KASHIN-BECK'S DISEASE

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A case of Kashin-Beck's disease is presented. It is an acquired, disabling, polyarthritic, degenerative condition of early onset, sometimes leading to a variable degree of dwarfism.
It occurs endemically in certain Asian areas. Treatment is, if possible, preventive. In the established disease the therapy is that of any other form of secondary osteoarthritis. Reconstructive surgery and removal of loose bodies may be needed in severe cases (11). Early detection, facilitated by careful family history taking, is a prime requisite, especially in non-endemic areas (7).

Keywords : Kashin-Beck ; osteoarthritis ; secondary osteoarthritis.
Mots-clés : Kashin-Beck ; polyarthrite ; polyarthrite secondaire.

CASE HISTORY

A 49-year-old Chinese man was admitted to the Ghent University Hospital because of acute and painful swelling of the right knee. He also reported chronic pain involving both shoulders, elbows, fingers, hips, the contralateral knee and the ankles since his childhood. The pain was mechanical in nature with inflammatory episodes. He had no complaints in the axial region.

From the family history it appeared that two of his brothers, aged 50 and 51, and his 48-year-old sister had similar complaints. His eldest brother, 64 years of age, was symptom free. When the patient was about 3 years old, the family moved from Central China to a province in the Northeast. During the three years they stayed there, the four youngest children caught the disease like many others of their age in that area.

The clinical examination showed a markedly limited mobility of all large joints, except for the axial skeleton. The x-ray examination disclosed multiple deformations of the joint surfaces with secondary arthrotic changes. The 99mTc-MDP total body scan was consistent with these findings.

The patient also had gastroscopic evidence of a bulbar ulcer, for which he was treated with famotidine, 40 mg/d, for a period of 3 months.

INTRODUCTION

A case of Kashin-Beck's disease is presented. A brief description is given of one of the few diseases of the osteoarticular system in which an environmental toxic food substance has been implicated.

Kashin-Beck's disease is a chronic, disabling, symmetrical and generalized osteoarthritis involving peripheral joints and sometimes the spine, unassociated with systemic or visceral manifestations (5, 9).

Careful history taking and a thorough investigation are essential to the diagnosis, especially in nonendemic areas (7).

Biochemical investigations did not reveal any relevant data. The standard rheumatologic parameters, routine urinanalysis and urine sediment were normal.

The patient was treated conservatively, as is customary for secondary uncomplicated osteoarthritis.

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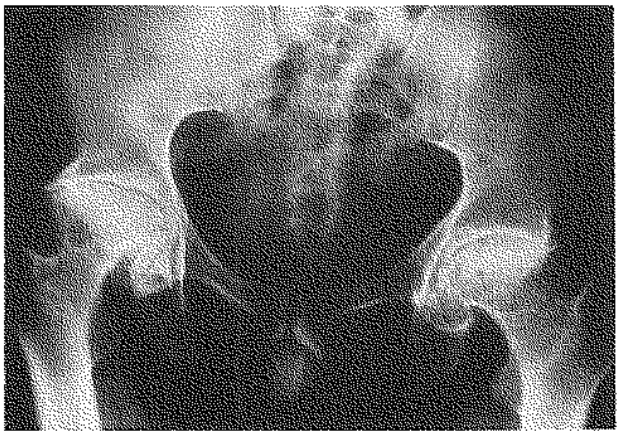

Fig. 1. — Bilateral localisation at the hip.

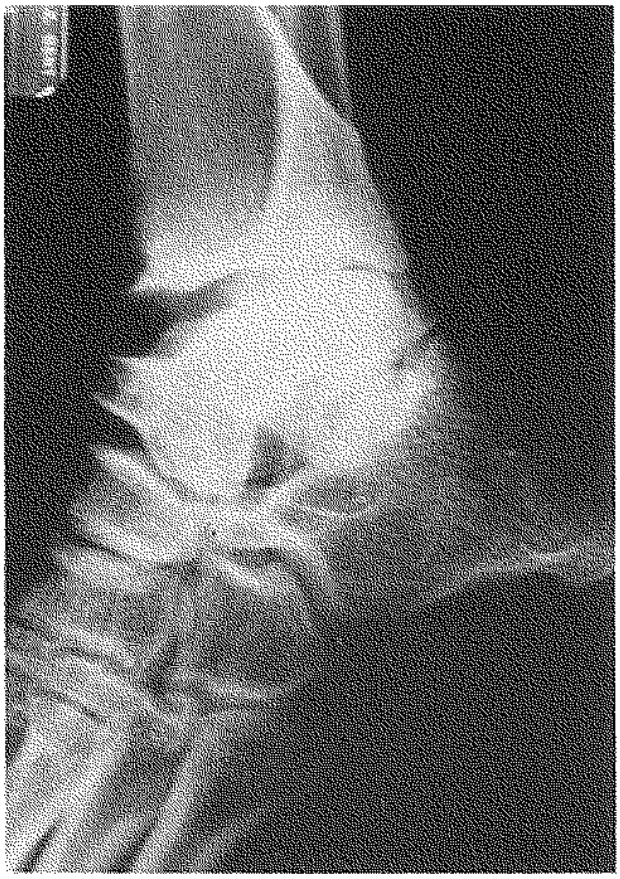

Fig. 2. — Localisation at tibio-talar joint.

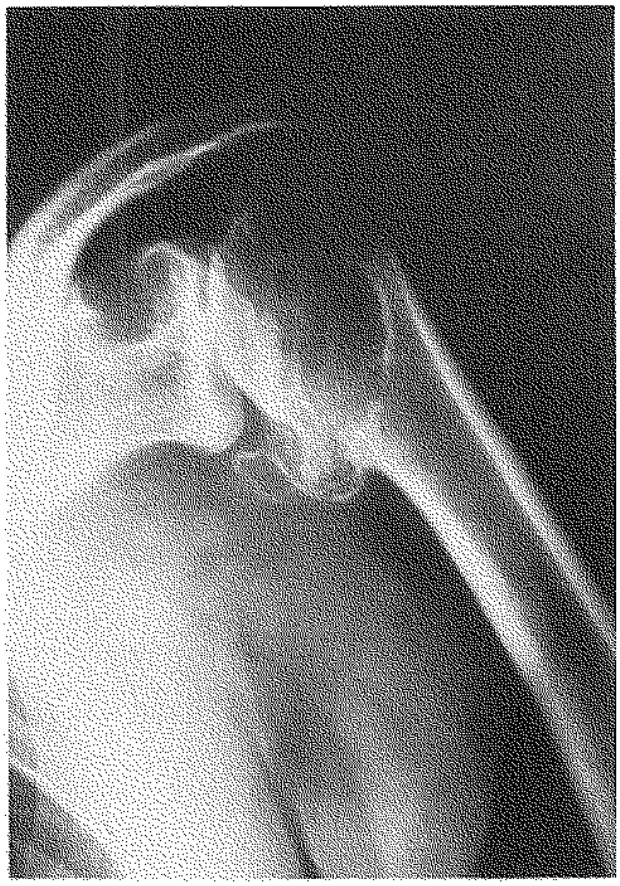

Fig. 3. — Localisation at the shoulder.

DISCUSSION

Kashin-Beck's disease was first described in the late 19th century (4). The condition is endemic in certain areas of northern China, northern Korea and eastern Siberia, and is characterized by episodes of recurrent joint swelling which eventually lead to deformities and accelerate the development of osteoarthritis.

An onset in the first or second decade of life causes osteochondrosis with epiphyseal necrosis, defective growth and the occurrence of loose osteochondral bodies in the joints, which are responsible for the acute attacks or locking of the joints. The patients do not grow to full adult height. The parts of the growing skeleton that are associated with hyaline cartilage are subject to pathologic changes. Conversely, when the onset

occurs in adult life, the changes seem to be confined to the articular cartilage. The necrosis is of the coagulative type with a characteristic distribution starting at the base of the cartilage and extending to the surface. It may involve the whole thickness of the cartilage (2, 4). These disturbances cannot be found in fetuses in the endemic areas, and thus they are not congenital (11).

In those areas where cereals are frequently infested by *Fusarium sporotrichiella*, the climatic conditions are special : very cold, dry winters and wet, hot summers. By the use of an experimental animal model scientists found that animals fed with these cereals developed changes in the articular cartilages, epiphyses, and metaphyses similar to those found in Kashin-Beck's disease (19, 20, 22). The pathogenetic factors relate both to grain and to water in the diet in endemic areas (11).

A second hypothesis concerns a lack of sodium selenite. Some scientists found no evidence that chondrocytes have idiosyncratic requirements for selenite nor did they find arguments to support the hypothesis that selenite deficiency is a major etiologic factor in the disease (11, 16, 18). There is evidence that hyposelenosis could be responsible for a common childhood cardiomyopathy Keshan disease, in Shaonxi province (1). Tibial dyschondroplasia in chickens can be caused experimentally by admission of a water-soluble mycotoxin of *Fusarium roseum* "graminearum" (6, 15). The mycotoxin, fusarochromanone, is cytotoxic but chondrocytes are not more sensitive than other cell types (11). Hence, it does not seem to be a major etiologic factor in Kashin-Beck disease (17).

An increase in activity of lactic dehydrogenase, aspartate transaminase, alanine transaminase, alkaline phosphatase, and creatinine phosphokinase in the serum, an increase in creatine and a decrease in creatinine in the urine, may be present, but not as a rule. The relation between the changes in activity of the serum enzymes and the pathological alterations requires further study (4, 14).

Kashin-Beck's disease must be distinguished from other forms of secondary osteoarthritis (table I). In young people it should be distinguished from the several types of juvenile chronic arthritis (7).

Table I. — Secondary arthrosis

Articular deformity
Mucopolysaccharidosis
Spondyloepiphyseal dysplasia
Multiple epiphyseal dysplasia
Congenital dysplasia of the hip
Protrusio acetabuli
Slipped femoral capital epiphysis
Malunited fracture
Deformity due to metabolic bone disease
Meniscectomy
Aseptic bone necrosis
Idiopathic
Corticoid therapy
Thiemann's disease
Occupational
Hemoglobinopathies
Gaucher's disease
Mseleni's disease (3, 8, 11, 12, 13)
Articular inflammation
Articular abuse
Occupational
Neuropathic
Joint laxity
Long-leg arthropathy
Metabolic disorders
Hemophilias

In its early phases, the disease is reversible by leaving the endemic area (21). This proves the importance of early detection (7). Reconstructive surgery and removal of loose bodies may be needed in severe cases (11).

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SAMENVATTING

J. MOERMAN, D. UYTTENDAELE, W. VAN DEN BROECKE en H. CLAESSENS. Ziekte van Kashin-Beck.

Naar aanleiding van een klinisch geval wordt een beschrijving gegeven van de enige ziekte van het osteoarticulair stelsel waarin een nutritionele toxische factor een rol zou spelen. De ziekte van Kashin-Beck is een chronische, degeneratieve osteoarthrose die reeds vroeg het jonge groeiende individu kan aantasten. De ziekte is endemisch in bepaalde gebieden van Azië. Preventie is de beste behandeling, behoudens complicaties, conservatief zoals bij elke andere vorm van secondaire osteoarthrose. Een vroegtijdige diagnose is belangrijk en wordt vergemakkelijkt door een diepgaande familiale anamnese, vooral in de niet-endemische gebieden.

RÉSUMÉ

J. MOERMAN, D. UYTTENDAELE, W. VAN DEN BROECKE et H. CLAESSENS. Maladie de Kashin-Beck.

Les auteurs présentent un cas de maladie de Kashin-Beck. Celle-ci est acquise pendant la jeunesse et mène à une affection dégénérative, polyarticulaire et invalidante. Elle est endémique dans certaines régions de l'Asie. Le meilleur traitement reste la prévention. Quand la maladie est installée, le traitement est celui de toute forme d'ostéoarthrite secondaire. Le diagnostic précoce est primordial. Il est facilité par une anamnèse familiale approfondie, spécialement dans les régions où la maladie n'est pas endémique.