GROWTH PROGRESSION OF FORESTIER'S DISEASE IN THE CERVICAL SPINE

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ABSTRACT

Forestier's disease is known as a diffuse idiopathic skeletal hyperostosis (DISH) and the nature of its progression is not completely understood. A 50-year-old female was admitted with neck pain, headaches for 22 years and non-progressive difficulty in swallowing for 10 years. In 1977, there was a small osteoid formation in front of the C5-6 intervertebral disc space. By 1985, this had grown larger and had come into contact with new osteophytes in C5 and C6 bodies. New osteophytes subsequently combined with the ossification in the anterior longitudinal ligament (ALL) at C5 and C6. In 1991, new osteophytes were observed in C4 and C7 bodies. In 1999, there was a large skeletal hypertrophy in ALL at C4-5-6 associated with large new osteophytes in C3, C7 and T1 bodies. This report illustrates gradual radiological progression of a small osteoid formation to DISH. It should be considered that small osteoid formations may have a potential growth progression to DISH.

Key Words: Diffuse idiopathic skeletal hyperostosis, Dysphagia, Progression

ÖZET

SERVİKAL OMURGADAKİ FORESTİER HASTALIĞINDA BÜYÜME PROGRESYONU

Forestier hastalığı diffuz idiopatik iskelet hiperostozisi (DISH) olarak bilinir. Paravertebral ligamentlerin, kasların enkondral ossifikasyonu ve yaygın spinal osteofitler ile karekterizedir. DISH'in doğal progresyonu tam olarak anlaşılamamıştır.

50 yaşındaki bayan hasta 22 yıldır baş ve boyun ağrıları şikayetleri ile kliniğimize başvurdu. Hastanın 10 yıldır da yutkunma güçlüğü varmış ve 4 yıldır Grave's hastalığı nedeni ile tedavi görüyormuş. Nörolojik muayene bilateral exoftalmus dışında normaldi. DISH tanısı servikal düz grafilerle kondu ve 1977'den bu yana çekilen grafileri ile gelişimi izlendi. 1977'de C5-6 disk mesafesi önünde küçük bir osteoid gelişim vardı. 1985'deki filmlerinde bu oluşum daha genişti ve longitudinal ligament (ALL) içinde birleşik bir ossifikasyon gösterdi. 1991'de C4 ve C6 korpuslarında yeni osteofitler gözlendi. 1999'da C3, C7 ve T1 korpuslarındaki yeni osteofitlerle beraber C4-5-6 daki ALL'de geniş iskelet hipertrofisi vardı. Son grafilerinde lomber ve trosik yeni osteofitler de gözlendi.

Hasta yutma güçlüğünün ilerleyici olmaması nedeni ile operasyonu kabul etmedi. Graves' hastalığı tıbbi tedavi ile düzelmişti. Uzun süreli takibinde nörolojik defisit gözlenmemiştir. Uzun yıllar içinde osteoid formasyonun gelişimi izlenebilmiştir. Bu olgu sunumu küçük bir osteoid formasyonun DISH'e kademeli ilerlemesini göstermektedir. Graves' hastalığı ile birlikte olması, DISH'ın multi sistemik ve immuno-patolojik bir hastalık olabileceğini akla getirmektedir.

Anahtar sözcükler : Forestier hastalığı, Disfaji, Radyolojik progresyon

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INTRODUCTION

Forestier's disease is known as a diffuse idiopathic skeletal hyperostosis (DISH) and is characterised by a bone-forming tendency with prominent radiographic findings, including proliferation of bone throughout the ligaments and tendons of the spine and extremities(1,2). DISH occurs commonly in the thoracic spine, less frequently in the lumbar and cervical spine and commonly in the sixth and seventh decades of life(3). Its prevalence in adults over 40 years of age is 3.8% for men and 2.6% for women(4). Signs and symptoms include stiffness, pain, dysphagia due to direct oesophageal compression and myelopathy related to spinal cord compression associated with ossification of the posterior longitudinal ligament (OPLL)(5).

We present a case with Forestier's disease, which began as a small osteoid formation at the cervical ALL and then became a diffuse skeletal hyperostosis. Follow-up over 22 years allowed an opportunity to observe the insidious growth progression of this disease.

CASE REPORT

This 50-year old female had a 22-year history of neck pain and headache. She had also suffered a difficulty in swallowing for ten years, and had been receiving treatment for Graves' disease (diffuse nodular goitre) for four years. Thyroid scintigraphy and ultrasonography showed a solitary right nodular gland, and her free T4 level was elevated. She is now in remission of Graves' disease, under conventional antithyroid drugs (propylthiouracil) and beta blocking agents. On examination, there was no neurological abnormality, except for bilateral exophtalmos. Her swallowing complaint was progressive and oesophageal barium examination showed a narrowing at the level of C6-7.

The progression of DISH had been monitored with serial plain roentgenograms since 1977. In 1977 she had only neck pain and the cervical spine roentgenogram showed a small osteoid formation in front of the C5-6 intervertebral disc space (ALL) associated with loss of cervical lordosis (Figure 1). In 1985, this osteoid formation in the ALL had grown larger and had come into contact with newly-formed osteophytes in anterior margins of C5 and C6 vertebral bodies. Then all of these showed a larger, combined ossification in ALL, at the level of C5 and C6 (Figure 2).

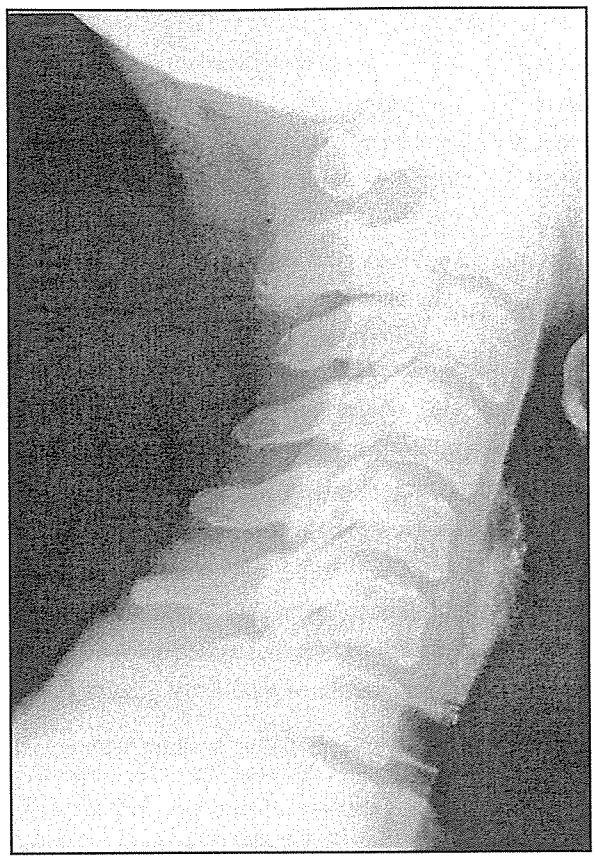

Figure 1. At the age of 28, a small osteoid formation was observed in ALL at the level of C5-6, which was not taken seriously.

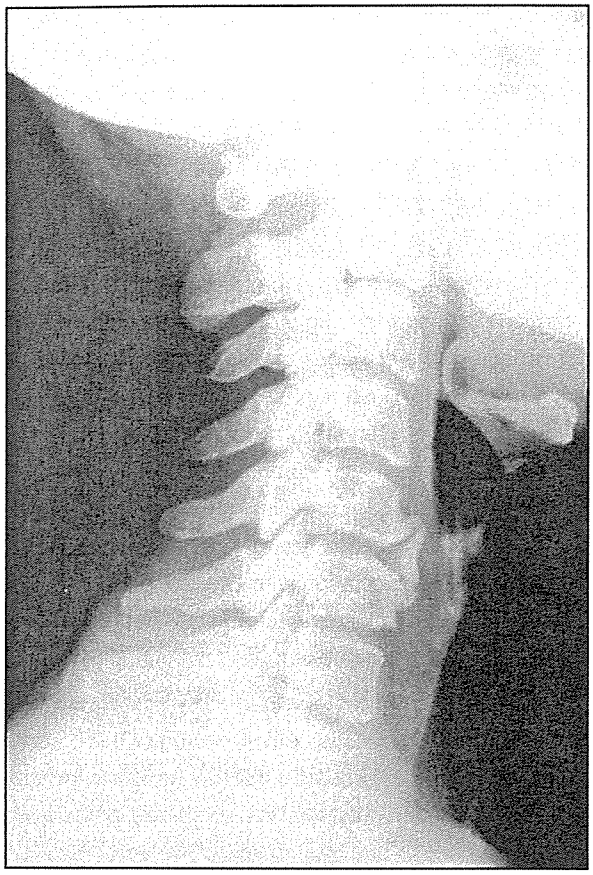

Figure 2. At the age of 36, osteoid formation in the ALL had grown larger and come into contact with newly-formed osteophytes in anterior margins of C5 and C6 vertebral bodies.

n 1991, new osteophytes were again observed to have formed in C4 and C7 that subsequently began growing (Figure 3) and the osteophyte in C4 later combined with the neighbouring osteoid formation at the level of C5-6. In 1999 there was a large skeletal hypertrophy in ALL at C4-5-6 associated with new osteophytes in C3, C7 and T1 bodies (Figure 4). Heights of cervical intervertebral spaces were not changed. At the level of C6-7, skeletal hypertrophies showed a protrusion through the oesophagus. Cervi cal computerised tomography verified that there was no OPLL. There was also no loss of cervical lordosis and no restricted cervical movements. Small new thoracic and lumbar osteophytes were also observed in the latest roentgenograms. In addition to these, bilateral osteoid spurring in ulna at the elbow and in scapula at the shoulder were also observed. All these new osteoid formations led us to consider that abnormal growth progression of bone in our case had started to invade the entire skeletal system. There was no sacroiliitis or HLA B27 in our case.

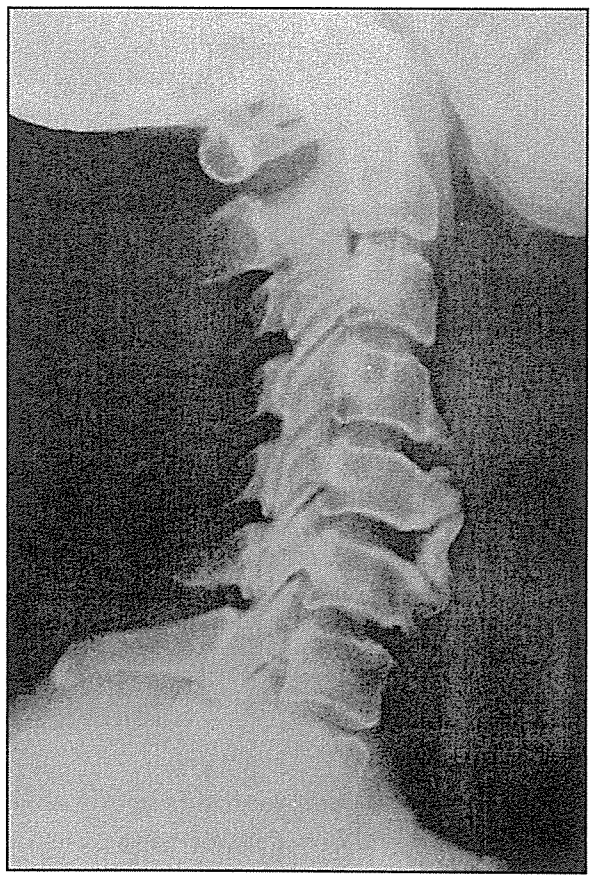

Figure 3. At the age of 42, new osteophytes in C4 and C7 were observed. Old osteoid formation showed enlargement.

The patient was advised of the necessity of an operation to remove cervical osteophytes in order to reduce oesophageal narrowing. Due to her progressive complaint in swallowing, she was operated and cervical osteophytes removed. Now, she has no complaint in swallowing. Further follow-up was planned to observe the progression of oesophageal narrowing.

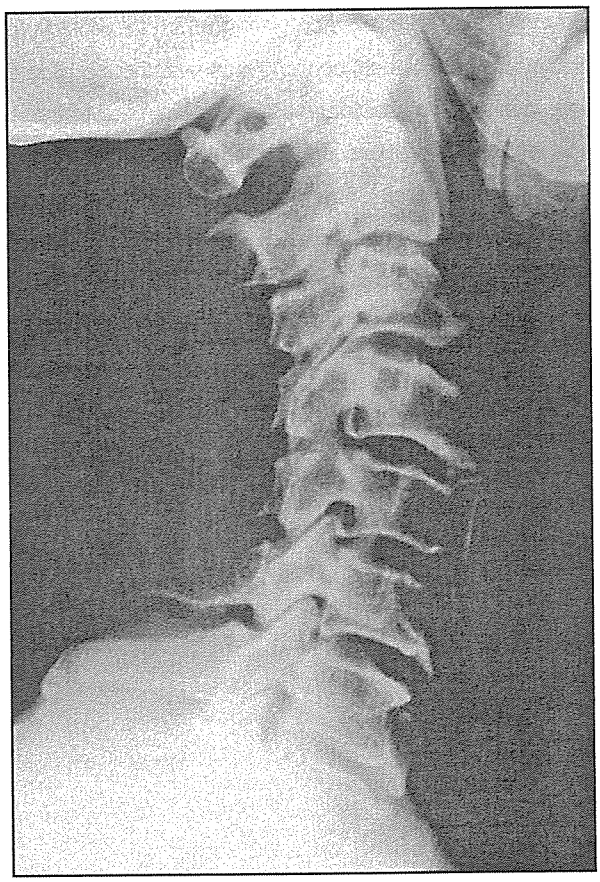

Figure 4. At the age of 50, there was a large skeletal hypertrophy in ALL at C4-5-6 associated with large new osteophytes in C3, C7 and T1 bodies. Note the protruded osteophytes in ALL at the level of C6-7, causing swallowing problems.

DISCUSSION

DISH is a progressive disease and long-term follow-up of the insidious growth of skeletal hypertrophies is not commonly possible. In our case, the osteoid formation began as a small structure 22 years ago and new skeletal hypertrophies then occurred adjacently to this. Some of these new formations combined with the old skeletal hypertrophy. It has been reported that ossification grows in thickness at mobile segments and no growth of ossification is found at immobile segments(6). In our case, growth progression and even remodelling in skeletal hypertrophy continued at the immobile segment. Bone remodelling is necessary to maintain the structural integrity of the skeleton(7). It is our opinion that new osteoid formations may appear in adjacent mobile segments, and old skeletal hypertrophies, even those at immobile segments, continue growing in cases of DISH. Extraspinal manifestations of DISH commonly occur at muscle-attachment sites in bones; for that reason, investigations of whole skeletal system will be necessary in the future(8).

Changing mechanical forces or metabolic and nutritional stress may stimulate abnormal osteophytic formation(7). One of these may have been the etiological triggering factor in our case. DISH has also been considered a multisystemic hormonal or metabolic disorder due to the high incidences, in cases with DISH, of common metabolic conditions such as dyslipidemia, hyperuricemia, increased growth hormone and diabetes mellitus(9,10). Association with Graves' disease in our case may support this consideration, although Graves' disease was a later development.

Ankylosing spondylitis is generally associated with HLA B27 and sacroiliitis, which exclude DISH; some reports, however, present DISH in association with ankylosing spondylitis(11). We excluded ankylosing spondylitis due to she has no sacroiliitis and HLA B27.

She was operated to remove the osteophytes, to reduce oesophageal narrowing. Stridor, aspiration pneumonia, dysphagia secondary to giant cervical osteophyte formation may be the major symptoms in DISH(3,5,12).

This report illustrates gradual radiological progression of a small osteoid formation to DISH. Long-term radiological follow-up gave us an opportunity to increase our understanding of the nature of bone growth in DISH. We conclude that small osteoid formations should be considered to have a potential for growth progression, since DISH has a prevalence in adults over 40 years of age. DISH should be suggested at the differential diagnosis in dysphagia. In cases with DISH, accompanying immunopathological and hormonal diseases should also be investigated.

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