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Kashin-Beck disease involving the ankles

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We present a case of Kashin-Beck Disease primarily affecting the ankles. Our findings are consistent with other reports of the symptoms, incidence, and progression of these diseases. At this time, our patient has chosen conservative treatment, but is considering eventual osteoarticular allografts. According to the available literature, surgical treatment is the only option that will not only alleviate her symptoms but halt her joint degeneration. Additionally, osteoarticular allografts have been shown to be an effective treatment and are able to be converted to an ankle arthrodesis if necessary.

Case report

This 41-year-old female had a sudden onset of bilateral ankle pain in 1993, after playing basketball. She was unable to walk without pain for a week, and sought medical care. She said that radiographs at that time showed "small fractures" and "dead cartilage" in her ankles. Her pain improved gradually over the next six months with conservative care, but then began recurring more and more frequently, interfering with her previously active lifestyle of running and playing tennis. She had no history of ankle trauma.

Over the same time period, she also developed pain, stiffness and swelling of her fingers. This was considered to represent seronegative rheumatoid arthritis. These symptoms have been well-controlled with a low carbohydrate, no sugar diet. She had not taken steroids for this.

The patient was born in Northeast China, and reported that a number of family members and friends from Northeast China have had similar symptoms. She was concerned that she and they have Kashin-Beck disease.

Her ankle pain became progressively worse over the next 12 years, and in 2005 she sought podiatric care. Radiographs revealed osteochondral defects of both talar domes (Fig. 1).

Magnetic resonance imaging (MRI) of both ankles showed osteochondral irregularity and collapse in both talar domes (Fig. 2).

Computed tomography (CT) of both ankles was then performed to assist with surgical planning, and also showed osteochondral irregularity and collapse in both talar domes (Fig. 3).

She was then seen by an orthopedic foot and ankle specialist, who discussed several treatment options, including talar-dome drilling and bone grafting, osteoarticular allograft, total ankle arthroplasty, and ankle arthrodesis. At present, our patient is continuing conservative care, but is considering eventual osteoarticular allografts.

Discussion

Kashin-Beck Disease (KBD), also known as "big bone disease" in China, is a chronic endemic osteochondropathic condition. It is found primarily in agricultural regions of eastern Siberia, northern Korea, and the central regions of China (1, 2) (Fig. 4).

The disease was named after Dr. Nicolai Ivanowich Kashin and Dr. Evgeny Vladimirovich Beck, the Russian military physicians who discovered and studied it beginning in 1849 (3, 4). An estimated 2 to 3 million people in China are believed to be affected by KBD (5). However, due to prevention measures instituted by the KBD Foundation, the incidence of new KBD cases is decreasing.

Affected individuals present with bilateral joint pain due to joint destruction affecting the ankles, knees, wrists, and elbows (5, 6). KBD joint degeneration is generally accepted to be due to chondrocyte necrosis. Chondrocytes facilitate

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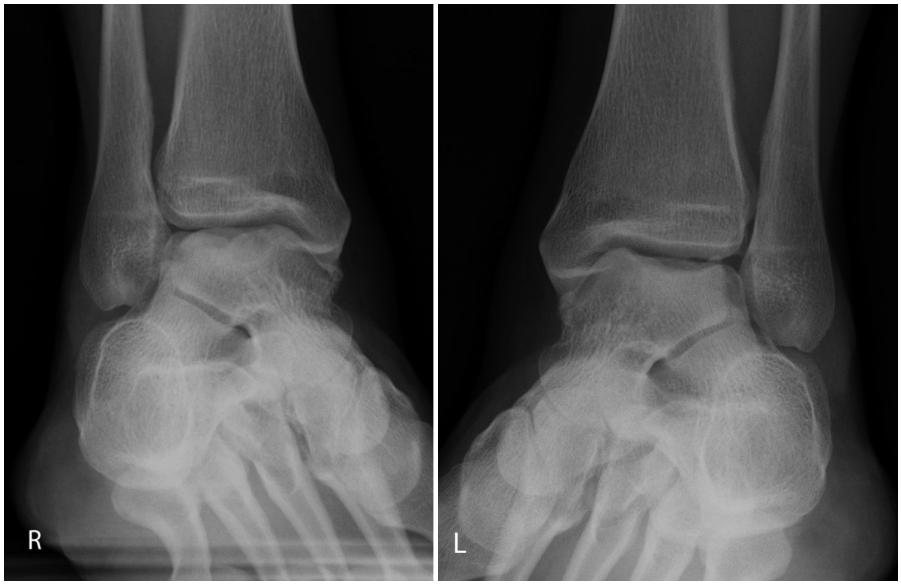

Figure 1. 41-year-old female with Kashin-Beck disease. Radiographic mortise views of right and left ankles show irregularity and flattening of both talar domes, consistent with subchondral collapse.

cartilage repair by producing and secreting collagens, proteoglycans, and enzymes. Without viable chondrocytes, cartilage is not maintained, and this leads to inadequate mechanical support causing joint pain.

Although much is known about pathophysiologic nature of KBD, etiological factors are not clear (7). The disease is generally thought to be linked to a variety of factors: nutritional, environmental, immunological, genetic, and infectious (5). The three leading environmental hypotheses proposed are endemic soil selenium deficiency in affected regions, cereal grain contamination by mycotoxin-producing fungi, and high humic acid levels in drinking water (7).

Based on a 2001 study conducted on 2,560 subjects in China's Shaanxi Province, the distribution between males and females with KBD is about equal, 51% and 49% respectively (8). The study distinguished between various stages of KBD in children (1-15 years) and adults (16-20 years) and was based on primarily on hand x-rays. Enlarged joints and metaphyseal lesions were used to determine KBD severity. The study revealed no adult participants classified as having early-stage KBD, showing that the onset of KBD begins during childhood. That our patient reports her symptoms began at the age of 29, is unusual, as all participants in the study with KBD showed symptoms before the age of 16. Another possibility is that our patient did have minor symptoms of KBD at an earlier age but did not notice them until they progressed further.

Xiong recorded 20 different clinical sites of KBD, including the ankles, as in our patient (8). In this study, 66.1% of adults showed enlarged finger joints, a common finding in rheumatoid arthritis. In addition, 69.3% of participants in this study showed limited ankle motion. Despite her severe ankle pain, our patient had full range of motion in her ankles. KBD is more common in the distal appendicular skeleton; that is, fingers, wrists, and ankles are more likely to be affected than hips and shoulders (9).

KBD progression begins with smaller joints in the early stages of disease; over time, larger joints become involved (8). While lesions exist in both upper and lower extremities, damage is more frequent and severe in the lower limb. There is a significant relationship between the radiological appearance on an anatomical site and the clinical stage of KBD (9). The joint involvement pattern of KBD is similar to that of rheumatoid arthritis (9).

Multiple treatments and therapies have been tried for KBD, including physical therapy, medication, and surgical procedures such as osteotomy of the knee (10, 11, 12). Of these options,

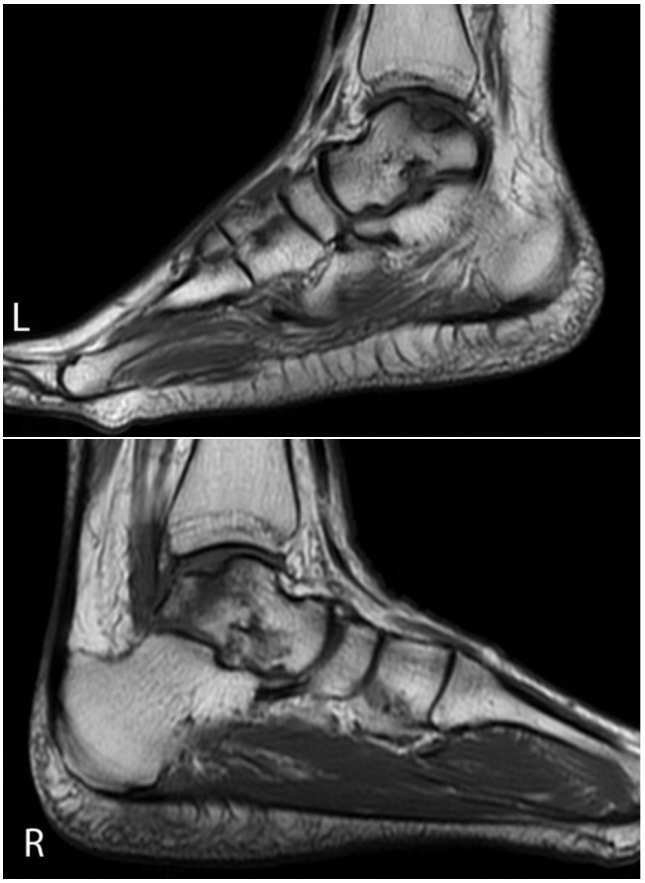

Figure 2. 41-year-old female with Kashin-Beck disease. Sagittal T1W images of left and right ankles show irregularity and flattening of both talar domes, consistent with subchondral collapse.

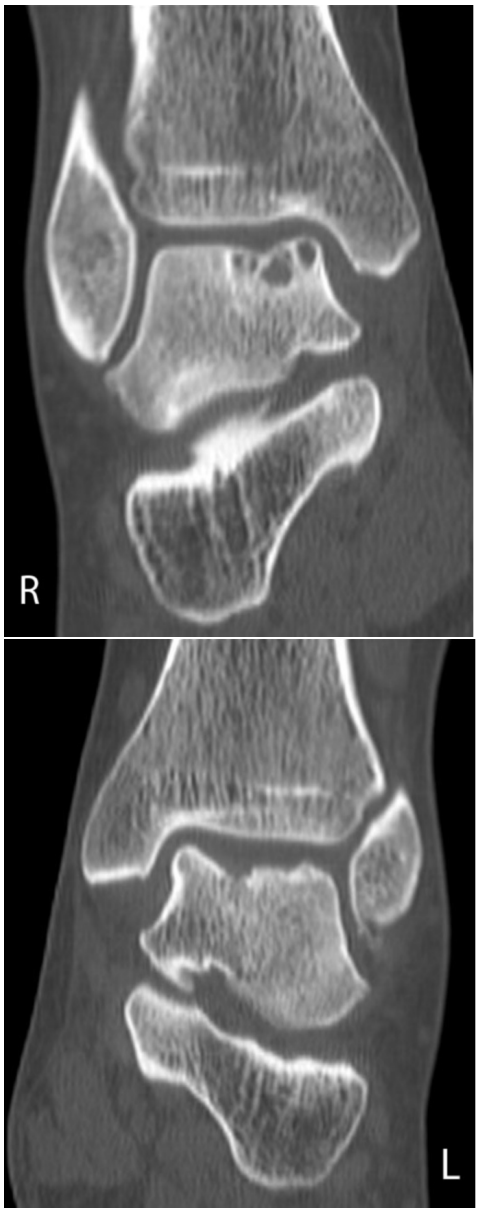

Figure 3. 41-year-old female with Kashin-Beck disease. Coronally reformatted CT images of right and left ankles show irregularity and flattening of both talar domes, consistent with subchondral collapse.

Figure 4. 41-year-old female with Kashin-Beck disease. Distribution of KBD in China. The prevalence of KBD varies from less than 10% to greater than 40% within the area outlined by the red line (4).

all three may offer some symptomatic relief, but only osteotomy improves anatomical deformities and bone deterioration. Other treatment options include osteoarticular allograft, total ankle arthoplasty, and ankle arthrodesis (13). Other less invasive methods include arthroscopic debridement and microfracture, transfer of autologous osteoarticular tissue from the knee or talus, and autologous chondrocyte implantation (14).

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