



Diffuse Idiopathic Skeletal Hyperostosis (DISH) – a Rare Presentation of the Disease

KEYWORDS

Diffuse idiopathic skeletal hyperostosis, Enthesopathy, Flowing spinal calcification, Forestier's disease.

Dr. Dodia Apoorva V

4th year Resident in Orthopaedics, M.P.Shah Medical College, Jamnagar. "Apoorva"; 65, Digvijay Plot; Near Jolly Bunglow; Summair Club Road; Jamnagar, Gujarat.

Dr. Pratik D. Shah

3rd year Resident in Orthopaedics, M.P.Shah Medical College, Jamnagar. Room No.38, New PG Hostel, M.P.Shah Govt. Medical College, Jamnagar, Gujarat.

Dr. Soman Shardul M.

3rd year Resident in Orthopaedics, M.P.Shah Medical College, Jamnagar. Room No.28, New PG Hostel, M.P.Shah Govt. Medical College, Jamnagar, Gujarat.

ABSTRACT Diffuse idiopathic skeletal hyperostosis (DISH) also known as Forestier's disease, is a disorder of unknown etiology that is characterized by back pain and spinal stiffness. The condition is recognized radiographically by the presence of "flowing" ossification along the anterolateral margins of at least four contiguous vertebrae (candle wax dripping appearance) and the absence of changes of spondyloarthropathy or degenerative spondylosis. Even in patients who present with either lumbar or cervical complaints, radiographic findings are almost universally seen on the right side of the thoracic spine. It is characterized by a tendency toward ossification of ligaments (enthesopathy). It most characteristically affects the spine. Ossification of the longitudinal ligaments (especially the anterior ligaments) of the spine produces a tortuous paravertebral mass anterior to and distinct from the vertebral bodies. The zygapophyseal and sacroiliac joints are not involved in DISH, and the intervening intervertebral disk space is preserved. But our patient presented with hip and knee pain rather than the usual spine complaints.

• Introduction:-

Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a skeletal disorder that produces characteristic degenerations in both spinal and extra-spinal structures. It is an idiopathic form of degenerative arthritis. It is usually characterized by exuberant bony growth along the anterior longitudinal ligament and typically affects males over 60 years of age.

Diffuse idiopathic skeletal hyperostosis, previously known as Forestier's disease, is the most common enthesopathy. In 1950 it was originally described by Forestier and Rotes-Querol as a senile ankylosing hyperostosis of the spine. This condition is associated with extraaxial involvement (i.e., ossification of the nuchal ligament or tendons in the extremities). Patients with this diagnosis can suffer from heel spurs or ligamentous calcification at the elbow or calcification of the intra-pelvic ligaments.

Exact aetiology of DISH syndrome is unclear. However it is more common in type 2 diabetes and prolonged hyperinsulinaemia may be an explanation. Among other possible factors, elevated levels of insulin growth factor or elevated levels of growth hormone may play a part in the new bone formation. One half of all patients with DISH syndrome will have manifest diabetes or impaired glucose tolerance. DISH syndrome may be associated with obesity, hyperlipidaemia and hyperuricaemia, coronary heart disease in addition to its association with diabetes. It does not show an association with the duration of diabetes.

The disease is most common in the thoracic spine and occurs less frequently in the lumbar and cervical spine. The disease most commonly presents in the sixth and seventh decades of life and its estimated frequency in the elderly is 5-15%. Signs and symptoms include stiffness and pain in the back, dysphagia due to direct esophageal compression/distorsion, pain related to associated tendinitis, myelopathy related to core compression associated to the ossification of the posterior longitudinal ligament, and pain related to vertebral complications--e.g. fracture/subluxation. While conventional radiography clearly confirms the diagnosis of diffuse idiopathic

skeletal hyperostosis, CT and MRI better detect associated findings (e.g. ossification of the posterior longitudinal ligament) and complications (e.g. spinal cord compressive myelomalacia).

Some researchers feel that the extra bone is made because of extra blood supply near the spine. Growth factors that affect the formation of calcium are likely to play a role. DISH generally affects the ligament in front of the spine called the anterior longitudinal ligament. This is the most common ligament that turns into bone with DISH, but it can affect other ligaments as well. This condition can also affect the ligaments and tendons in the heels, ankle, hips, knees, shoulder, elbows, and hands. Another common feature of DISH is that it can cause syndesmophytes (a bony outgrowth attached to a ligament) and tendonitis at sites other than the spine. These calcification sites are most often in the heels, elbows, ankles, knees, and shoulders. Bone spurs and inflammation develop where the tendon attaches to the bones. Although DISH is considered a form of osteoarthritis, it is not considered degenerative, from wear or tear. The disc heights and facet joints do not show wear and tear as with other forms of osteoarthritis. Medications that are used to treat acne may increase the risk of having DISH. These medications are called retinoids. They are similar to vitamin A.

• Clinical features:-

Stiffness and decreased range of motion are the most common symptoms of DISH. The stiffness usually happens in the morning upon waking. It also happens after a long period of rest. Often the decrease in motion in the spine is with side-bending. DISH most commonly affects the mid back, but can also affect the neck and low back. It also seems to affect the right side of the thoracic spine more often than the left side. Pain may be a symptom of DISH, but not always. In addition to changes in the spine, DISH can also cause stiffness, pain, and inflammation in tendons throughout the body. When DISH affects areas other than the spine, it feels like tendonitis. Ligaments and tendons where they attach to the bone near the joints develop extra bone growth (spurs). Most common joints outside of the spine that can have bone spurs

are the joints of the finger, elbow, shoulder, hip, knee, ankle and foot. Difficulty swallowing (dysphagia), or a hoarse voice, can happen when people have DISH. The pressure can also cause a hoarse voice or difficulty in breathing. In rare cases, this can become serious. This may require surgery to remove the bone spurs. Neurological problems are rare in DISH. In severe cases, the extra bone growth around the spine can cause problems with the spinal cord or nerves. Squeezing of the spinal cord can cause loss of feeling and paralysis.

• **Diagnostic criteria:-**

- 1) The presence of flowing calcification and ossification along the anterolateral aspect of at least four contiguous vertebral bodies.
- 2) Relative preservation of intervertebral disc height in the involved vertebral segment and the absence of extensive radiographic changes of degenerative disc disease.
- 3) The absence of apophyseal joint bony ankylosis and sacroiliac joint erosion, sclerosis or intra-articular osseous fusion.

• **Radiological abnormalities:-**

Spinal	<ol style="list-style-type: none"> 1) Anterolateral flowing ossification 2) Bumpy spinal contour 3) Radiolucent disc extension 4) Radiolucent area beneath deposited bone
Extra-spinal	<ol style="list-style-type: none"> 1) Bony proliferation 2) Ligament calcification/ossification 3) Para-articular osteophytes

• **Treatment modalities:-**

Nonsurgical Treatment: While there is no cure for DISH, there are treatments that can help the symptoms. Nonsteroidal anti-inflammatory drugs (NSAIDs) may help manage pain or tendonitis-like inflammation. More severe pain may be treated with corticosteroid injections.

Surgery: Rarely is surgery necessary. However, if the extra bone growth compresses the spinal cord or nerve roots, surgery may be needed. Surgery is done to relieve pressure on the spinal cord or nerve roots. Surgery to take out the extra bone growth (spurs) in the neck may help with symptoms of difficulty swallowing.

• **Case report:-**

Our patient was a 60 year old hindu female patient, a housewife, coming from low socioeconomic class. She had a complaint of pain in both hips and knee since past 1 year, which was insidious in onset, gradually progressive in nature, dull aching, localised, non-radiating, of mild intensity to start with, aggravated by sitting cross legged and squatting and prolonged walking, and partially relieved by rest and oral medications. She consulted multiple physicians who treated her condition as myalgia and gave her oral analgesics.

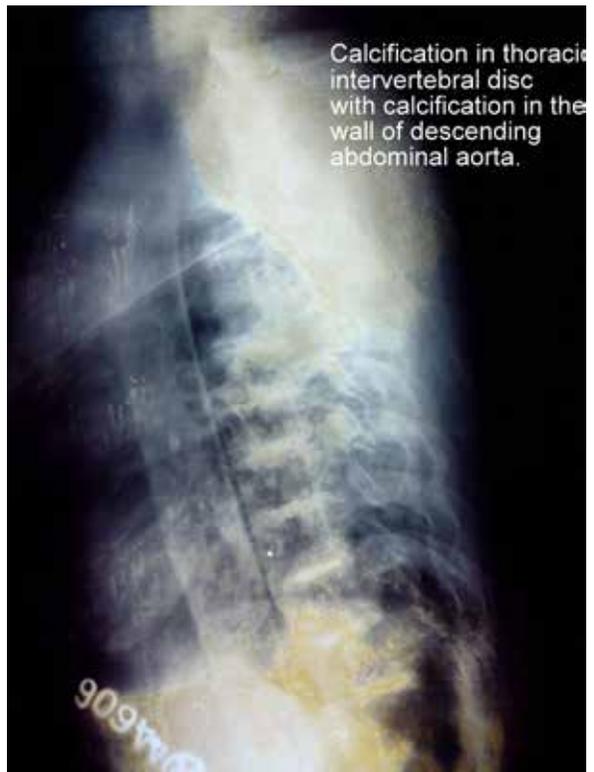
She was then referred to an orthopaedic surgeon who got x rays of the hip and knee of the patient and commented that the x rays were normal and he also treated the patient symptomatically using oral meds. But pain was progressing in intensity and was then associated with difficulty and at a later date inability to squat and sit cross legged. She also developed difficulty in walking.

She was then referred to us. We started off by assessing her clinically, which gave us the clue that something was wrong with her spine as well as both her hips. She had tenderness in the base of both scarpa's triangle, left side more in intensity than the right. Galeazzi test revealed left sided femoral shortening. There was a 30 degree fixed flexion deformity of the left hip and 10 degrees of fixed flexion deformity of the right hip. Left hip also showed a 20 degree of fixed adduction deformity and 10 degree of fixed internal rotation deformity. There was a 3 centimetre of apparent shortening of the left lower limb as compared to the right one but only 0.5 cen-

timetre of true shortening. There was associated exaggerated lumbar lordosis and restricted spinal movements. On detailed history work-up we found that she also had chronic backache since many years and associated spinal stiffness.

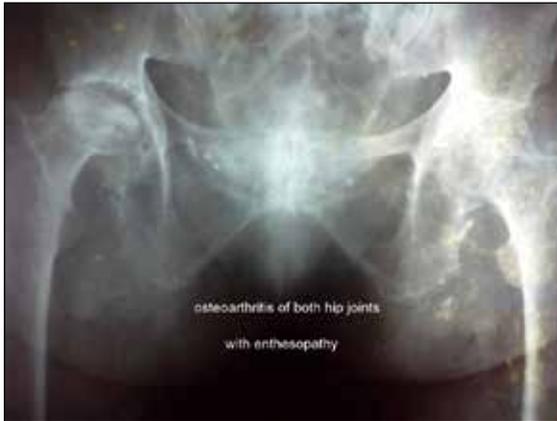


We ordered x rays of pelvis with both hips and lumbo-sacral spine and to our surprise we found a full blown enthesopathy in the pelvis with cortical irregularities in both femoral heads, left one involved more severely than the right one. The spine x-rays showed typical changes of DISH in the form of anterolateral flowing ossification, bumpy spinal contour, radiolucent disc extension and radiolucent area beneath deposited bone. She also showed calcification on the right side of the vertebral bodies of dorsal vertebrae. Then radiographic evaluation of the whole body was done and typical extra-spinal x-ray changes of DISH were observed. The x-rays of the patient met all the 3 criteria for diagnosing DISH. Ultrasonography of the abdomen and chest was done and incidental findings of multiple liver cysts and gall bladder stones were noted. MRI of the spine and both hips was done which proved the diagnosis of DISH and also showed arthritis of both hips. Her tox-screen was negative and lab reports showed that she was non-diabetic. She had no associated comorbidities.



Patient was advised for total hip replacement for both hips but patient was not willing for it. So she was treated con-

servatively for DISH in the form of analgesics.



• **Discussion and conclusion:-**

The usual presentation of a patient with DISH is spine stiffness and chronic backpain. But our patient presented with signs and symptoms of bilateral osteoarthritis of hip and DISH remained asymptomatic. Hence it may be prudent to say that not all patients of DISH present in the typical manner, some may not have any complaints at all and some may even present with signs and symptoms of some other disorder that does not bear any correlation with DISH.

REFERENCE

1. Forestier J, Rotes-Querol J: Senile ankylosing hyperostosis of the spine. *Ann Rheum Dis* 9:321- 330, 1950 || 2. Gibson T, Schumacher HR: Ankylosing hyperostosis with cervical spinal cord compression. *Rheumatol Rehabil* 15:67-70, 1976 || 3. Mazieres B, Rovensky J: Non-inflammatory enthesopathies of the spine: A diagnostic approach. *Baillieres Best Pract Res Clin Rheumatol* 14:201-217, 2000 || 4. McCafferty RR, Harrison MJ, Tamas LB, et al: Ossification of the anterior longitudinal ligament and Forestier's disease: an analysis of seven cases. *J Neurosurg* 83:13-17, 1995 || 5. Resnick D, Niwayama G: Radiographic and pathologic features of spinal involvement in diffuse idiopathic skeletal hyperostosis (DISH). *Radiology* 119:559-568, 1976 || 6. Tsukahara S, Miyazawa N, Akagawa H, et al: COL6A1, the candidate gene for ossification of the posterior longitudinal ligament, is associated with diffuse idiopathic skeletal hyperostosis in Japanese. *Spine* 30:2321-2324, 2005 |