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Evolutionary particularities in Diffuse Idiopathic Skeletal Hyperostosis associated with Spinal Stenosis – case report

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ABSTRACT

Diffuse idiopathic skeletal hyperostosis is a non-inflammatory disease, that affects mostly males and the principal manifestations are calcification and ossification of spinal ligaments, calcification of the tendons and ligaments in the vertebral body insertion. Clinical manifestations of the disease are mainly in the thoracic spine, but it may involve also the cervical and the lumbar spine. Extraplural involvements are frequently present at patients with diffuse idiopathic skeletal hyperostosis. Spinal stenosis is associated with this disease.

The present case is of a 56 years old man, initially diagnosed with Ankyloses spondylitis, who was under a treatment with Sulfasalazin for approximately 7 years, afterwards the diagnosis was infirmed and he was diagnosed with “Diffuse idiopathic skeletal hyperostosis”.

The patient has multiple extra spinal manifestations of the disease (shoulders, elbows, hips, knees), cervical and lumbar spinal stenosis with clinical manifestations and imagistic confirmation, and also treated in neurosurgical service for several times, for the lumbar spine pathology. Particularities of this case are related with the delay of a correct diagnosis which means that the anterior treatments, both medical and surgical are under the questions.

Keywords: diffuse idiopathic skeletal hyperostosis, spinal stenosis, extra spinal manifestations

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Introduction

Diffuse idiopathic skeletal hyperostosis (DISH) has several synonyms, including ankylosing hyperostosis and Forestier’s disease. It is a non-inflammatory disease, with the principal manifestation being calcification and ossification of spinal ligaments and the regions where tendons and ligaments attach to the bone (entheses). Radiographic changes characteristic of DISH may occur in the absence of any musculoskeletal symptoms. It is more common in men than women and the incidence varies by population and increases with age, rarely being diagnosed before the age of 40[1].

Case report

A 56 year old male, from urban area, known in the clinic from 2011 when he was diagnosed with Diffuse idiopathic skeletal hyperostosis, was admitted to the Rehabilitation Hospital Eforie Nord for pain in the cervical, thoracic and lumbar spine with mechanical characters, mainly in the lumbar spine with irradiation on the lateral side of both legs, with paresthesia at the level of the calf and the foot, for urinary incontinence, erectile dysfunction and mechanical pain in the knees and both shoulders. The patient’s symptoms started insidious, in 1982, with

pain in the thoracic and lumbar spine, apparently almost permanent, irradiation of the pain in the right leg unsystematically and functional thoracic and lumbar spine disability.

The patient was diagnosed in 1993 with Ankylosing spondylitis and was treated with Salazopirin and anti-inflammatory nonsteroids (NSAI) for 7 years, in 2010 after detailed investigations the diagnose was invalidated. The patient was known to have multiple pathologies: tonsillectomy, meniscectomy on the left knee (1992), right inguinal hernia with surgical treatment for several times (re-operated for postsurgical infection), herniated disc at L4-L5 level compressible at the right side with surgical treatment for several times (twice in 2003 and once in 2006), postsurgical thrombophlebitis on the left leg (2006), hypothyroidism (being under treatment with levothyroxine 100µg per day).

The patient told us he was a mechanic in a chemical fertilizer factory, being a toxic environment especially with phosphorus and fluorine.

On hospitalization the physical examination showed that the patient had a relatively good condition, obese : body mass index (BMI) = 34.7, consciously, co-operating, cardio-pulmonary, digestive and mentally balanced, normally colored, warm and wet teguments, with supple, unpainful postsurgical scars at lumbar spine, right inguinal and left knee.



Figure 1 - Lumbar hyperlordosis

The spine examination highlights that the patient have lumbar hyperlordosis (as shown in Figure 1), with obvious sensitiveness at the palpation of the cervical, thoracic and lumbar spinous processes, retraction of the paravertebral muscles of the cervical, thoracic and lumbar spine; mobility of the cervical spine: value of the menton-sternum distance = 7cm, value of the occiput- thoracic spine distance = 8cm, value of the tragus – left shoulder distance = 12 cm, value of the tragus-right shoulder distance = 13cm; mobility of the thoracic spine: Ott sign = +3cm; mobility of the lumbar spine: fingertips-to-floor distance = at the knee, Schöber sign = +3cm, lateral lumbar inflexibilities and extension of the lumbar spine slightly painful and limited, without nerve root syndrome at the cervical spine, with bilateral L5 nerve root syndrome, negative nerve stretching signs, normal deep tendon reflexes on the superior and inferior limbs, excepting the Achilles reflex and the plantar reflex that are slightly diminished, without motor deficits, positive un-systematized Romberg test.



Figure 2 - Cervical spine radiography in posterior-anterior angle of incidence and profile incidence.

The shoulders examination shows pain on palpation and mobilization on both shoulders with cracking on mobilization, normal passive mobilization and painful limitation of the active mobilization on maximum amplitude movements; painful bilateral elbows on palpation without limitation in active and passive mobilization, Heberden nodes at the second fingers bilateral and on the Vth right finger. The hips examination shows positive Patrick test on the right hip and a slight painful limitation of the mobility.

The knees are in axle with pain on palpation and mobilization, cracking on mobilization with negative instability tests.



Figure 3 - Lumbar spine and sacroiliac joints radiography in posterior-anterior angle of incidence

The radiographic investigations shows: the cervical spine radiography in postero-anterior angle of incidence and profile incidence (Figure 2), the lumbar spine and the sacroiliac joints radiographies (Figure 3) in postero-anterior angle of incidence are without osteo-articular changes. The left shoulder radiography (Figure 4) and the left elbow radiography (Figure 5) highlights the narrowing of the articular spaces with periarticular marginal osteophytes; the right knee radiography shows the narrowing of the articular space, osteophytes at the marginal tibial plateau and a calcified Baker cyst.



Figure 4 - Left shoulder radiography in posterior-anterior

angle of incidence.



Figure 5 - Left elbow radiography – profile.



Figure 6 - Right knee in posterior-anterior angle of incidence

The lumbar spine magnetic resonance imaging (MRI) (Figure 7) highlights disk protrusion at L4-L5 level with the formation of disco-osteophytes complex

at L4-L5 bodies that has an intracanal contact with the right L5 nerve root and stenosis type changes at the medullary canal from L3 to S1 levels.

The cervical spine MRI (Figure 8) shows stenosis type changes at the medullary canal from C5 to C7 levels, with disco-osteophytes complex that compress the left C6 nerve root and the right C7 nerve root.

The patient does not have an inflammatory syndrome, parathyroid hormone, thyroid-stimulating hormone, serum calcium, creatine kinase, serum phosphorous are all in normal limits, except the minimal intolerance at oral glucose tolerance test and a slight hypercholesterolemia.



Figure 7 - Lumbar spine MRI (without colouring agent)

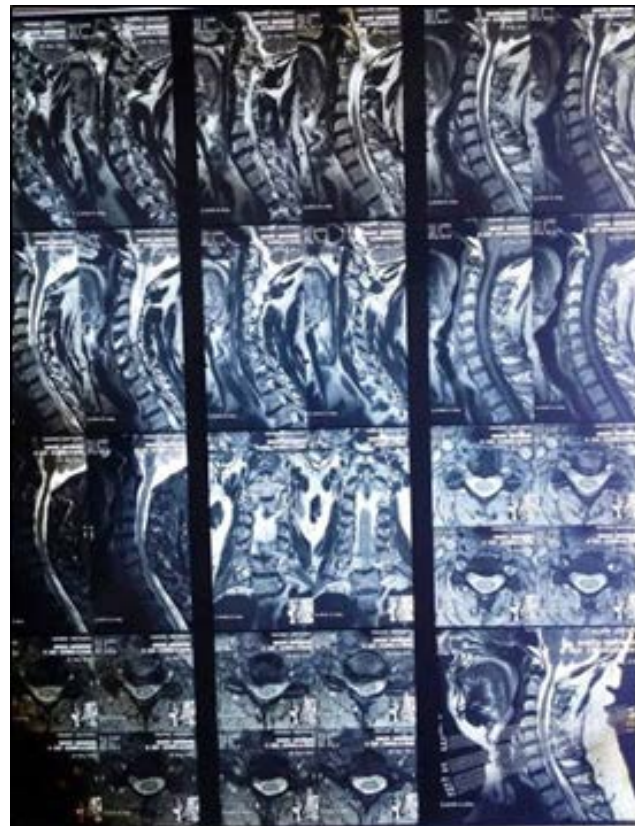


Figure 8 - Cervical spine MRI (without colouring agent)

Discussion

Forestier's disease was first described by Jacques Forestier and his student Jaume Rotes-Querol in 1950 under the name "senile ankylosing vertebral hyperostosis". In 1976, Resnick and Niwayama coined the term "diffuse idiopathic skeletal hyperostosis" (DISH) for the systemic involvement[2].

Resnick and Niwayama defined DISH as the:

- 1) radiographic findings of calcification or ossification along the anterolateral aspects of the least four contiguous vertebral levels across 3 disc spaces;
- 2) preservation of the disc height in the involved vertebral segments without degenerative disc disease;
- 3) absence of sacroiliac inflammatory

changes[1,3,4].

Later on Utsinger defined other criteria to diagnose DISH:

1) continuous ossification along the anterolateral aspect of the least four contiguous vertebral bodies, primarily in the thoracolumbar spine. The ossification begins as a fine wave of bone but commonly develops into a broad band of bone;

2) continuous ossification along the anterolateral aspect of at least two contiguous vertebral bodies;

3) symmetrical and peripheral enthesopathy involving the posterior heel, superior patella, or olecranon, with the enthesal new bone having a well-defined cortical margin. The exclusion criteria: 1) abnormal disc space height in the involved areas; 2) apophyseal joint ankyloses. Defining criteria for DISH = criteria 1. Criteria of probability for DISH = criteria 2 and 3[1].

The etiology of the disease remains unknown, but some factors have been discovered as being important, such as: mechanical factors (the presence of marginal osteophytes, continuous ossification along the anterolateral aspect of two-four contiguous thoracic vertebral bodies), external environmental and diet factors (the presence of fluoride in the water and air, the excess of vitamin A), certain drugs (long-term medications with derivatives of vitamin A), metabolic conditions (the insulin-like growth hormone stimulates the osteoblasts, obesity, diabetes, elevated blood pressure, elevated levels of insulin are all associated with the disease, and in 20% of the cases it is associated acromegaly)[1,2,4].

Forestier's disease affects mainly males aged between 40 and 60 years[1].

The clinical manifestations of the disease depend on the affected regions of the body, but sometimes it can be asymptomatic for a long time. Most symptoms occur due to biomechanical damage of the spine, leading to pain, stiffness and a degree of decrease range of motion of the spine and of the peripheral joints[1]. The most frequently affected joints are: pelvis, hips (bone proliferation, ligament ossification, osteophytes), knee and patella, ankle and foot, shoulder and hands[1,2,5]. There are some other symptoms that may occur such as: respiratory symptoms (dysphonia), gastrointestinal (dysphagia) and even neurological due to bone compression on

adjacent structures[4,6,7,8].

The most frequently affected region of the spine is the thoracic region (96%), followed by the lumbar (90%) and cervical (78%)[2]. Regarding the cervical region the most affected are C4-C7 vertebrae and there are some associated symptoms beside the limitation of mobility, patients can suffer due to the compression of the spinal canal some neurological symptoms such as dizziness, sore throat, ear pain, dysphagia, stridor, sleep apnea or luxation of the atlanto-axial joint[8]. The osteophytes on the right side of the vertebral bodies are characteristic for the thoracic spine, with preservation of the intervertebral spaces [1,2]. In lumbar spine we find characteristic the bone bridges, intervertebral space narrowing and lumbar fractures[8,9,10].

Many studies revealed the coexistence of DISH with gout, rheumatoid arthritis, Paget's disease, and because the prevalence of the disease is after the age of 45-50 years old, it may be associated with other diseases, the most frequently is the metabolic syndrome[1,2].

In the evolution of the Forestier's disease it may occur the spinal stenosis. Spinal stenosis refers to the factors or conditions that lead to the compression of the spinal canal, the lateral recesses and by reduction of the foramen diameter of the spinal canal, all these can lead to the compression of the spinal cord [11]. Symptoms that can occur due to constriction of the nerve root: paresthesia in the upper or lower limbs that are intensified with effort and improved with rest, muscle weakness or muscular atrophy, neurogenic claudication, sphincter disorders (urinary incontinence)[11,12,13].

There is no specific treatment for DISH, only symptomatic treatment and there wasn't any controlled study to assess the effects of certain drug therapies. The balneophysiokinetotherapy treatment is indicated due to the effects on the pain relief and increasing mobility. The surgical treatment is indicated in some cases for dysphagia or nerve root compression[1,13].

Conclusions

The DISH diagnostic was delayed, after an uncertain diagnosis of ankylosing spondylitis for which the patient followed a prolonged treatment with Sulfasalazine, he suffered neurosurgical treatment several times for a herniated disc at L4-L5 level and the last MRI highlights cervical and lumbar spinal stenosis. In this case the disease affects most the peripheral joints (shoulders, elbows, hips and knees). The patient has associated a metabolic syndrome which is a predisposing factor for the Forestier's disease.

References

1. Helfgott, S.M., Tugwell, P. & Romain, P.L. (2015). *Diffuse idiopathic skeletal hyperostosis (DISH)*, UpToDate, Retrieved Aprilie 6, 2015, from UpToDate database on the World Wide Web : <http://www.uptodate.com>
2. Nascimento, F.A., Gatto, L.A.M., Lages, R.O., Neto, H.M., Demartini, Z. & Koppe, G.L. (2014). Diffuse idiopathic skeletal hyperostosis : A review, *Surg Neurol Int*, 5(Suppl 3), S122-S125, DOI: 10.4103/2152-7806.130675
3. Holton, K.F., Denard, P.J., Yoo, J.U., Kado, D.M., Barrett-Connor, E. & Marshall, L.M., Osteoporotic Fractures in Men (MrOS) Studz Group (2011), Diffuse idiopathic skeletal hyperostosis (DISH) and its relation to back pain among older men: The MrOS Study, *Semin Arthritis Rheum*. 41(2), 131-138, DOI: 10.1016/j.semarthrit.2011.01.001
4. Zhang, C., Ruan, D., He, Q., Wen, T., Yang, P. (2014), Progressive dysphagia and neck pain due to diffuse idiopathic skeletal hyperostosis of the cervical spine: a case report and literature review, *Clinical Interventions in Aging*. 9: 553-557, DOI: 10.2147/CIA.S60146
5. Terzi, R. (2014). Extraskelatal symptoms and comorbidities of diffuse idiopathic skeletal hyperostosis, *World J Clin Cases*. 2(9): 422-425, DOI: 10.12998/wjcc.v2.i9.422
6. Pillai, S. & Littlejohn, G. (2014), Metabolic factors in diffuse idiopathic skeletal hyperostosis – a review of clinical data, *The Open Rheumatology Journal*. 8, 116-128 , DOI: 10.2174/1874312901408010116
7. Najib, J., Goutagnz, S., Pezre, M., Faillot, T. & Kalamarides M (2014 Mar.) Forestier's disease presenting with dysphagia and dysphonia, *PanAfrican Medical Journal*. 17, 168, DOI: 10.11604/pamj.2014.17.168.2453
8. Storch, M.J., Hubbe, U. & Glocker, F.X., (2007). Cervical myelopathy caused by soft-tissue mass in diffuse idiopathic skeletal hyperostosis, *Eur Spine J*. 17(Suppl 2), S243-S247, DOI: 10.1007/s00586-007-0508-6
9. Hasegawa, K., Takahashi, H., Iida, Z., Zokozama, Z., Fukutake, K., Takamatsu, R., Nakamura, K., Suzuki, D., Shishikura, W., Tsuge, S., Sekiguchi, M. & Wada, A. (2013). Spontaneous Symptomatic Pseudoarthrosis at the L2-L3 Intervertebral Space with Diffuse Idiopathic Skeletal Hyperostosis: A Case Report, *Orthopedics*. 2013, Article ID 497458, 4 , DOI: 10.1155/2013/497458
10. Diederichs, G., Engelken, F., Marshall, L.M., Peters, K., Black, D.M., Issever, A.S., Barrett-Connor, E., Orwoll, E., Hamm, B., Link, T.M. & Osteoporotic Fractures in Men (MrOS) Research Group, (2010). Diffuse idiopathic skeletal hyperostosis (DISH) relation to vertebral fractures and bone density. *OsteoporosInt*. 22, 1789–1797, DOI: 10.1007/s00198-010-1409-9
11. Levin, K., Aminoff, M.J., Atlas, S.J. & Wilterdink, J.L., (2013). *Lumbar spinal stenosis: pathophysiology, clinical features, and diagnosis*, UpToDate, Retrieved Aprilie 9 2015, from UpToDate database on the World Wide Web : <http://www.uptodate.com>
12. Levin, K., Aminoff, M.J. & Wilterdink, J.L., (2013), *Cervical spondylotic myelopathy*, UpToDate, Retrieved Aprilie 9 2015, from UpToDate database on the World Wide Web : <http://www.uptodate.com>

13. Lucescu, V., (2009). *Afecțiunile degenerative ale coloanei vertebrale – Clinica, diagnosticul și tratamentul de recuperare, Note de curs*, Constanța: Editura Dobrogea.