankylosing spondylitis
Affection	Thoracic vertebra	Lumbosacral vertebra
Laterality	Unilateral	Bilateral
Age group	Elderly	Younger
Genetics	No association with HLA-B27	Strong association with HLA-B27
Radiograph	Candle wax dripping appearance	Bamboo spine appearance
Disc space	Affected disc space preserved	Affected disc space reduced
Degenerative changes	Absent	Present
Sacroiliac joint involvement	Absent	Present

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.<sup>5</sup> 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.<sup>7</sup>

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