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Diagnostic, clinical and radiological characteristics of Kashin-Beck disease in Shaanxi Province, PR China

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Abstract Twenty clinical symptoms and four radiological signs of different joints in patients with Kashin-Beck disease (KBD) were studied in 2560 subjects from endemic and non-endemic areas of the Shaanxi Province in China. It is suggested to classify the symptoms into five groups representing different manifestations of the disease. The association of some of the symptoms appears to provide significant criteria for use in the diagnosis of KBD.

Résumé Dans la maladie de Kashin Beck, vingt symptômes cliniques et quatre signes radiologiques au niveau de différentes articulations ont été étudiés chez 2560 personnes des régions endémiques et non-endémiques de la province du Shaanxi en Chine. Il est suggéré de classer les symptômes dans cinq manifestations différentes. L'association de quelques symptômes paraît être des critères diagnostic pour la maladie de KB.

Introduction

Kashin-Beck disease (KBD) is a chronic endemic osteoarthropathy in which the epiphyseal growth plate and the articular cartilage are the most commonly affected sites. Focal chondronecrosis in the deep zone and impaired endochondral ossification in this cartilage results in a secondary chronic deforming osteoarthrosis and impaired skeletal development. As the histological diagnosis is particularly difficult, clinical and radiological examinations have provided the best means of identifying KBD until now. Little is known about its early stages which occur before the appearance of the characteristic destruction of the joint. Previous reports of investigations have summarised the main findings and thus allowed the development of criteria for diagnosing KBD [3]. This paper summarises the clinical and radiological findings which occur in joints damaged by KBD.

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Materials and methods

From among 2560 subjects in Shaanxi Province we studied 1992 from eight different counties (Bin, Yongshou, Linyou, Qian, Yao, Yulin, Huanglin and Liquan) that have either a mild (less than 30%), middle (31–50%) or high (more than 50%) prevalence of KBD. In addition, 560 subjects came from one non-endemic county (Fhang-an) close to Xi'an city. The patients were divided into seven groups and have been compared according to where they lived, their age and their diagnosis (Table 1). The diagnosis of KBD was established on the basis of criteria presented previously in China [3] and this was classified in four stages (Table 2). We used the physical examination protocol described previously [1, 2] and we recorded 20 clinical signs and symptoms. An antero-posterior radiograph of the right hand of each patient was taken, and if the patient was then classified into the first, second or third stage an antero-posterior and a lateral radiograph of the elbow, knee, ankle, hip and spine were added. Abnormal radiological signs in the metaphysis, epiphysis and the end of the phalanges of the hand and in the carpal bones were noted. Data entry and statistical analyses were carried out with the SPSS (version 8.0) software package. The chi-square test was used to analyse the different variables.

Table 1 Distribution of the population between the different groups following place of residence, age and diagnosis of the patients in Shaanxi Province, PR China

	Children (1–15 years)	Adults (16–20 years)	Total
KBD area			
Healthy	902	223	1125
Early stage	334	0	334
1st–3rd stage	285	248	533
Non-KBD area			
Healthy	388	180	568
Total	1909	651	2560

Table 2 Kashin-Beck disease. Classification in four stages [3]

Stage	Clinical symptoms	X-ray signs
Early stage	Flexion of the terminal part of the fingers or crooked fingers and arthritic pain in knee and ankle joints without enlarged finger joints	Metaphyseal lesions in phalanges
First stage	Enlarged finger joints and clinical symptoms of early stage	Metaphyseal lesions in phalanges
Second stage	Shortened fingers and clinical symptoms of first stage	Metaphyseal lesions in phalanges
Third stage	Retarded growth or dwarfism and clinical symptoms of second stage	Metaphyseal lesions in phalanges

Table 3 Prevalence of the clinical symptoms and X-ray alterations in children and adults in non-KBD and KBD areas in eight counties of Shaanxi Province, PR China

Clinical symptoms and X-ray signs	Children in non-KBD		Children in KBD areas						Adults in non-KBD		Adults in KBD areas			
	Healthy		Healthy		Early stage		KBD I–III		Healthy		Healthy		KBD I–III	
	388	%	902	%	334	%	285	%	180	%	223	%	248	%
Arthritic pain	0	–	3	0.3	12	3.6	45	15.8	0	–	10	4.5	176	71
Morning stiffness	7	1.8	5	0.5	1	0.3	1	0.3	5	2.8	10	4.5	139	56
Enlarged finger joints	0	–	0	–	0	–	126	44.2	0	–	0	–	164	66.1
Deformed finger joints	0	–	0	–	0	–	26	9.1	0	–	0	–	79	31.8
Short humerus	0	–	0	–	0	–	3	1	0	–	0	–	2	0.8
Short fingers	0	–	0	–	0	–	8	2.8	0	–	0	–	97	39.1
Short toes	0	–	0	–	0	–	10	3.5	0	–	0	–	63	25
Limited motion														
Finger flexion	17	4.4	141	15.6	110	32.9	157	55.1	0	–	2	0.9	78	31.4
Wrist	0	–	0	–	0	–	20	7	0	–	0	–	148	59.7
Prosupination	0	–	0	–	0	–	2	0.7	0	–	0	–	49	19.8
Elbows	0	–	0	–	0	–	32	11.2	0	–	0	–	158	63.7
Shoulders	0	–	0	–	0	–	1	0.3	0	–	0	–	3	1.2
Ankles	2	0.5	0	–	0	–	98	34.4	3	1.7	0	–	172	69.3
Knees	0	–	0	–	0	–	8	2.8	0	–	0	–	31	12.5
Hips	0	–	0	–	0	–	0	0	0	–	0	–	5	2
Squat down	2	0.5	0	–	0	–	98	34.4	3	1.7	0	–	174	70.2
Flat feet	5	1.3	0	–	0	–	10	3.5	4	2.2	0	–	13	5.2
Deformed knees	0	–	0	–	0	–	4	1.4	0	–	0	–	5	2
Scoliosis	0	–	0	–	0	–	0	0	0	–	0	–	3	1.2
Dwarfism	0	–	0	–	0	–	4	1.4	0	–	0	–	6	2.4
X-rays alterations														
Metaphyseal	13	3.3	0	–	255	76.3	245	86	0	–	0	–	0	–
Epiphyseal	0	–	0	–	35	10.5	49	17.2	0	–	0	–	0	–
Bone end	0	–	0	–	36	10.8	156	54.7	0	–	0	–	239	96.4
Carpal	0	–	0	–	11	3.3	26	9.1	0	–	0	–	41	16.5

Results

There were 1,307 males (51%) and 1,253 females (49%). The majority of the patients were young, and the age distribution is shown in Fig. 1. The incidence of clinical symptoms and radiological signs in the different groups is summarised in Table 3, and these are mentioned below.

Arthritic pain (without redness, swelling or heat) in ankles and knee joints was the most common symptom in the KBD patients (71% adults, 16% children) and was significantly higher than in the other groups ($P<0.001$).

Morning stiffness of joints mainly affected the KBD adults (56%, $P<0.001$). Fixed flexion of the terminal

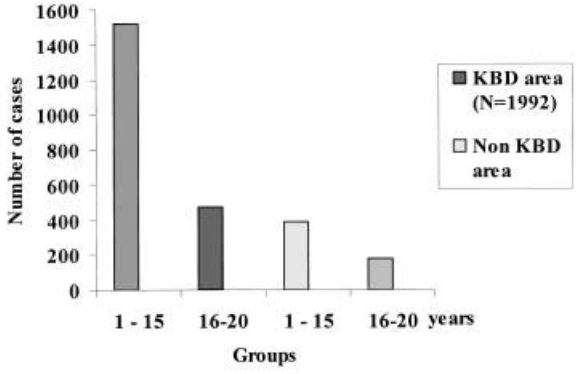

Fig. 1 Age distribution of the population studied (n=2560) in endemic and non-endemic areas of Shaanxi Province, PR China

Fig. 2 Radiographic findings in the right hand of a 13-year-old boy with KBD

Table 4 Description of the different lesions in phalanges and carpal bones in cases of KBD children. Shaanxi Province, PR China

Location	Description	%
Metaphysis	Blurred and interrupted	39.8
	Thick marginal sclerosis	36.2
	Small defect (<2 mm)	28.5
	Large defect (>2 mm)	4.8
	Irregular marginal sclerosis	4
Epiphysis	Cone shaped	12.4
	Premature closure of epiphyseal line	7.3
	Thinner	2.6
	Fragmented	2.9
	Irregular marginal sclerosis	1.3
	Absence	0.7
End of phalanges	Flattening	19.1
	Marginal spur	11.5
	Small defect with sclerosis	11.7
	Irregular	9.2
	Blurred and interrupted	7.8
	Marginal defect	5.2
	Enlarged	4.9
	Marginal sclerosis	1.9
	Pouch-like changes	1.6
Carpal joints and bones	Absence	7.1
	Irregularity with sclerosis	6.6
	Marginal interruption	5.7
	Narrowed joint space	1.9
	Impaired development	1.4
	Marginal blurred bones	0.6

joint of the fingers and deformed fingers appeared more frequently in the group of advanced KBD children (first to third stages: 55%, $P<0.001$). The incidence of deformed fingers was higher in the group of healthy children living in an endemic area than in the group living in non-endemic areas ($P<0.01$). Limited motion when squatting and in the ankle joints was more common in both the KBD children and adults ($P<0.001$). Neither flat feet nor limited motion of the hip joints and spine were observed in the groups of children. The incidence of clinical symptoms and of radiological signs (except in the growing area) increased with age and were more common in the adult groups ($P<0.01$). Radiological changes in the metaphyseal area were the most commonly observed lesions in KBD children in the early stage (76%) and also in the more advanced (first, second and third stage) group of children (86%). Abnormal radiological signs appeared in the metaphyseal area in 70% of patients. The distal parts of the middle and proximal phalanges of the index and ring fingers showed radiological changes in 51%. A detailed description of radiological changes in the phalanges and carpus (Fig. 2) is summarised in Table 4.

Fig. 3 Clinical findings in a 45-year-old man with KBD: enlarged interphalangeal joints, shortened fingers and limited motion of the elbows

Discussion

Twenty clinical symptoms and four radiological changes were used to characterise effects of KBD on the joints. These can be grouped into five different manifestations of the disease: flexion of the terminal joint of the fingers or deformed fingers; arthritic pain in knee and ankle joints; enlargement of small and middle-sized joints (Fig. 3); limited motion in the affected joints (wrist, elbow, ankle, knee); and deformity and retarded growth (shortening of fingers and humerus, shortened stature).

Joint damage in patients with KBD shows an increasing proximo-distal gradient. The distal joints (fingers, wrists, ankles) are more often affected than the proximal ones (hips and shoulders). The number of symptoms is greater in adults. The irregular and impaired development of bones is the consequence of the premature closure of the epiphyseal plate often leading to short fingers, femur and humerus.

It was noted that abnormal radiological changes at the extremities of the finger phalanges were present in a high percentage of patients (96%) in the KBD adult group but that this did not occur in the healthy adult group. It is suggested that this might be a specific criterion for diagnosing KBD. In early stage diagnosis abnormal radiological changes in the metaphysis of finger phalanges must be considered with any clinical symptoms.

In summary, the four most significant criteria for the diagnosis of KBD are:

1. Radiological changes in the distal end of the bones of the middle and proximal phalanges of the index and ring fingers.
2. Focal or irregular premature closure of the epiphysis.
3. Limited motion and enlargement of peripheral joints, deformities and dwarfism.
4. Involvement of multiple joints by non-inflammatory lesions.

These manifestations in an individual who has resided in an endemic area for at least 6 months make a diagnosis of KBD highly probable.

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