



Diffuse Idiopathic Skeletal Hyperostosis: a Distinct Clinical Entity

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Abstract

Diffuse idiopathic skeletal hyperostosis is often incorporated into osteoarthritis. Although DISH often coexists with OA, patients affected by this disorder differ from patients with primary OA in several aspects: prevalence in the general population, gender distribution, anatomic site of primary involvement, magnitude and distribution in the spine and the peripheral joints. DISH is a distinct clinical entity. Its recognition as such should stimulate clinicians and researchers to focus on its pathogenesis, treatment and prevention.

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Diffuse idiopathic skeletal hyperostosis is a condition characterized by calcification and ossification of soft tissues, mainly ligaments and entheses. This condition was described by Forestier and Rotes-Querol over 50 years ago [1] and was termed senile ankylosing hyperostosis. There is a marked predilection to the axial skeleton, particularly the thoracic spine. However, the recognition that the condition is not limited to the spine and may involve peripheral joints led researchers to coin the name DISH, a term widely used [2]. The disease is characterized by the production of flowing osteophytes involving, in particular, the right side of the thoracic spine with preservation of the intervertebral disk space [Figure 1], and by ossification of the anterior longitudinal ligament. Other enthesal regions in the peripheral joints might be affected, such as the peripatellar ligaments, at the Achilles tendon insertion, plantar fascia, olecranon and others [3,4].

In the absence of validated diagnostic criteria the diagnosis is usually based on the definition suggested by Resnick and Niwayama [4]. This radiographic approach requires the presence of right-sided, flowing, coarse osteophytes in the thoracic spine, connecting at least four contiguous vertebrae, or ossification of the anterior longitudinal ligament, preserved intervertebral disk height in the involved segment, and the absence of apophyseal joint ankylosis and sacroiliac joint involvement [4].

The inflammatory spondyloarthropathies are usually easily distinguishable from non-inflammatory conditions. The clinical history, physical examination, extra-articular features, laboratory results, and various imaging modalities help in reaching the correct

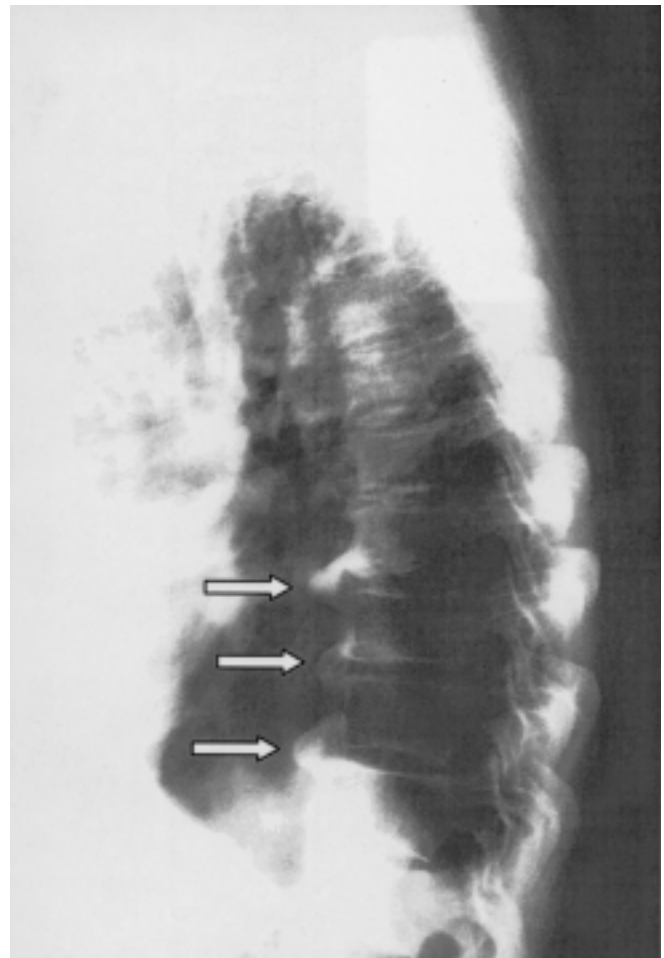


Figure 1. Lateral radiography of the thoracic spine. Anterior course, flowing osteophytes (arrows) with preserved intervertebral height.

diagnosis. This review assesses whether DISH can be distinguished as a separate entity and not confounded with primary osteoarthritis, although the two conditions may coexist. The pathogenesis is not fully understood, but several factors have been implicated in the disease based on frequent associations with various metabolic conditions. Some of these factors are: hyperinsulinemia with or without diabetes mellitus [5], obesity, gout [6], dyslipidemia [7], and prolonged use of isotretinoin [8].

DISH = diffuse idiopathic skeletal hyperostosis
 OA = osteoarthritis

In most textbooks DISH is described as a subgroup of osteoarthritis. Often the disease is asymptomatic and is accidentally found by radiographs of the thoracic spine performed for diverse reasons. Recently, the use of chest X-rays for the diagnosis of DISH has been validated [9]. Because of the lack of specific symptoms and signs on the one hand, and the radiographic diagnostic basis on the other, it was considered by some to be a condition and not a disease [10]. A thorough review of the existing literature suggests that DISH, although affiliated to OA, is a distinct clinical entity with somewhat different characteristics.

Spinal involvement

Classically, the portions of the spine that are usually involved in OA are the lower portions of the cervical spine and the lumbar spine, as opposed to the mandatory involvement of the thoracic spine in DISH. Furthermore, T-spine involvement in DISH is characterized by preserved intervertebral height, while in spinal OA a reduced intervertebral disk involvement is common. These differences in the radiologic appearance and anatomic spinal distribution are probably due to different pathogenetic mechanisms. It is presumed that the primary target for the osteoarthritic process is the cartilage, represented in the spine by the intervertebral disks and the cartilage of the facet joints. The wear and tear forces operating in the very mobile lower cervical and lumbar portions of the spine might explain the frequent involvement of these segments in OA. The T-spine is the least mobile of the spinal segments, suggesting different pathogenetic mechanisms affecting different anatomic structures in DISH. In fact, the main targets of the disease in DISH are the spinal ligaments and entheses [11]. The ossification and calcification of these particular structures is responsible for the characteristic radiographic appearance. These sites of ossification and the subsequent production of large osteophytes may cause severe clinical manifestations, especially when the C-spine is affected. These manifestations include dysphagia [12], quadriplegia [13], esophageal obstruction [14], dyspnea and hoarseness [15], atlantoaxial subluxation [16] and others. The high prevalence of coexisting intervertebral disk damage in relatively young patients with DISH suggests an important role for this condition in the pathogenesis of spondylosis in this group of patients [17].

Although the disease may be asymptomatic, it was reported to be associated with dorsolumbar pain and stiffness in the majority of patients [3,4]. A recent study comparing DISH patients with healthy controls confirmed these findings but failed to show a significant difference when the patients were compared to patients with spondylosis [18]. However, a significant reduction in the range of spinal movement was noted.

Peripheral joint involvement

There is no doubt that clinical manifestations similar to or identical to those of OA are prominent features of DISH in the peripheral joints. However, the peripheral joints affected by DISH have

features that distinguish them from primary OA. One is the more frequent involvement of joints that are not usually affected in OA, such as metacarpophalangeal joints, elbows and shoulders [19–22]. Another feature is a more severe hypertrophic disease in the joints affected by DISH [23]. These peculiar characteristics have important pathogenetic implications. OA is perceived as a disease of the cartilage. This is usually accepted as the primary event in weight-bearing joints such as the knees and hips. However, this assumption alone does not hold for small, non-weight-bearing joints such as the proximal interphalangeal joints and distal interphalangeal joints. It was suggested that the osteoarthritic process in these small non-weight-bearing joints is caused by an increased intraarticular pressure and subsequent development of “crash” forces [24]. This was attributed to thickening of the collateral ligaments of these joints, which enforce a constraint movement, and not to primary damage to the cartilage. As described previously, the primary event in DISH is thickening, calcification and/or ossification of ligaments and entheses. In particular, enthesopathy affecting the peripheral joints has been described [25]. The radiographic appearance of peripatellar, cruciate ligament insertion, and pericapsular osseous enthesopathies are just some examples of the contribution of DISH to stiffening of the soft tissues surrounding a joint [26]. Therefore it seems reasonable that the joints affected by DISH develop or worsen the same “crash” forces operating in small joints, as a result of this additive mechanism. This might explain both the involvement of “atypical” joints not commonly affected by OA, and the hypertrophic osteoarthritic changes in the commonly affected joints.

Epidemiology

OA is the most common joint disease in the world. Its prevalence rises with advanced age, reaching 75% in patients over the age of 70. DISH is also more common in those over 50 years old. The reported prevalence for DISH varies among different studies. It was reported by some to be slightly above 10% in patients over 70 years old [27,28]. In other studies with different populations, the prevalence in men and women over age 50 was reported to be 25% and 15% respectively, and for ages above 70 years 35% and 26% respectively [29]. Therefore, the prevalence of OA is 2–7 times that of DISH, and while OA invariably occurs in advanced age, DISH is by far less frequent and therefore is not related solely to the process of aging. In contrast to primary OA, males are more often affected by DISH [27,30], supporting the hypothesis of different pathogenetic mechanisms in these two conditions.

Conclusion

Since both primary OA and DISH are diseases of the older age groups, it is not surprising that they often coexist or enhance some of the clinical manifestations of OA. In recent years our understanding of OA has greatly improved, and some prospective for effective treatments other than palliation are emerging. Although DISH might contribute to the generation of OA, or worsen primary OA, it should be regarded as a separate clinical entity, with different prevalence, joint distribution, gender

C-spine = cervical spine
T-spine = thoracic spine

distribution, and pathogenetic mechanisms. Its recognition as a distinct clinical entity will stimulate a more targeted research and, hopefully, better solutions based on a true understanding of the pathogenetic mechanisms underlying this common, yet under-recognized disease.

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