

Diffuse idiopathic skeletal hyperostosis presenting as cervical and thoracic myelopathy

Nikhil Gladson^{1,*}, Shaji C. V², Vishal V. Panicker³, Meena Kumari⁴

^{1,3,4}Senior Resident, ²Professor, Dept. of Neurology, Govt. T D Medical College, Alappuzha, Kerala, India

***Corresponding Author:**

Email: nikhilgladson@gmail.com

Abstract

Introduction: Diffuse idiopathic skeletal hyperostosis (DISH), also termed Forestier's disease, occurs in about 12% of middle-age and elderly men.¹ Anterolateral ossification of the anterior longitudinal ligament leads to ankylosing hyperostosis of continuous vertebral bodies. Neurological problems due to Ossification of Posterior Longitudinal Ligament [OPLL] or Ossification of Ligamentum Flavum [OLF] associated with DISH have been reported.

Case Report: A 42 year old male presented with insidious onset neck pain restriction of neck and back movements of 1 year duration. For last 2 weeks presented with weakness and sensory symptoms of both lower. On examination was found to have features of cervical myeloradiculopathy. He was found to have bridging ossification of cervical and thoracic vertebrae in X Ray. MRI showed ossification of posterior longitudinal ligament in cervical spine and ossification of ligamentum flavum in thoracic spine causing compressive myelopathy. He was diagnosed to have DISH with OPLL and OLF causing cervical and thoracic compressive myelopathy.

Keywords: Diffuse idiopathic skeletal hyperostosis, Ossification of posterior longitudinal ligament, Cervical myeloradiculopathy, Ossification of ligamentum flavum.

Introduction

Diffuse idiopathic skeletal hyperostosis (DISH), also termed Forestier's disease, occurs in about 12% of middle-age and elderly men.¹ Anterolateral ossification of the anterior longitudinal ligament leads to ankylosing hyperostosis of certain continuous vertebral bodies.² Ossification and calcification of the anterolateral aspect of the thoracic spine is considered to be the hallmark of the disease.³

The most commonly used classification criteria were defined by Resnick and Niwayama, and require involvement of at least four contiguous thoracic vertebral segments, preservation of intervertebral disc spaces and the absence of apophyseal joint degeneration or sacroiliac inflammatory changes.²

Reports of cervical myelopathy in DISH are rare. neurological problems due to ossification of posterior longitudinal ligament or ossification of ligamentum flavum associated with DISH also have been reported. In DISH, most reported cases of myelopathy due to OPLL occurred in the subaxial cervical spine.⁴

Case Report

A 42 year old man drama artist by profession presented to us with a history of neck pain for last 1 year with pain increases on movement of neck sideways and restriction of neck movements to either sides and difficulty to extend the neck for last 6 months. For last 6 months he developed low back ache which increases on bending down or doing some work. There was no history of radiation of pain to limbs. He developed lower abdominal pain and girdle sensation below umbilicus for last 2 weeks. For last 2 weeks he developed insidious onset dragging of both lower limbs

Rt> Lt with heaviness of limbs and log of wood sensation. Along with this he also developed slipping of chappals with knowledge. He also had 3 episodes of tripping and fall in the last 2 weeks. Since last 1 week he has difficulty assuming squatting position and can get up from squatting position with the support of 2 persons. He walks with the support of 1 person for the last 1 week. He has urgency of micturition and urge incontinence with hesitancy for last 1 month. He also noticed burning sensation of lateral thigh, lateral aspect of right leg and lateral aspect of left thigh. No history of weakness or sensory symptoms of upperlimbs. No history of perianal sensory loss, symptoms suggestive of cranial nerve involvement or cognitive dysfunction.

He had a history of fall from bike when the bike which he was riding lost control and he fell with face hitting the ground one and a half year back.

On examination he was well built and nourished with a blood pressure of 130/90 mm of Hg. Cranial nerve examination showed a normal optic fundus with full extraocular movement and no nystagmus. Motor system examination showed spasticity of both upper and lower limbs. He had normal power in upper limb and Grade 3 power in lower limbs Right >Left and beevors sign was positive. Deep tendon reflexes were normal in upper limbs and exaggerated in lower limbs with bilateral extensor plantar response.

Abdominal reflex was absent in all 4 quadrants and vibration was impaired upto T10 with intact pain and temperature sensations. He could walk with the support of one person with dragging of both lower limbs.

His lateral and flexion extension neck movements and spine movements were restricted. Modified schobers test was positive showed 2 cm increase only.

His blood investigations showed Hb-14.1 gm% Total WBC count-8700 Polymorphs 58% Lymphocytes30% and monocytes11% Platelet count-2.76 lakhs ESR-8 mm/1st Hr. His Random blood sugar was 114 gm%, Blood urea 19 Ser Creatinine-1.0,

Ser Sodium-140, Ser Pottasium-4.6, Total Protein-7.6, Ser Albumin-3.8 SGOT-26, SGPT-27, Alkaline Phosphatase-60

His X Ray Chest PA view was normal ECG was Normal

His X ray Cervical spine [Fig. 1] showed contiguous bridging ossification of C2 to C6 cervical vertebrae. His X Ray of Thoracic spine [Fig. 2] also showed bridging ossification in anterolateral aspects of right T10 to T12 vertebrae.

So a possibility of diffuse idiopathic skeletal hyperostosis was considered.

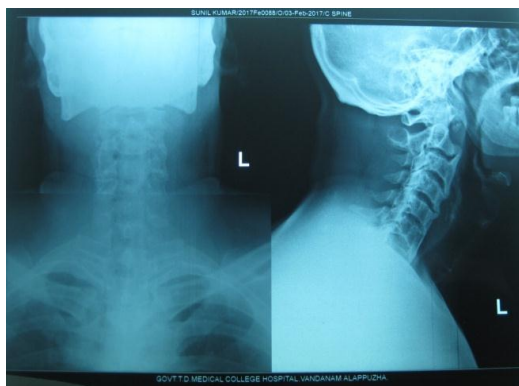


Fig 1: X ray cervical spine showing contiguous bridging ossification of C2 to C6 vertebrae



Fig 2: X ray thoracic spine showing bridging syndesmophytes in T10 to T12 vertebrae right side

His MRI cervical Spine [Fig. 3, 4] showed ossification of posterior longitudinal ligament from C2 to T2 vertebrae causing compressive cervical myelopathy with contiguous bridging ossification from C2 to C6. His MRI thoracic spine [Fig. 5, 6] showed nodular calcification and ossification of ligamentum

flavum causing compression of thoracic spinal cord at T7 and T10.

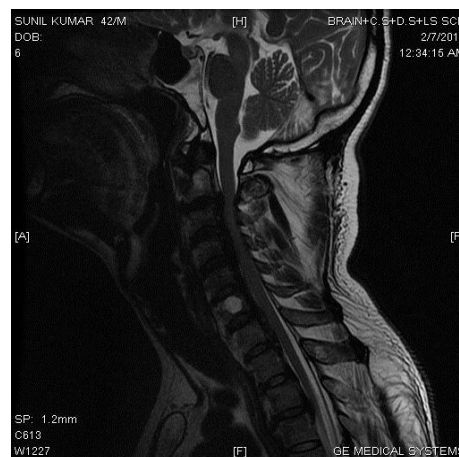


Fig. 3: MRI cervical spine T2 sagittal showing cervical compressive myelopathy with ossification of posterior longitudinal ligament from C2 to T2 with hyperintense signal in cervical spinal cord



Fig. 4: MRI T2 axial sequence showing spinal cord compression secondary to OPLL



Fig. 5: MRI T2 Sagittal Sequence of thoracic spine showing ossification of ligamentum flavum at T7 and T10 causing compression of thoracic spinal cord



Fig. 6: MRI T2 Axial film of thoracic spine showing compression by ossified ligamentum flavum

Further evaluation was done for differentiating from ankylosing spondylitis. His CRP was negative. X Ray Pelvis with SI Joint was normal. MRI of both sacroiliac joint [Fig. 7] was taken which showed no evidence of sacroillitis. HLA B27 was also done which was negative.



Fig. 7: MRI Sacroiliac joint coronal image showing normal signal intensity from both sacroiliac joints

So a final diagnosis of diffuse idiopathic skeletal hyperostosis with compressive cervical and thoracic myelopathy secondary to ossification posterior longitudinal ligament and ligamentum flavum was made.

Discussion

DISH is most commonly observed in individuals over the age of 50 with a reported prevalence between 2.5 and 28%.⁵ The diagnosis of DISH is currently based on classification criteria that require the involvement of

the spinal thoracic segment. To differentiate DISH from AS and degenerative disease, Resnick et al.² proposed the following criteria: 1) “flowing” ossification extending over four contiguous vertebrae; 2) relative preservation of intervertebral disc height in relation to age; and 3) absence of apophyseal joint ankylosis or sacroiliac changes.

Our patient fulfilled the Resnick criteria of DISH.

Neurological symptoms which are rarely seen include decreased flexibility of the spine and narrowed spinal canal-related myelopathy due to anterior and posterior longitudinal ligament calcification. Myelopathy is often associated with narrowing of the lower cervical vertebral canal.⁶

Neurological problems due to OPLL or OLF associated with DISH also have been reported. In DISH, most reported cases of myelopathy due to OPLL occurred in the subaxial cervical spine.⁴

In the previous studies, OLF associated with DISH played an important role in the development of neurological deficits in the thoracic or lumbar spine.⁷

Our patient had cervical OPLL and thoracic OLF causing compressive myelopathy at both cervical and thoracic level.

The incidence of OPLL is 2.4% in the Asian population and 0.16% in non-asian populations.⁸ DISH with OPLL is not common, 8/49 patients with OPLL (16%) had DISH with cervical myeloradiculopathy.⁹

Conflicting Interest: Nil

References

1. Smythe H, Littlejohn G. Diffuse idiopathic skeletal hyperostosis. In: Kippel JH, Dieppe PA (eds) *Rheumatology*. Mosby, London, 1984; pp 1–6.
2. Resnick D, Niwayama G. Diffuse idiopathic skeletal hyperostosis (DISH): ankylosing hyperostosis of forestier and rotesquerol. In: Resnick D, Niwayama G (eds) *Diagnosis of bone and joint disorders with emphasis on articular abnormalities*. WB Saunders, Philadelphia, 1988; pp 1562–1602.
3. Resnick D, Niwayama G. Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* 1976;119:559–68.
4. Griffiths ID, Fitzjohn TP. Cervical myelopathy, ossification of the posterior longitudinal ligament, and diffuse idiopathic skeletal hyperostosis: problems in investigation. *Ann Rheum Dis* 1987;46:166–168.
5. Boachie-Adjei O, Bullough P G. Incidence of ankylosing hyperostosis of the spine (Forestier's disease) at autopsy. *Spine (Phila Pa 1976)* 1987;128: 739–743.
6. Epstein NE. Simultaneous cervical diffuse idiopathic skeletal hyperostosis and ossification of the posterior longitudinal ligament resulting in dysphagia or myelopathy in two geriatric North Americans. *Surg Neurol* 2000;53:427–431.
7. Reisner A, Stiles RG, Tindall SC. Diffuse idiopathic skeletal hyperostosis causing acute thoracic myelopathy: a case report and discussion. *Neurosurgery* 1990;26:507–511.

8. Tsuyama N. Ossification of the posterior longitudinal ligament of the spine *Clin Orthop Relat Res* 1984;184:71–84.
9. Ryoji Tauchi, Sang-Hun Lee, Colleen Peters, Shiro Imagama, Naoki Ishiguro, K. Daniel Riew. Cervical Myeloradiculopathy due to Ossification of the Posterior Longitudinal Ligament with versus without Diffuse Idiopathic Spinal Hyperostosis. *Global Spine Journal* 2016;6,4:350-356.