ISSN: 2690-9189

Editorial

International Journal of Orthopaedics Research

Tarsal Navicular Osteonecrosis in Children

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Submitted: 30 Nov 2019; **Accepted**: 16 Dec 2019; **Published**: 24 Dec 2019

Abstract

Osteonecrosis (avascular, aseptic or ischemic bone necrosis) of the tarsal navicular in children may develop either spontaneously (primary, idiopathic, atraumatic or non-traumatic) or secondary to trauma (post-traumatic) and osteochondrosis. In both groups, of primary and secondary osteonecrosis, the clinical findings as well as the radiographic abnormalities are self-limited and usually resolve spontaneously irrespective of weight-bearing and immobilization treatment modalities. Köhler's disease has been defined either as atraumatic navicular osteonecrosis or as an osteochondrosis process, based on the similar radiographic appearance of increased sclerosis and flattening detected in both asymptomatic and symptomatic children. Post-traumatic tarsal navicular osteonecrosis in children may follow microtrauma or overuse injuries, stress fractures, acute fractures, osteochondritis dissecans and severe foot injuries. This editorial aims to present the primary and secondary causes of osteonecrosis of the tarsal navicular in children and to describe the difficulties of the clinical and radiological evaluation in order to define an accurate diagnosis.

Keywords: Tarsal navicular, Osteonecrosis, Köhler's disease, Children.

Editorial

Osteochondrosis is a group of disorders that affect the growing skeleton and are usually due to a vascular insult leading to secondary osteonecrosis. This term is preferred than that of osteochondritis, which more specifically refers to infection or inflammation of bone and cartilage. There is no consensus on the etiology and multiple contributing etiologic factors, such as microtrauma, rapid growth, genetic, and/or hormonal factors, may be involved. The condition may be either in the primary deformans or in the dissecans form. The former affects the entire primary ossification center, while the latter affects a more limited bone and cartilage portion of weight-bearing areas in older children [1-5].

Osteochondrosis of the tarsal navicular in the pediatric patients or Köhler's disease was named after the German radiologist who described it in 1908. In children, the navicular bone is the last tarsal bone to ossify. In girls, the ossification center appears between 18 to 24 months and in boys between 30 to 36 months of age. As the child grows and becomes heavier, the unossified navicular may be compressed between the already ossified talus and cuneiform bones. The compression of the non-ossified navicular may result in the squeezing of the perforating vessels in the central spongy bone, which could potentially lead to ischemia and avascular necrosis. Köhler's bone disease is most commonly seen in males (80%) aged 4 to 7 years. It is usually unilateral, although it may be bilateral in up to 25 to 30% of cases. Patients typically present with sudden medial-sided foot pain, swelling of the medial foot, and/or a limp. There may be point tenderness over the navicular bone on examination. When intermittent limping is the only clinical manifestation, the diagnosis may be considerably delayed.

On plain films, the standard characteristics of osteonecrosis including patchy sclerosis, fragmentation, and flattening (thinning, decreased anteroposterior diameter) of the tarsal navicular are evident. Multiple centers of ossification as well as accessory ossicles, such as the accessory navicular bone, os supranaviculare and os infranaviculare, may be seen on the radiographs. There have been no reported cases of Köhler's disease developing long-term clinical or radiologic abnormalities. In most cases, the navicular assumes a relatively normal appearance within 1 to 3 years after symptom onset. Cortical irregularity may be the only late radiographic finding, but almost all of these cases remain asymptomatic.

There is a divergence of opinions whether Köhler's disease may be asymptomatic or not. Most authors believe that the diagnosis of navicular osteochondrosis requires a combination of clinical and radiological signs. Therefore, symptomatic or true navicular osteochondrosis should be differentiated clinically from asymptomatic roentgenographic changes resembling Köhler's osteochondrosis. In addition, it has been suggested that the isolated presence of radiological signs represents abnormalities of ossification, which are very common in the tarsal navicular bone in children, particularly those who have late-onset ossification. In asymptomatic cases the radiographic findings of the tarsal navicular bone may also be completely remodeled after the disease has run its course. It should be emphasized that it still remains unclear whether Köhler's disease represents a self-limiting symptomatic abnormality of ossification or a spontaneously resolving osteonecrotic process.

Recent reviews have shown that a short leg cast for up to eight weeks accelerates resolution of symptoms, although long-term outcomes are favorable regardless of treatment. Köhler's disease may often be misdiagnosed as osteomyelitis in children. A diagnosis of Köhler's disease does not require a bone biopsy, and a biopsy is not recommended for the diagnosis unless there is the need to rule out infection or malignancy [6-23].

Köhler's disease should be differentiated from the spontaneous navicular osteonecrosis of adults (Müller-Weiss syndrome also known as Brailsford's disease), which is more common in females and results in chronic mid-foot pain. Differential diagnosis of the post-traumatic tarsal navicular osteonecrosis in children includes a wide array of disorders, but most likely microtrauma or overuse injuries of the navicular bone, stress fractures, acute fractures, osteochondritis dissecans (OCD) and severe foot injuries, all of which should be readily differentiated from the typical presentation of osteochondrosis by conventional radiography. The usual radiographic findings of post-traumatic tarsal navicular osteonecrosis are loss of trabecular pattern, bone collapse, increased density, sclerotic bone, flattening and fragmentation [24-29].

Repetitive microtrauma or stress from overuse may lead to bone or soft tissue ankle and foot disorders. Overuse foot injuries are the most prevalent injuries in ballet dancers. Professional ballet dancers suffer different types of injuries depending on their age and years of professional practice. Female ballet dancers have a higher incidence of injuries than male ballet dancers or modern dancers, in part because they dance on the tips. Repetitive microtrauma from overuse may be indicated by navicular bone sclerosis, usually appearing as a single late radiographic finding [30-32].

Stress fractures are increasingly reported in the pediatric population. Stress fracture occurs when normal stress is applied to abnormal bone or abnormal stress is applied to normal bone and represents a disturbance between bone resorption and bone regeneration. In the case of normal bones, it is felt to be a fatigue fracture or overuse injury [33].

The location of the navicular predisposes it to unique patterns of stress and impingement between the head of the talus and the cuneiforms during running or jumping, resulting in fatigue failure of the navicular bone, which is defined as the cornerstone of the medial arch. Navicular stress fractures account for about 15% of all stress fractures, 14 to 35% of all foot and ankle stress fractures, while track and field athletes account for 59% of all navicular stress fractures. However, a non-athlete's complaint of medial-sided foot pain should never be discounted [34-38].

Navicular stress fractures are typically diagnosed with an average four month delay from the initial symptom onset. Various factors contribute to the common delay in diagnosis of navicular stress fractures. The navicular bone does not heal with callus formation, so the radiographic appearance of periosteal apposition, commonly seen in the healing of fractures of the long bones at the end of 3 weeks, is not expected. Athletes can often continue activity until pain increases too much by altering their gait and decreasing use of the forefoot. Pain also resolves rapidly with rest, making it possible for athletes to resume participation after a week of respite from activity. Patients who present with navicular stress fractures typically have a normal range of motion and strength to manual muscle testing. History usually elicits risk factors such as overuse with an increase in exercise duration and intensity as well as poor nutrition. The pain typically increases with activity such as running and jumping. Patients generally exhibit increased pain while standing on their toes in the equinus position. With continued participation, the pain occurs sooner during activity and lasts longer into post-activity rest periods. Symptoms are rarely bilateral [29, 39-46].

When suspicion justifies diagnostic studies, the initial step is typically plain radiographs. Unfortunately, only 33% of plain radiographs have sensitivity for navicular fractures, because the majority of fractures are incomplete. In addition, since bone resorption requires 10 days to 3 weeks to allow visualization of fractures on plain radiographs, even complete fractures are often missed on initial films. However, plain films are useful if positive, and they also assist in ruling out other etiologies. Early diagnosis may be evident by ultrasound, which is a good screening test for stress fractures. Computed tomography is the best tool in the diagnosis of navicular stress fracture. It allows differentiation between stress fracture and stress reaction, a periosteal stress-induced reaction, and enables accurate fracture definition [47].

If cost is not an issue, and time is of the essence, a magnetic resonance imaging can give information similar to that of bone scan plus computed tomography scan and it may rule out a stress reaction. Early diagnosis of these lesions and proper management usually yields a favorable outcome. There must be a high incidence of suspicion for these fractures because the navicular has poor blood supply over the middle one third, and most of the impingement force is focused at the central third of the navicular bone during stride. Thus, a combination of structural and vascular anomalies may make some individuals more prone to the formation of navicular osteonecrosis than the broader population. Compression of the small vascular foramina that help to supply the central area of the tarsal navicular, which are smaller than 1 millimeter in adults, could result in decreased blood flow and osteonecrosis. An untreated fracture may be complicated by poor healing with delayed union or nonunion. Non weight-bearing cast immobilization for 6 weeks usually results in fracture healing. The usual time to return to athletic activity may be as long as 5 to 6 months [48-58].

The most frequent navicular fracture in children is a dorsal proximal chip fracture, which is best seen on a lateral radiograph of the foot. This injury may represent an avulsion pull-off of an apophyseal fragment from the dorsal tarsal ligament [59, 60]. Complete displaced fractures of the navicular in children, involving either the tuberosity or the body, are very rare and they often result from high-energy trauma; therefore, associated injuries, such as those of the Lisfranc complex, are common. Since much of the bone surface is intra-articular, closed or open reduction and internal fixation may be indicated for displaced fractures. Assessment of the soft-tissue envelope is important in these high-energy injuries, and compartment syndrome should be ruled out [61, 62].

The cause of OCD of the tarsal navicular bone remains unknown. It has been suggested that mechanical stress may be an essential factor. Compression fracture of the secondary center of ossification is an important factor in the pathogenesis of osteochondrosis dissecans

[63]. In particular, axial loading in plantar flexion causes a high degree of mechanical stress on the navicular bone, which is crunched between the talus and the cuneiforms in this position. Following this theory, it would be conceivable that the history of ballet dancing could have contributed to the development of OCD of the navicular by repetitive microtrauma, as it is often reported in OCD of the knee in athletic adolescents. It has previously been suggested that it would be interesting to conduct a study to examine the incidence of OCD of the tarsal navicular in active ballet dancers versus participants in a control group. Diagnosis of OCD may often be made by conventional radiography. Focal lucency that disrupts the subchondral cortical line with surrounding sclerosis is characteristic of OCD. Computed tomography has the ability to uncover small lesions that may be indistinguishable on plain radiographs and to determine the exact size and localization of lesions in preparation for surgical treatment. Magnetic resonance imaging is useful to assess the integrity of the articular cartilage that covers the lesion as well as the viability of the subchondral bone and thus can help to guide treatment decisions [64-70].

Figure 1: Anteroposterior and oblique images in a 5-year-old boy showed flattening and fragmentation of the tarsal navicular indicative of Köhler's disease on the right side. His parents reported mild discomfort on the medial side of the foot, especially after overactivity, since 3 months ago

Our experience related to the tarsal navicular osteonecrosis in children indicates that only Köhler's bone disease may be diagnosed and treated properly, although a usually significant period of time is required for the patient to seek medical advice (Figure 1). On the contrary, in the young growing athletes involved in dancing or sport injuries, including participation in ballet dancing, taekwondo, kung fu fighting, basketball players and runners, that were examined in our outpatient clinic the diagnosis of tarsal navicular osteonecrosis, other than Köhler's bone disease, was considerably delayed and in most cases it was incidentally made, since the patients were referred for an acutely suffered ankle or foot injury. Moreover, it was impossible to differentiate the true nature of the navicular bone post-traumatic osteonecrosis in most of these cases, as well as to define an accurate radiographic diagnosis, such as a lesion following microtrauma due to overuse injuries, a healing stress or acute fracture and OCD, since there was no specific relevant information from the history in any of the patients. The difficulty to establish a radiographic diagnosis in these patients was most likely due to the considerable period of time that had elapsed from the initial insult, so that the findings of navicular bone remodeling on plain films is more prominent than the findings of the primary lesion (Figure 2, 3, 4, 5).

Figure 2: Anteroposterior and lateral radiographs in a 5-year-old girl following a lateral ankle injury indicated a navicular bone deformity on the right side. She participated in artistic gymnastics but reported no history of a foot injury or complaint. The radiographic appearance of the navicular bone lesion was indicative of healing following osteonecrosis of the dorsal part that could be due to an overuse injury, a stress fracture potentially associated with an os supranaviculare, an acute avulsion fracture, and less likely, due to the age of the patient, an OCD

Figure 3: Lateral radiographs in a 6-year-old girl following an ankle injury demonstrated a bilateral navicular bone deformity. She participated in ballet dancing since 3 years ago, twice a week. There was no history of a foot injury or complaint. The radiographic appearance of the navicular bone lesion on both sides indicated bone healing following osteonecrosis of the dorsal part that could be due to a potential acute fracture, a stress or overuse injury, and less likely, due to the age of the patient, an OCD

Figure 4: Anteroposterior and lateral radiographs in a 9-year-old boy following an ankle injury demonstrated a bilateral navicular bone deformity. He participated in taekwondo since 3 years ago, for an hour three times a week. There was no history of a foot injury.

He reported minor complaints following intense athletic activities during the last 2 years. The radiographic appearance of the navicular bone lesion on both sides indicated healing following osteonecrosis of the dorsal part that could be due to a potential acute fracture, a stress or overuse injury, and an OCD

Figure 5: A 10-year-old boy injured his right foot after a fall from a height, during athletic training. There was no obvious fracture line on the initial radiographic evaluation. No restriction of physical activities was undertaken. He was referred 3 months post-injury with medial-sided discomfort after intense athletic activity. Anteroposterior and oblique radiographs revealed a nondisplaced navicular fracture through the body associated with flattening and fragmentation of the fragments, which were indicative of post-traumatic osteonecrosis. In an asymptomatic patient with no trauma history a developmental variant of ossification would be the most likely diagnosis. A thin translucent line was evident on the medial cuneiform, on the oblique view, that could be consistent with bipartition

In conclusion, it is clearly evident that the clinical and radiographic distinction of the non-traumatic and post-traumatic osteonecrosis of the tarsal navicular in the growing skeleton may occasionally be demanding. In the non-traumatic lesions the diagnosis of Köhler's bone disease is usually based on the clinical and the typical radiographic findings. Repetitive microtrauma from overuse may be indicated by the navicular bone sclerosis, usually appearing as a single late radiographic finding. In the post-traumatic osteonecrosis the diagnosis is usually significantly delayed mainly due to the late radiographic appearance of stress fractures, acute undisplaced fractures and OCD of the tarsal navicular bone. Therefore, the examination of these patients, involving both clinical and radiological findings, is usually significantly delayed and, therefore, it is not sufficient to make a distinct differential diagnosis of the post-traumatic lesions.

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