PATTERN OF JOINTS INVOLVEMENT IN KASHIN-BECK DISEASE: A LOCAL OSTEOCHONDROPATHY IN CHINA

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Background: Kashin-Beck Disease (KBD) is an endemic osteochondropathy. The disease starts in childhood and attacks the growth of joint cartilage, the joints become deformed and painful, and the worst forms result in dwarfism. The most frequent joints involved are the finger, wrist, ankles, knees and elbows. In this study the pattern of joints involved in KBD was studied. **Methods:** A total of 368 patients aged above 13 from 6 villages in endemic areas of the Shaanxi province, located in the north-west of China, were selected through multistage stratified sampling. The patients were diagnosed based on the clinical criteria for diagnosis of KBD. The patients were administered with a battery of questionnaires along with clinical examination for joint involvement. **Results:** The patients presented both as oligoarticular as well as polyarticular pattern. Smaller joints were more frequently involved with a proximal-distal pattern. The number of joints involved increased with the severity of the disease. **Conclusion:** Proximal smaller joints are more frequently involved in the disease. The involvement of bigger joints takes place in later stages of disease, i.e., in the second and third degree. The pattern of joint involvement shows some correlation with Rheumatoid Arthritis which needs further investigation and comparative studies.

Keywords: Kashin-Beck Disease, Endemic, Osteochondropathy

INTRODUCTION

Kashin-Beck disease (KBD) is a chronic, endemic, degenerative osteoarthropathy with severe skeletal deformation and dwarfism.[1] Geographically KBD has a typical endemic distribution in Eastern Siberia of Russia, the diagonal broad belt extending from the north-eastern to the south-western China and North Korea.[2] The etiology of the disease is still not established. Three major environmental hypotheses including endemic selenium deficiency, cereal contamination by mycotoxin-producing fungi and high humic acid levels in drinking water have been proposed in the past 150 years.[3–6]

The climate in KBD endemic areas usually includes a long period of frost and remarkable differences between the daily minimum and maximum temperatures. In China, there were 0.74 millions patients with KBD and 10.342 millions of people at risk in 354 counties of 15 provinces or autonomous regions in 2006.[7] The disease effects the villages in small groups having proximity to each other. In certain well-defined areas, such as some valleys of the Tibetan plateau and the Shaanxi province, a prevalence as high as 80% of the population has been reported.[8–10] Mostly people related to agriculture and those consuming locally produced grain as their staple diet are affected. Kashin-Beck disease is a disabling osteoarticular disorder locally called as big bone disease. Kashin-Beck disease usually starts in children between the ages of 5 and 13 years and usually involves the metaphysis in growing bone. It is equally prevalent in males and females.[3]

Clinically, the disease manifests itself in arthritic pain, morning stiffness, enlarged and shortened fingers and deformed, enlarged joints with limited motion in the extremities. In seriously affected patients, this disease leads to shortened stature or dwarfism and disability in their daily life.[11]

There is no cure, and no clear preventive measures exist. The most frequently affected joints are ankles, knees, wrists, and elbows leading to atrophied muscles, making manual farming work difficult and painful causing greater economic and social difficulties for rural population.[12–13]

Kashin-Beck disease severity can be defined by the deterioration of joints: the early stage typically involves flexion of the terminal joints of fingers or deformed fingers; the first degree includes enlargement of small and middle-sized joints, with arthritic pain in the knee and ankle joints; the second degree includes those symptoms noted for the first degree plus shortening of the fingers; and the 3rd degree includes degree one and two symptoms plus dwarfism.[14]

Joint damage in patients with KBD shows an increasing proximal-distal gradient. The distal joints (fingers, wrists, ankles) are more often affected than the proximal ones (hips and shoulders), in 66.1% of the KBD patients the finger joints are enlarged while 39.1% of the patients have short fingers. As compared to the fingers a lesser percentage of patients, i.e., 25% show short toes. The wrist joint is involved in 59.7% of the patients while elbow in 63.7%, shoulder in 1.2%, ankle 69.3%, knee in 12.5%, and hip joint in 2% of the patients. Studies reveal that 2.4% of the patients were dwarf.[11,14,15]

The current study was done to look in to the pattern of joint involvement in KBD patients and its association with the severity of the disease.

MATERIAL AND METHODS

The data for this paper was taken from a cross sectional study conducted in the KBD endemic areas of Shaanxi province in China. Located in the north-west of the country Shaanxi province is one of the most endemic provinces for KBD having a population of 37.2 million with 63% population living in the rural area.[16] The study was designed to measure health related quality of life and disease burden of KBD. The current paper, however, focus on the pattern of joints involvement in KBD only. Target population was the population aged above 13 years living in endemic areas in Shaanxi province. Randomly selected participants were screened for KBD based on the 'diagnostic criteria of Kashin-Beck disease' in China.[11–14] The clinical criteria included a positive living history in a KBD area and typical clinical symptoms in the extremities which include arthritic pain, flexion of distal fingers, enlargement of finger joints, shortening of fingers, wrist joint dysfunction, elbow joint dysfunction, knee joint dysfunction, ankle joint dysfunction, difficulty squatting, short stature, sitting height, hand length and hand width. Those having any other osteoarthritic disease like Rheumatoid Arthritis or Osteoarthritis were excluded from the study.

A total of 684 participants were recruited in the study. Multistage stratified Random sampling was employed for the study. The sample unit was a village in Shaanxi province. In the first stage two counties from endemic areas of Shaanxi province were randomly selected and then the villages of each selected county were divided in to 3 strata based on the prevalence of KBD: low (<10%), middle (10–20%) and high prevalence (>20%) villages. One village was selected from each strata and all population aged above 13 in the selected villages were invited to participate.

A general questionnaire and a check list were used to ascertain the joints involved. The check list was prepared after going through the available pattern of joints involvement and was finalized after discussion with the disease experts.

Data was analysed using SPSS version 16. The basic characteristics of the KBD patients were described as frequencies, percentages and means depending on the type of variable. The frequency of joints involvements were measured for each joint and then the pattern of each joint involved were assessed with the severity of the disease. The difference in joint involvement in different degrees of KBD patents were compared using chi-square test with $p \leq 0.01$.

RESULTS

A total of 684 individuals were recruited in the study from six villages of the two counties of Shaanxi province by multistage stratified sampling. There were 347 from Linyou and 337 from Yongshou counties. Of the total, 368 patients were diagnosed as KBD cases with 225 patients having first degree KBD while 128 had second, and 15 with third degree KBD.

Out of the total number, 48% patients were male and 51.4% were female. The patients had a mean age of 55.9±10.08 years. Patients who never went to school were 39.9%. Out of the rest, majority have primary education, some had middle and only a few (6%) had ever attended the high school. Most of the patients (96.7%) were farmers, and living in properly built houses (89.7%). Patients having their water supply through wells were 48.1%, and rain water (29.3%) while others utilised the tap water and river water. The mean income of the KBD patents was 6331 RMB per year. (Table-1).

In 293 (79.6%) of the KBD patients multiple joints were involved while the rest 75 (20.4%) had only one joint involved. Fifty-three (14.4%) KBD patients reported to have 2 joints involved while 72 (19.6%), 65 (17.7%), 63 (17.1%), 26 (7.1%), 8 (2.2%) and 6 (1.6%) patients reported to have 3, 4, 5, 6, 7, and 8 joints involved respectively. Among the patients who reported the involvement of only a single joint ,finger was the most frequently involved joint 36 (48%) followed by Knee 19 (25.3%) and wrist joint 15 (20%). A small proportion of patients reported the involvement of ankle 4 (5.3%) and shoulder joint 1 (1.3%) while none reported the isolated involvement of hip, elbow or spine.

Figure-1 shows the percentages of different joints involved. The finger joints were involved in 293 (79.6%) of the KBD patients while wrist in190 (51.6%), elbow in 171 (46.5%), shoulder in 57 (15.5%), spine in 29 (7.9%), hip in 30 (8.2%), knee in 221 (60.1%) and ankle in 242 (65.8%) of the KBD patients

Table-2 shows involvement of joints in different stages of KBD. In case of finger joints the frequency of patients involved was 74.7%, 87.5% and 86.7% in first, second and third degree respectively. The 44.4% of first degree, 60.2% of the second degree and 86.7% of the third degree KBD patients had their wrist joint involved. In case of elbow joint the frequency of joint involvement was 40.9%, 52.3% and 80% in the 3 degrees of the disease in order of severity. Only a fewer patients, i.e., 12.9% and 15.6% in 1st and 2nd degree of KBD had their shoulder joints involved but it increased with severity, and in 3rd degree patients the shoulder joint was involved in 53.3% of the patients.

The involvement of spine was 7.1%, 8.6% and 13.3% in the 3 degrees of KBD in order of severity. The hip joint was involved in 6.2% of 1st

degree, 6.2% of the 2nd and 53.3% of the 3rd degree KBD patients. Knee joint was involved in 57%, 64.1% and 86.7% respectively in 1st, 2nd, and 3rd degree patients. The ankle joint involvement didn't vary much across the 1st, 2nd and 3rd degree, i.e., 65.8%, 64.8% and 73.3% respectively.

Table-1: Characteristics of KBD patients

Characteristic		No. (%)
Gender	Male	179 (48.6)
	Female	189 (51.4)
Education level	No schooling	147 (39.9)
	Primary	118 (32.1)
	Middle	81 (22.0)
	High School	22 (6.0)
	University	0 (0)
Profession	Farmer	356 (96.7)
	Factory Worker	3 (0.8)
	Business	2 (0.5)
	Technician	6 (1.6)
	Others	1 (0.3)
Marital Status	Single	9 (2.4)
	Widow	43 (11.7)
	Married	316 (85.9)
Housing	Cave	36 (9.8)
	House	33027 (89.7)
	Flat	2 (0.5)
Water resources	Tap Water	48 (13)
	Well	177 (48.1)
	Rain Water	108 (29.3)
	River Water	2 (0.5)
	Other	33 (9)
Income (Mean)	6331 RMB/year	

Table-2: Severity of KBD and Joints involvement

Joint	1st degree No. (%) n=225	2nd degree No. (%) n=128	3rd degree No. (%) n=15	*p**
Finger	168 (74.7)	112 (87.5)	13 (86.7)	0.013
Wrist	100 (44.4)	77 (60.2)	13 (86.7)	0.000
Elbow	92 (40.9)	67 (52.3)	12 (80.0)	0.003
Shoulder	29 (12.9)	20 (15.6)	8 (53.3)	0.000
Spine	16 (7.1)	11 (8.6)	2 (13.3)	0.642
Hip	14 (6.2)	8 (6.2)	8 (53.3)	0.000
Knee	126 (57.0)	82 (64.1)	13 (86.7)	0.033
Ankle	148 (65.8)	83 (64.8)	11 (73.3)	0.807

*Asymp. Sig. (2-sided) Pearson Chi-Square test

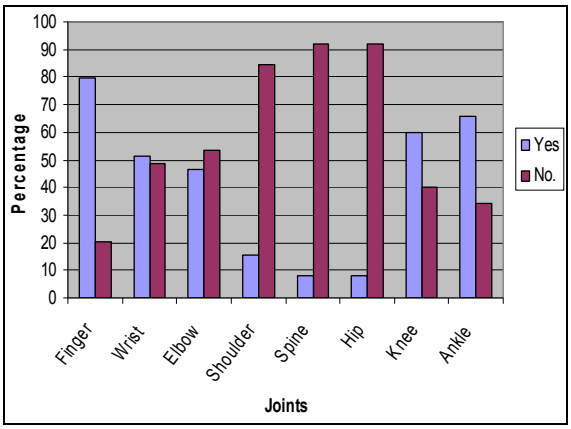

Figure-1: Frequency of Joints Involvement in KBD

Figure-2: 3rd degree KBD patient

DISCUSSION

Kashin-Beck disease is an osteoarthropathy prevalent in rural china. The population in the endemic areas for KBD is largely poor having low socioeconomic indicators.[9] Our study population followed the same trend having low level of education, low income and water supply sources.

It is important to ascertain the pattern of joints involvement in KBD patients so that better preventive and rehabilitative measures can be adopted to overcome the sufferings of the patient. KBD presents itself either as an oligoarticular or a polyarticular disease and the joint involvement usually follows proximal-distal pattern. The smaller joints are more commonly involved then the larger joints. The first joint to involve is usually the finger joints followed by wrist, ankle and knee.[11–14] the same pattern was seen in our study, finger joints showed the most frequent involvement followed by ankle, wrist and elbow in the few of the patients have their larger joints involved, i.e., hip, spine and knee as shown figure-1.Majority of the patients79.6% have their multiple joints involved. Among the polyarticular KBD patients 19.6% of the patients have their 3 joints involved which is the most frequent among this category. Those patients who have only one joint involved, among them the finger joint involvement is most common followed by knee and wrist joint.

With the increase in severity of the disease the number of joints involvement of increased. Although all the joints showed an increase involvement with increase in severity but this is significant only in

case of wrist, shoulder and hip joints. The possible explanation for this is the increase number of patients in the 3rd degree having the bigger joints involved while the smaller joints are involved from the beginning of the disease so equally prevalent in all the degrees of KBD.

If we look at the pattern of joint involvement in other osteo-articular diseases like RA we found that the involvement of metacarpophalangeal joints is 96%, wrist is 86%, elbow 80%, shoulder 58%, spine 46%, ankle 74%, knee 80% and hip 22%.[17] The involvement of the joints in KBD patients in our study correlates with that in RA. It may suggest some common phenomena which paves way for future investigations.

CONCLUSIONS

Kashin-Beck disease is a local osteochondropathy endemic in rural areas of the shaanxi province. It mostly effected the poorer communities. The proximal smaller joints were more frequently involved in the disease. The involvement of bigger joints takes place in later stages of disease, i.e., in the second and third degree. The pattern of joint involvement showed correlation with RA which needs further investigation and comparative studies.

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