Diffuse idiopathic skeletal hyperostosis: case report and literature review

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Abstract

Background: Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a common disorder of unknown aetiology that is characterized by back pain and spinal stiffness. Diffuse idiopathic skeletal hyperostosis is a common disease, which is most prevalent in persons over 50 years of age. Several metabolic derangements and concomitant diseases associated with DISH include obesity, increased waist circumference, hypertension, dyslipidaemia, diabetes mellitus, hyperuricemia, metabolic syndrome and an increased risk for cardiovascular diseases. There is paucity of literature on case reports and prevalence studies in Africa especially with the increase of metabolic diseases. The condition is identified radiographically by the presence of “flowing” ossification along the anterolateral margins of at least four contiguous vertebrae and the absence of changes of spondyloarthropathy or degenerative spondylitis. However, DISH is not limited to the spine it may affect multiple peripheral sites independently. Extra-spinal entheseal ossifications are common and observing their isolated presence may lead to the diagnosis of DISH. Treatment should be aimed at symptomatic relief of pain and stiffness, and measures such as analgesics, NSAIDs, local applications and physiotherapy, might also prove to be useful in patients with DISH. Large-scale controlled studies are needed in order to delineate the entire spectrum of this condition. The role played by the metabolic and constitutional derangements as well as its impact on the diagnosis and treatment of DISH awaits further studies. In order to raise awareness of DISH this article tackles various aspects of DISH from symptomatology, pathophysiology to its management

Case presentation: A 55 year old obese man presented with a 12 month history of lower back pain. The pain was worse in the morning and associated with progressively worsening early morning stiffness. He had noted neck stiffness with forward stooping and some mild odynophagia to solid foods. He had no history of peripheral joint involvement, fevers, cough, bowel dysfunction or psoriasis. On examination he was noted to be obese with restricted movement both active and passive throughout the spine more marked in the neck and lower back. X-rays of the spine showed flowing mantles of ossification in the anterior longitudinal ligament extending from C2 to C6 and L1 to L4 vertebrae consistent with diagnosis of DISH.

Key words: DISH, Enthesopathy, Calcification, Metabolic syndrome

Introduction

Diffuse Idiopathic Skeletal Hyperostosis (DISH) is a skeletal disease characterized by the ligamentous ossification of the anterolateral spine. Forestier and Rotes–Querol¹ first described it more than 50 years ago as a disorder characterized by spinal stiffness, osteophytosis, and “flowing” new-bone formation about the thoracic spine. They termed it “senile ankylosing hyperostosis” and distinguished it from ankylosing spondylitis. Resnick et al² later termed this condition Diffuse Idiopathic Skeletal Hyperostosis (DISH). For diagnosis he used three strict radiographic features of the spine:

(i) Flowing calcification and ossification within the anterior longitudinal ligament involving at least four contiguous vertebral bodies, most commonly the thoracic spine
(ii) A minimal degree of degenerative disc disease
(iii) Absence of apophyseal joint ankylosis and sacroiliac erosions, sclerosis, or intra-articular osseous fusion.

Pathological entities that can be confused with DISH are osteophytes accompanying degenerative disease of the cervical spine, and ankylosing spondylitis.
Case presentation

A 55 year old obese man presented with a 12 month history of lower back pain. The pain was worse in the morning and associated with progressively worsening early morning stiffness. It radiated to the hips and upper back up to the neck and relieved by activity. He had noted neck stiffness with forward stooping and some mild odynophagia to solid foods. He had intermittent paraesthesias of the lower limbs with normal muscle power and normal sphincter function. He had no history of peripheral joint involvement, fevers, cough, bowel dysfunction or psoriasis. On examination he was noted to be obese with kyphosis and forward stooping of the neck. He had restricted movement both active and passive throughout the spine more marked in the neck and lower back. Laboratory work up revealed normal haemogram, ESR, CRP, urea, electrolytes, uric acid levels and calcium levels. X-rays of the spine showed flowing mantles of ossification in the anterior longitudinal ligament extending from C2 to C6 and L1 to L4 vertebrae consistent with diagnosis of DISH.

Figure 1: Lumbar radiograph of the case

Discussion

Diffuse idiopathic skeletal hyperostosis is most prevalent in persons over 50 years of age. The prevalence has been reported to be as high as 15% in women and 25% in men over the age 50 years and 26% in women and 28% in men over 80 years. Boachie-Adjei et al reported that 28% of the spines of subjects with an average age of 65 years had evidence of DISH. Mata et al reported a frequency of 2.5% to 10% in persons over age 70 years, with a slight male predominance. There is paucity of literature on case reports and prevalence studies in Africa. One study done in South Africa on black Africans prevalence of DISH was 3.9% (males 3.8% and females 4.2%). There was a rise in the prevalence of DISH with increasing age from 1% in the 40–49 year age group to 13.6% in those over 70 years. Diabetes was the most commonly associated risk factor at 52.4% in the 21 patients with DISH. Our patient was male and was 55 years which is in keeping with literature.

The aetiology of the condition remains unknown. It’s postulated that genetic, metabolic, endocrinology, anatomic, environmental, and toxic factors play a possible pathogenetic role in the new bone growth characterizing it. Metabolic disorders such as obesity, hyperlipidaemia, diabetes mellitus, and hypertension are frequent in patients with DISH. Our patient was obese. We have planned to do the above tests to rule out the above comorbidities to complete the evaluation of the patient.

DISH typically presents as a middle-aged or older patient with chronic mild pain in the middle to lower back associated with spinal stiffness that is worse in the morning or evening and the typical radiographic changes in the spine. The axial manifestations of DISH are the most frequent characteristic. Thoracic vertebrae are involved in almost 100% of affected individuals, lumbar vertebrae in 68-90% of these persons, and cervical vertebrae in 65-78% of affected individuals. Dysphagia secondary to large cervical osteophytes is an occasional complication of DISH. Interestingly, majority of the patients are asymptomatic, and DISH is an incidentally finding. The clinical findings are milder in comparison to the dramatic radiographic findings. Possible reasons for the minimal pain experienced by some patients is as a consequence of the relative stabilization of spinal segments through ankylosis. Our patient had a long history of the back pain and actually only reported to us when the back pain worsened and had developed the odynophagia.

Patients with DISH are at high risk for fracture and instability from even minor trauma. The increased incidence of fracture instability in DISH patients is due to ankylosis of the vertebral segments proximal and distal to the fracture, which creates increased lever arms that can cause displacement of the spine even in low-energy injuries. This may explain the delay in diagnosis and a high rate of immediate and delayed neurologic consequences. Hyperextension injuries are frequent, involving either disk disruption or fracture through the middle of a vertebral body. Patients with DISH, neck pain, and a history of trauma must be evaluated for occult fracture with Computed Tomography (CT) or Magnetic Resonance Imaging (MRI). The management plan of this patient should include education, fracture prevention and danger signs to look out when suspecting fracture. Treatment of fractures is similar to that of patients with other ankylosing conditions. The physician must consider the additional instability caused by poor ligament integrity and increased lever arms. It is important to note that the degree of instability is likely to be underrepresented on radiographs. Cervical traction may result in excessive
distraction due to the lack of ligamentous structures and should be used cautiously. The use of open reduction and internal fixation is recommended to prevent progression and delayed neurologic compromise.

Common extra-spinal manifestations include tendinitis and enthesophytes (osseous outgrowths at the sites of attachment of tendon, ligament, or capsule to bone). Many joints can be affected, and some patients have diffuse, vague aching similar to that of polymyalgia rheumatica. Subtle periostitis at the site of ligament or tendon insertion is often seen.

**Diagnosis**

A careful history delineating the nature and location of back pain is necessary. Back pain that is severe or acute in onset is unlikely to be related solely to DISH. The presence of extra-spinal musculoskeletal symptoms should be sought. There are no diagnostic laboratory findings, but evaluation may exclude other potential diagnoses (Table 1). The erythrocyte sedimentation rate and C-reactive protein, rheumatoid factor, and antinuclear antibody levels are typically normal. If DISH is suspected in an adult, the thoracic spine should be evaluated radiographically to establish the diagnosis (Table 2). Chest radiographs are adequate as screening tests for DISH. The lumbar spine is usually evaluated radiographically, as also the sacroiliac joints on these films can be helpful in ruling out other entities, such as seronegative spondyloarthropathies. Other sites of pain should be imaged with plain radiography, especially the heel, elbow, sacroiliac joints, and cervical spine. In non-traumatic situations, bone scans are not often helpful and can falsely give the appearance of multiple periarticular metastases.

**Table 1: Differential diagnosis of back pain and spondylophytosis**

<table>
<thead>
<tr>
<th>Diagnosis</th>
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<tbody>
<tr>
<td>DISH</td>
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<tr>
<td>Spondylosis deformans</td>
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<td>Ankylosing spondylitis</td>
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<td>Acromegaly</td>
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<td>Hypoparathyroidism</td>
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<td>Fluorosis</td>
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<td>Ochronosis</td>
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<td>Charcot spine</td>
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<td>Sternocostoclavicular hyperostosis</td>
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<td>Intervertebral osteochondrosis</td>
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<td>Spondylitic variants (e.g., psoriasis, Reiter’s syndrome, inflammatory bowel disease, Whipple’s disease)</td>
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<td>Pachydermoperiostosis</td>
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<td>Pseudogout</td>
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<td>X-linked hypophosphatemic</td>
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<td>Ostomalacia</td>
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**Table 2: Diagnostic criteria for DISH**

1. Flowing ossification along the anterolateral aspect of at least four contiguous vertebrae
2. Preservation of disk height in the involved vertebral segment; the relative absence of significant degenerative changes, such as marginal sclerosis in vertebral bodies or vacuum phenomenon
3. Absence of facet-joint ankylosis; absence of sacroiliac erosion, sclerosis, or intra-articular osseous fusion

**Therapeutic considerations**

The pathogenesis of the disease is not clear, and therefore the current therapeutic interventions are empirical. Aim of treatment is symptomatic relief of pain and stiffness. For patients with isolated back pain or enthesopathies treatment options include activity modification, physical therapy, corset or brace wear, nonsteroidal anti-inflammatory medications, and bisphosphonate therapy are the mainstays of treatment. The literature on efficacy of these modalities has not been well established. Control of associated metabolic disorders, including obesity, hypertension, hyperinsulinaemia (with or without hyperglycaemia), dyslipidaemia, hypertriglyceridaemia and hyperuricemia, may reduce the morbidities associated with these disorders. These may retard future cardiovascular disease and possibly slow down the progression of soft tissue ossification. Therapeutic interventions are aimed at a reduction of insulin secretion and insulin resistance. In patients with non-insulin-dependent diabetes, the use of biguanides, which induce a better usage of insulin, may offer an advantage over the use of sulphonylureas that increase insulin secretion. Coexisting hypertension the choice of drugs that might improve insulin resistance such as angiotensin-converting enzyme inhibitors, calcium channel blockers and beta-blockers should be preferred over drugs that might worsen insulin resistance, such as thiazide diuretics. Generally, surgery is not indicated for DISH in the absence of some other diagnosis, such as fracture, stenosis, tumour, infection, or painful deformity. Fortunately, debilitating pain is rare in the absence of neurologic or visceral impingement, probably because bony ankylosis prevents painful motion.

**Conclusions**

Diffuse idiopathic skeletal hyperostosis is a very common, often occult bone-forming diathesis with many musculoskeletal manifestations. Data from Africa is rare. There has been an increase in metabolic disorders associated with DISH. We hope this article will raise the awareness of this disease. A diagnosis of DISH should be suspected in adult patients who present with back pain.
pain and spinal stiffness especially in the background of metabolic disease. The diagnosis is based on the presence of flowing ossification along the anterolateral aspects of at least four vertebrae, typically in the thoracic spine. Radiographic evaluation of the thoracic spine must be performed, even if pain is localized to the lumbar or cervical areas. All other areas of musculoskeletal pain should be evaluated radiographically, looking for hyperostosis or enthesopathy. Awareness of the disorder and a high index of suspicion can add a great deal to the clinical acumen of practitioners in rheumatology and orthopaedic subspecialty. Treatment should be aimed at symptomatic relief of pain and stiffness, and measures such as analgesics, NSAIDs, local applications and physiotherapy, might also prove to be useful in patients with DISH. Large-scale controlled studies are needed in order to delineate the entire spectrum of this condition.

The highlights of the study are as follows:
(i) DISH is a common disease, but is under reported in our local set up. Doctors need to have a high index of suspicion
(ii) Musculoskeletal manifestations of DISH are not limited to the spine and often affect peripheral sites
(iii) DISH is often associated with metabolic and constitutional derangements, leading to an increased cardiovascular risk
(iv) Prevention of sequale in DISH is paramount
(v) Management is multi-disciplinary comprising of rheumatologists, surgeons and physiotherapists.

Conflict of interest

The authors confirm that this article content has no conflict of interest.

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References