EDITORIALS

CLINICAL MANIFESTATIONS OF DIFFUSE IDIOPATHIC SKELETAL HYPEROSTOSIS (DISH)

In 1948, Forestier and Rotés-Querol [1] reported on nine older patients (50–73 yr of age) who suffered from spinal rigidity, only a third of them having had mechanical spinal complaints. Radiological studies showed exuberant osteophytes, with a tendency of spreading in front, and particularly the right side of the vertebral bodies. Neither the sacroiliac nor the posterior interapophyseal joints showed arthritis. These exuberant osteophytes were felt to represent an ossification of the vertebral ligamentous system (the 'perirraquitis' of Reiner [2]). The condition appeared different from ankylosing spondylitis and vertebral spondylosis, from which it needs to be distinguished. They named it hiperostosis anquilosante vertebral senil (HAVS) (senile vertebral ankylosing hyperostosis), and it was later coined diffuse idiopathic skeletal hyperostosis (DISH).

With the passing of time, the condition was recognized as an independent disease, particularly when it was noticed that it did not share with ankylosing spondylitis its association with HLA B27, and that some of its sufferers also had ossification of the periostium at the site of attachment of the enthesis and ligaments of peripheral joints, producing on occasions a peripheral joint disease with precise radiological characteristics [3]. At the spine, the process begins with calcification of the anterior longitudinal ligament, followed by cartilaginous metaplasia and posterior ossification [3]. Prevalence studies based on the radiological characteristics have shown that from 2.4 to 5.4% of those older than 40 yr suffer from it, as well as 11.2% of those above 70 [4, 5]; necropsy studies have shown similar figures.

Initially, Forestier and Rotés-Querol considered HAVS as a radiological alteration, spectacular but innocuous. It was usual then, and still is, for these images to be overinterpreted by the radiologist—and often by the clinicians—who frequently ignore its nature, and/or mistake them for vertebral spondylosis. The radiological characteristics are now generally well known, and it has become apparent that DISH is a cause of clinical alterations, some of them severe [6], which have received little attention. In this editorial, I review the clinical manifestations of DISH, which can be systematized as follows.

(1) Spinal rigidity of variable degree, which may pass unnoticed by the patients. Whenever an older patient presents with significant cervical or lumbar loss of motility in the absence of a history of pain, the possibility of hyperostosis should be entertained.

(2) Syndromes caused by dynamic overload of the 'mobile segment'. Patients with DISH suffer from acute or chronic lumbar, cervical or dorsal pain, sometimes with associated root compression [5], with a higher frequency than those without it. During the years in which hyperostosis forms, ankylosed vertebral segments co-exist with adjacent discs or unaffected 'mobile segments'. Quite frequently, there is a topographic relationship between the clinical manifestations and some of these overloaded 'mobile segments', which on occasions develop destructive or spondylotic changes.

(3) Syndromes produced by space occupation. Protrusion of hyperostotic formations into the spinal canal can occur, producing medular compression, or outwards, pressing on the oesophagus, trachea or larynx.

(i) The syndromes of medular compression due to ossification of the posterior longitudinal ligament, described initially by Japanese authors [7, 8] are, after spondylosis, the second cause of cervical myelopathy. Initially it was felt to be a process restricted to the Japanese population, but its more widespread occurrence is becoming recognized, particularly after the introduction of CAT scan and MRI, which allows the identification of the typical band due to the ossified posterior longitudinal ligament, located along the posterior surface of the vertebral bodies; the ligament is much thicker in the cervical spine than in the lower segments [9]. Occasional compression of the medulla of a similar nature has been reported in the dorsal or lumbar spine, or even in the cauda equina due to ossification of the yellow ligaments. Frequently, surgeons do not recognize the radiological images of hyperostosis, and treat them similarly as they do spondylosis. The recognition of hyperostosis is not without importance when planning surgery, since generally it is not necessary to fix the cervical spine—already fixed by hyperostosis—and the procedure could be limited to a section of the posterior laminae, opening a window to widen the canal, without removing the ossified posterior longitudinal ligament. Myelopathy may also occur in the dorsal spine; here the hyperostosis lies more frequently in the posterior structures of the canal: yellow ligament or capsules of the interapophyseal joints.

(ii) Dysphagia. In 1973, Meeks and Renshaw [10] published on two patients with dysphagia due to compression by large hyperostotic growths in patients with hyperostosis. The oesophagus is fixed at its extremes in the cricoid bone and the diaphragm. Dysphagia due to external bone compression is produced close to the superior fixation point at C6, either above it, due to posterior pressure on the hypopharynx or even the pharynx, at which posterior wall a protuberance can be appreciated, or below it.
The intensity of dysphagia can be very variable, from very mild, to a severe difficulty allowing only the swallowing of liquids, and associated with hoarseness, respiratory difficulty or loss of weight mimicking an oesophageal tumour. A barium swallow clarifies the cause. It is of interest to the surgeon to distinguish between a hyperostotic formation and an osteoarthritic osteophyte, because in the first case it is sufficient at surgery to remove the hyperostotic formation, without the need for a more complex procedure such as fixing the underlying vertebrae, which may be necessary after removing an osteoarthritic osteophyte, to stabilize the vertebrae.

(4) Hyperostosis in peripheral joints. Hyperostosis is the cause of clinical manifestations at three different peripheral structures: hips, feet and shoulders. At the hip joint, hyperostosis produces characteristic bony proliferations. The most frequent of them is a square or triangular hyperostotic growth located just above the outer margin of the cotilum; its base is wide and the external borders are irregular and without a cortical. Also, in the inner and more inferior edge of the cotilum, another bony proliferation surrounds the more inferior and internal part of the femoral head. It has been debated whether these changes should be considered as an independent hip disease or simply as a variety of secondary hip osteoarthritis [11]. I think that it is of importance to note that in the majority of occasions, the hip joint space is preserved and that only \( \sim 15\% \) show some pain. A minority of these hips, generally in the latter stages, develop osteoarthritis, which can at times be severe and destructive. The diagnosis of osteoarthritis, and the associated probability of hip replacement surgery, should be avoided in those hips which simply show hyperostotic proliferations in the cotilum, without thinning of the joint space. Although the radiographic appearance of DISH affecting the hip joint is characteristic, antero-posterior and lateral radiographs of the dorsal spine may help to establish the diagnosis in doubtful cases. I have followed for as long as 30 yr a number of patients who were originally scheduled for hip replacement surgery, and who continue walking at present with their own hips, without any need for anti-inflammatory medication. In those patients in whom there is progress to severe osteoarthritis, requiring the placement of a hip prosthesis, the development of periarticular ossifications may hamper the results of the procedure [12]. These ossifications generally appear in the first year after surgery, and seem unrelated to the technique. They may progress to the point of fusing the new hip, generally in a flexed position. Attempts to halt them with indomethacin, diphosphonates or radiotherapy have been made without success.

Heel pain associated with large spurs under or behind the calcaneus may eventually require surgery. Just a curiosity, which may be bothersome, is the presence of abundant hyperostosis in the feet ('bristly feet') [13] or the 'late hyperostotic hallux rigidus'. In the shoulder, hyperostosis may produce ossification of the coraco-acromial ligament, contributing to the production of rotator cuff lesions.

The difficulty in clarifying some of the clinical manifestations of DISH, and understanding its pathogenetic mechanisms [14], lies in the diversity of its clinical expression, which presents to different specialists [15]. Probably, the formation and meeting of a group of physicians from the different medical fields in which the disease expresses itself would be useful better to define and understand all of its manifestations, as has been the case in other diseases of similar multidisciplinary characteristics, such as Behçet's disease.

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REFERENCES