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# "DISH" or "AS"?

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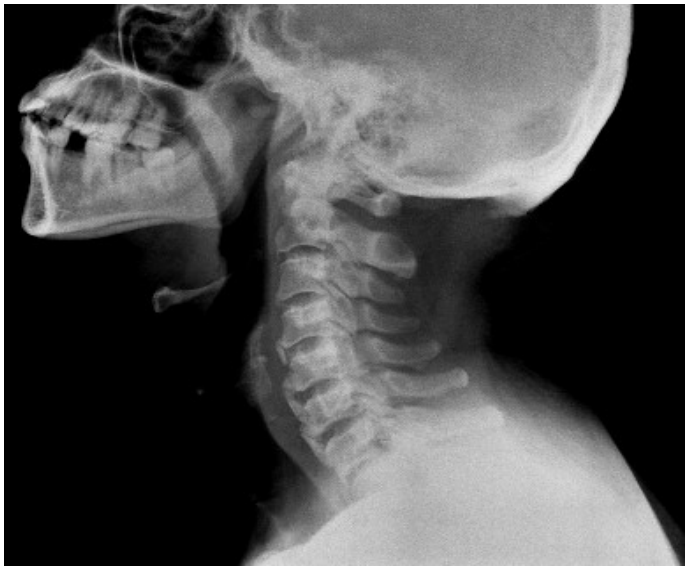
Exuberant ossifications and calcifications of bone and of entheses (the site of attachment of ligament or muscle to bone) is the characteristic feature of Proliferative Bone Disease, examples of which are Diffuse Idiopathic Skeletal Hyperostosis (DISH), Hypertrophic Osteoarthropathy (HOA), Osteoarthritis (OA), Seronegative spondyloarthropathies - Ankylosing Spondylitis (AS), Psoriatic arthritis (PsA), SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteomyelitis) and those associated with endocrine disorders - Acromegaly, Thyroid disorders and Hypoparathyroidism.

**Case 1:** A middle aged person had Hypertension and Hyperlipidemia of long standing. He presented with left sided hip pain of several months' duration. He was thin, had no clinical or biochemical features of Diabetes mellitus. The radiograph of his pelvis and spine showed osteophytes suggestive of Diffuse Idiopathic Skeletal Hyperostosis (DISH) (**Figure 1 a,b & c**).



**a**

**b**

**c**

**Figure 1:** X-rays of spine and pelvis showing osteophytes suggestive of Diffuse Idiopathic Skeletal Hyperostosis

**Case 2:** A middle aged woman with longstanding diabetes and back pain, had osteophytes arising from the right hip on her pelvic radiograph. She also had interspinous ligamental calcification along the entire thoracic spine.

**Case 3:** An elderly female had chronic back ache and severe restriction of neck movements of long duration. She was investigated extensively for metabolic disturbances and other arthritic conditions. Radiographs of cervical and thoracic spine and pelvis were taken.

Cervical spine AP and Lateral showed obliteration of cervical lordosis. There was extensive ossification of anterior longitudinal ligament and pre-vertebral soft tissues with retention of inter vertebral disk spaces and vertebral height. The anterior margins of the vertebral bodies merged imperceptibly with ossified prevertebral tissues. The posterior longitudinal ligament was heavily ossified with compromise of cervical spinal canal. There was evidence of calcification or ossification of interspinous or supraspinous ligaments. All the facet joints and unco-vertebral joints appeared normal. There was evidence for sclerosis along the right margin of the thoracic spine

In X-ray Pelvis AP view both Sacro iliac joints were normal. Whiskering was noticed in iliac crest, anterior superior, anterior inferior iliac spines and greater trochanters. Degenerative changes were present in both hip joints. There was evidence for pelvic ligaments calcification.

**Case 4:** An elderly male patient with a clinical diagnosis of advanced cervical spondylosis had stiffness of back and neck pain of long duration. A lateral cervical spine radiograph revealed exuberant osteophytosis in the cervical spine with preservation of inter vertebral disk spaces and vertebral height; facet joints were normal. He had no calcification of pre-vertebral soft tissues or spinal ligaments.

Excessive osteophytosis (‘Flowing osteophytes’) of the spine with preservation of disk spaces, vertebral heights, normal apophyseal joints and ossification of spinal ligaments and adjacent soft tissues in absence of clinical, radiological and serological evidences of other arthritis or arthritides favours the diagnosis of Diffuse Idiopathic Skeletal Hyperostosis (DISH) in all the cases described.

DISH is a common disorder which could baffle physicians who are unaware of this condition because it may be mistaken for one of the Spondyloarthropathies in symptomatology and radiological appearances.

Irregular bony outgrowths were first noted by Wenzel in 1824. It was Jacob Forestier who first described the entity in 7th International Congress on Rheumatology in New York in 1949, but it was eclipsed by Philips Hench's paper on the uses of Cortisone in Rheumatologic diseases and its importance recognized only after he published the radiologic findings of the condition the following year as ‘senile ankylosing disease of painless onset, distinct from Ankylosing Spondylitis’ [1].

Syndesmophytes are paravertebral ossifications with a vertical orientation, pathologically similar to osteophytes which have a horizontal orientation. Syndesmophytes are ‘marginal’ in AS and ‘non-marginal’ in DISH. Differentiating Syndesmophytes from Osteophytes may be difficult on orientation alone, but the former arise from the corner of the vertebral bodies in Ankylosing Spondylitis consequent upon inflammation [‘shiny corners’]. Bony growths originating inside a ligament, commonly in the ligaments of the spine, and specifically those in the intervertebral joints lead to fusion of vertebrae and ‘bamboo spine’ of AS.

DISH [Forestier's disease, ankylosing hyperostosis] is a benign non-inflammatory disorder of unknown aetiology, common in those aged more than 70 years (in 10% of men more than 65 years), and uncommon below the age of 45 years. Males are affected more than Females in a ratio of 2:1.

The prevalence of the disease is not known in India The overall prevalence of DISH was 17.0% in asymptomatic people above 50 years in Europe [2]. DISH was observed in 42% of men above 65 years in United state [3]. The osteophytes in the cervical region may cause dysphagia, hoarseness of voice, stridor, sleep apnoea and difficulties with intubation. Cervical myelopathy and stenosis may occur. Those with DISH are prone to hyper-extension injuries; the ankylosis of the vertebral segments proximal and distal to the fracture creates long lever arms which may result in instability and displacement even in low-energy injuries. It may be totally asymptomatic and picked up as an incidental finding on chance radiographs, with an incidence of 7/100 in men and 4/100 in women older than 30 year [4]. Although prevalence estimates vary in different population, the prevalence of DISH is consistently high and increases with age and obesity [6].

The exact aetiopathogenesis of DISH is not clear but genetic and environmental factors play a role. Most of the current theories focus on the pathologic calcification of the anterior longitudinal ligament of the spine due to the abnormal growth and function of the osteoblasts in the osteoligamentary binding [5]. Research into the pathophysiology of DISH has established that serum levels of the natural osteogenesis inhibitor Dickkopf-1 (DKK-1) are low in patients with DISH [6].

A relation has been described between DISH and obesity, Gout (upto 58% of men affected by gout may have DISH), Diabetes Mellitus, Hyperinsulinemia and raised Insulin Growth Factor -1. Lumbar canal stenosis may occur as an associated condition.

Mild chronic back pain or stiffness of the spine worse in the morning or on exposure to the cold may bring the patient to medical attention. The range of movement may be reduced or there could be features of cervical myelopathy or spinal stenosis. The entire spine may be affected, most commonly the thoracic spine on the right side, except in those with dextrocardia or situs inversus. The sparing of the left side of the spine is attributed to the pulsating aorta on the left side. This can be confirmed by isotope bone scan or other imaging techniques.

The cervical spine is affected more than the lumbar spine. Disc spaces and facet joints are not involved. Classically, advanced disease may have "melted candle wax"(‘flowing candle wax’) appearance along the spine on radiographic studies. Similar bone changes have been reported in human remains more than 4000 years old, also in dinosaurs, prehistoric reptiles, whales, dolphins, dogs, horses and monkeys. Ossification at muscle insertions results in ‘whiskering’; similar changes are described in the patella and foot. Bony sclerosis is a feature, and fractures are less common than in AS. A diagnosis of DISH can be made confidently by conventional radiography, but associated conditions and complication like spinal canal stenosis, cord compression and compressive myelomalacia etc are better made out by CT and MRI scans.

The diagnostic criteria for the DISH were described by Donald Resnick, one of the greatest musculo- sketetal radiologists, in 1976 as flowing osteophytes of variable thickness (upto 2 cm) coursing over the anterolateral aspect of at least 4 contiguous vertebral bodies in the thoracic spine as pathognomonic (4 vertebral body is only an arbitrary number) with preservation of the intervertebral disc space and height, absence of inflammatory changes in any apophyseal joint or the sacro-iliac joint. Facet joints show no bony ankylosis, sacro-iliac erosison, sclerosis or bony fusion.

Ossification of anterior longitudinal ligament can lead to a tortuous paravertebral thickening anterior to vertebral bodies at times with lucency between newly laid bone and vertebral margin. Bony Hyperostosis of posterior surface of the vertebral bodies, posterior spinal osteophytosis and ossification of posterior longitudinal ligaments are seen in a considerable percentage of patients with DISH. Exuberrant bone formations seen at entheses is the major extra vertebral manifestation and it is typically seen as whiskering in iliac crest, ischial tuberosity patella, calcaneum and olecranon.

Calcification and ossification of pelvic ligaments like sacro spinous, sacrotuberous and ilio lumbar

ligaments may even suggest the possibility of skeletal fluorosis, but for the absence of increased bone density.

Spinal DISH has characteristic pathologic features [8,9] - focal and diffuse calcification and ossification of the anterior longitudinal ligament, paraspinal connective tissue, annulus fibrosus; degeneration in the peripheral annulus fibrosus fibres, anterolateral extensions of fibrous tissue, hypervascularity, chronic inflammatory cellular infiltration; periosteal new bone formation on the anterior surface of the vertebral bodies. The new bone formed in DISH is normal bone with Haversian systems.

The human sacro-iliac (SI) joint has an antero-inferior synovial portion and a postero-superior cartilaginous portion. DISH causes bridging of the upper and cartilaginous non-synovial part. Involvement of the synovial portion of the SI joint excludes DISH. AS causes synovial inflammation and involvement of the SI joint starts at the antero-inferior portion with erosions first, ankylosis (and sclerosis) later. In AS usually there is bilateral and symmetric involvement of the SI joints, but in Psoriatic arthritis (PsA) it is asymmetrical. Changes characteristic of Ankylosing Spondylitis, i.e fusion of the facet joints, costovertebral joints and sacroiliac joints, are conspicuously absent in DISH.

**DISH or Ankylosing Spondylitis?**

Differentiating symptomatic DISH from Ankylosing Spondylitis may not be easy clinically, especially as about 34% of patient with DISH is positive for HLA B27, the antigen present in other arthropathies like AS. Hence a brief discussion about Ankylosing Spondylitis will be quite appropriate.

AS, on the other hand, is a chronic inflammatory disease with inflammation of subchondral marrow at the cartilage-bone interface. An auto-immune reaction to an environmental pathogen in a genetically susceptible person is postulated. Pain is less with exercise and more at rest. Its association with HLA B27 and bacterial infection of the gastrointestinal and genitourinary tracts are well described. There is marked osteoporosis with an increased tendency for fractures. Ankylosis of the spine starting at the SI joints in an ascending pattern eventually results in the “bamboo spine” [10]. At the sites of insertion of ligaments, muscle to bone (‘enthesis’) calcification occurs at the sites of insertion of the Tendo Achilles, plantar fascia, and in the supraspinatus. AS is a systemic illness and causes extra-articular involvement- uveitis, aortitis, upper lobe pulmonary fibrosis and renal amyloidosis.

The involvement of SI joint is noticed at the early stage of the disease itself. It may be unilateral to begin with, but bilateral symmetrical involvement is a rule as the disease progress. The antero inferior synovial part of the joint is involved in AS where as postero superior cartilaginous part is affected in DISH. Involvement of synovial part of SI joint excludes the diagnosis of DISH.

The characteristic radiological changes of the spine in AS (**Figure 2**) is entirely different from that of DISH. The earliest vertebral change is an erosive osteitis at the corners of vertebral bodies followed by reactive sclerosisc (Shiny corner or Romanus sign). The anterior concavity of the vertebral body is lost and it becomes squared, again due to the erosive osteitis. There is evidence of discitis and ballooning of the disc. The presence of marginal syndesmophytes, vertically oriented bridging bony spicules from the vertebral margin is pathagnomonic for AS. A syndesmophyte is different from osteophyte both in site of origin and orientation. Osteophytes are non marginal in origin and are usually oriented horizontally where as the syndesmophytes are marginal in origin and vertically oriented. Eventually there is ossification of the outer fibres of annulus fibrosus and fusion or ankylosis of the vertebrae ensue with typical Bamboo spine appearance (Due to bulging discs and

undulating syndesmophytes over them). In contrast with AS, the disc spaces are maintained and there is no reduction vertebral heights in DISH. The new bone formation in DISH is described as 'flowing' rather than osteophytes or syndesmophytes. Inflammation and fusion of the apophyseal joints of the spine is a rule in AS, but apophyseal joints are not involved in DISH. Calcification of interspinous and supra spinous ligament is common in AS (Dagger sign). Other differentiating feature of DISH from AS is the absence of erosion at the enthesis, a pathognomonic feature of Ankylosing Spondylitis. As mentioned already the ligamentous clacification / ossification may suggest an alternate diagnosis of skeletal fluorosis, but for the absence of increased bone density.

**Figure 2:** Ankylosing Spondylitis - Bamboo spine, fusion of both sacroiliac joints, erosion and joint space narrowing of left hip

## Other Differential Diagnoses of DISH

Ochronosis caused by deficiency of homogentisic acid dioxygenase, may be considered as one of the differential diagnosis for DISH, though both conditions have their own independent clinical and radiological presentations. Ochronosis (Alkaptunuria) is a congenital disorder, caused by accumulation of this organic acid and manifests usually by the 4th decade with pigmentation of sclera, ear cartilage, synovium. Pain and swelling affects large joints, the thoracolumbar spine, but the lumbosacral spine is usually spared. Urine darkens on standing (alkaptonuria). Degenerative changes may be seen in peripheral joints with erosions. Osteophytes are less marked than other degenerative arthritides. Radiographs show chondrocalcinosis of intervertebral discs with spondylosis progressing to ankylosis.

Synovial Chondromatosis (Reichel's syndrome) is a benign proliferation of synovium, cartilage metaplasia and multiple multi-faceted intraarticular loose bodies with a radiologically similar picture. Young adults of age 30-50 years are affected; the male : female ratio is 2:1. Chromosome 6 abnormalities have been found occasionally. It is typically monoarticular. The knee is the most common location with stiffness, pain and swelling of the joint, worse with activity. There may be redness, warmth, tenderness and decreased range of movement. The histological changes are discrete hyaline cartilage nodules with varying calcification and ossification, and may be identified on radiographs as multiple (upto 200) calcified periarticular bodies of hyaline cartilage 1-3 mm in size filling the joint. Synovectomy and loose body removal prevent degenerative changes long term.

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