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Nonsyndromic Massive Tarsal and Tarsometatarsal Coalitions in a Young Female

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ABSTRACT

Tarsal coalition is an abnormal connection between 2 or more tarsal bones, with the most common sites being between the talus and the calcaneus and between the navicular and the calcaneus. The occurrence of multiple and massive tarsal coalitions is rare. We describe a rare case of nonsyndromic bilateral tarsal coalition involving most of the tarsal bones and extending to the metatarsal bones in a 4-year-old female. The condition was not painful and did not affect her gait. The main concern was the abnormal shape of her feet. Tarsal coalition can occur as an isolated anomaly or in association with other congenital disorders, usually presenting around the age of 12 years. The coexistence of nonsyndromic bilateral multiple tarsal coalitions is seldom reported. Regardless of the presentation, treatment is not indicated in cases of asymptomatic tarsal coalition. Our patient had a unique presentation of tarsal coalition, involving both the tarsal and tarsometatarsal bones. To our knowledge, this is the first report of this type of presentation.

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Tarsal coalition is an abnormal connection between 2 or more bones at the back of the foot known as the tarsal bones, and the condition may produce pain and limit foot mobility (1). The connection can be fibrous, cartilaginous, or bony in nature (2), and it can be complete or incomplete. Multiple causes of tarsal coalition have been identified, including congenital, post-traumatic, infectious, and surgical, as well as joint disorders. In congenital cases, the coalition develops from failure of differentiation and segmentation of the primitive mesenchyme, which leads to failure of normal tarsal joint formation (3,4).

No clear data are present regarding the true incidence of tarsal coalition, because the disorder is usually asymptomatic and does not affect function; therefore, it goes unnoticed. However, the incidence has been estimated in a few studies to range between 1% and 12.7%, with bilateral presentation in 50% to 60% of cases (4,5).

The most common sites of tarsal coalition are between the talus and the calcaneus (medial talocalcaneal joint), which is seen in about 48% of cases, and between the navicular and the anterior process of the calcaneus, which is seen in about 44% of cases (2,6); coalition between the talus and the navicular or between the calcaneus and the cuboid is identified in

an additional 1% of cases each. Coalitions involving the 3 joints of the mid-foot are less frequent, with involvement of multiple joints and massive tarsal coalition being rare (6–11). Therefore, complete tarsal coalition, with synostosis manifested in all tarsal bones and the coalition extending to the metatarsal bones, is extremely rare. Moreover, tarsal coalition often remains silent; thus, surgery is rarely performed. When the disorder becomes symptomatic, it is typically between the ages of 8 years and 12 years as the coalition ossifies. Symptoms typically develop insidiously, beginning with vague pain; other complaints are an abnormally shaped foot, a rigid or spastic flat foot, and recurrent ankle sprains (3,9,12).

We encountered nonsyndromic bilateral tarsal coalitions in a very young female. Most of the tarsal bones were involved, and the abnormality extended to the metatarsals. To understand this case, we searched the MEDLINE and SciVerse Scopus databases by using the terms “multiple tarsal coalition,” “massive tarsal coalition,” and “tarsal synostosis,” but we found no report of a case that presented similarly.

In this report, we describe not only the condition but also our line of thinking and resulting clinical strategy.

Case Report

A 4-year-old female presented to our pediatric orthopedic clinic with her parents, who had noticed deformity of both of her feet. The parents first noticed that her left foot was abnormally shaped when she

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Fig. 1. Photographs obtained on the initial examination of the patient. The posterior view (A) shows both hindfeet in fixed varus, and anterior views (B, C) show both feet in fixed supination. The patient's big toes are very large, especially the left big toe.

had begun to walk at around 12 months of age. The female had not complained of foot pain either at rest or during walking and the parents had not observed any gait abnormality. There was no family history of a similar condition or other foot deformity.

On general examination, no dysmorphism or spinal anomaly was found. Both hips and knees had full range of motion and no other gross morphological abnormality was observed.

On physical examination, both feet were in fixed supination (left > right), and this remained during walking. The first ray of each foot was larger than the other rays and did not contact the ground during weightbearing. The hindfoot was in fixed varus (Fig. 1). No noticeable swelling and no tenderness on palpation were found. Both ankles had full range of motion, including full passive dorsiflexion range of motion, with the knee in flexion and extension, and there was no palpable tightness of the Achilles tendon on either side. However, subtalar motion was absent bilaterally. The medial arch of the midfoot was normal bilaterally, and there was no pes planus or evidence of antalgic peroneal spasm on either the right or left. The midfoot was rigid, with no movement possible. With supination of the forefoot, bilaterally, little forefoot abduction or adduction was possible. Owing to these limitations, the patient walked on the outer sides of her feet.

Plain radiography revealed coalition between multiple tarsal bones bilaterally but in different forms (Fig. 2). Computed tomography confirmed the presence of complex osseous coalition between the tarsal bones, extending to the metatarsals (Figs. 3 and 4; Table).

We decided to simply monitor the case until the patient attained physical maturity or the condition became symptomatic. Over an observation period of 30 months (February 2016–August 2018), the deformity did not progress in either foot and the condition remained asymptomatic.

Discussion

Tarsal coalition is an abnormal union of 2 or more adjacent tarsal bones that have failed to differentiate during embryonic development, with the union being fibrous (syndesmosis), cartilaginous (synchondrosis), or osseous (synostosis) in nature (2). Tarsal coalition can occur as an isolated anomaly or in association with other congenital disorders, such as fibular hemimelia, clubfoot, Apert syndrome, and Nievergelt-Pearlman syndrome (3,9). There are several reports of familial occurrence of coalition. Wray and Herndon (13) described occurrence of a calcaneonavicular coalition in 3 generations of a single



Fig. 2. Plain radiographs obtained on the first visit. Standing anteroposterior radiographs (A) and lateral radiographs (B) of both feet show massive tarsal and tarsometatarsal coalitions.

family and proposed that the inheritance pattern was that of autosomal dominance with reduced penetrance.

Patients with tarsal coalition usually present around the age of 12 years, when progressive ossification of the coalition occurs, leading to hindfoot stiffness and restricted subtalar motion. Some cases, though rare, have been reported in children as young as 6 years (10).

Although multiple tarsal and tarsometatarsal coalitions have been identified as a component of such rare congenital syndromes as Antley-Bixler syndrome and Fuhrmann syndrome, which derive from inheritable gene mutations for tarsal-carpal coalition and multiple synostosis (14–17), the occurrence of nonsyndromic multiple tarsal coalitions has been reported only rarely (18).

The bilateral, asymmetrical, massive tarsal and tarsometatarsal coalitions in our patient were unique, in comparison with the coalitions reported previously. Other unique features included the absence of a family history and the early presentation at 4 years of age. Neither fibular hemimelia nor carpal coalition were found (Fig. 5). No dysmorphia was identified and the patient's development and cognitive function were normal for her age. The salient feature in this case was the abnormal shape of the feet.

Radiographs of the feet in the 3 standard views (anteroposterior, lateral, and 45° oblique) should be obtained routinely for any patient with a suspected tarsal coalition, and computed tomography should be performed to confirm the diagnosis, to determine the type and severity

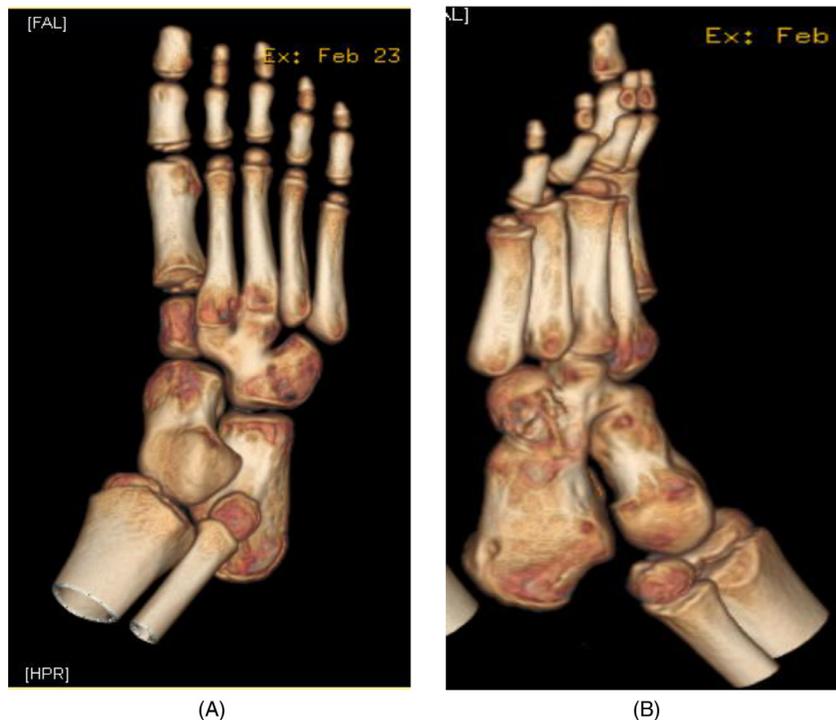


Fig. 3. Three-dimensional computed tomographic images of the right foot (A) and left foot (B), confirmed the massive tarsal and tarsometatarsal coalitions.



Fig. 4. Sagittal computed tomographic images of the right foot (A) and left foot (B) show the massive tarsal and tarsometatarsal coalitions.

Table

Sites of tarsal and tarsometatarsal coalition in the patient's right and left feet

	Site of Coalition
Right foot	Calcaneus with talus
	Talus with navicular
	Cuboid with cuneiform (lateral and middle)
	Cuneiform with second, third, and fourth metatarsals
Left foot	Calcaneus with talus
	Talus with navicular
	Navicular with cuneiforms
	Cuneiform with cuboid
	Calcaneus with cuboid
	Cuneiform with third metatarsal

of the coalition, and to identify the bones that are involved. Magnetic resonance imaging is useful in cases of fibrous coalition (9,11,19,20). The information derived from both computed tomography and magnetic resonance imaging is useful for surgical planning when surgical treatment is deemed necessary in symptomatic cases.

There is no evidence to support any treatment intervention for asymptomatic tarsal coalition. Treatment is indicated only for patients with pain or severe deformity (10,21). Even when the condition is painful, nonoperative treatment should be tried first, including activity modification, immobilization (with a cast or walking boot), use of a soft shoe insert, and use of nonsteroidal antiinflammatory drugs or steroid injections for pain control. Surgical treatment is indicated only after conservative treatment has failed, with the intent being pain relief. The success of surgical treatment depends on the ability of the surgeon to excise the coalition while minimizing damage to the adjacent structures to the fullest extent possible (22). Multiple surgical options are available, including resection of the coalition, osteotomy, and arthrodesis, depending on patient's age and presentation, foot flexibility, type or extent of coalition, and whether degenerative changes are present in the affected or adjacent tarsal bones (23). Massive tarsal and/or tarsometatarsal coalition in a nonsyndromic patient is a rare condition. Owing to its rarity, there are no specific recommendations regarding treatment in such cases. However, it is generally accepted that the treatment of the condition should be aimed at symptom relief. In our



Fig. 5. Anteroposterior radiograph of the right wrist obtained as part of the case evaluation shows normal carpal bone architecture without any sign of coalition.

opinion, immediate treatment is not necessary for patients presenting at a young age, before skeletal maturity, who do not have pain or a gait abnormality. We believe that when the only presenting symptom in such patients is deformity of the foot that is neither progressive nor painful, follow-up in the form of clinical observation is appropriate until the development of any debilitating symptoms.

In conclusion, the clinical presentation of tarsal coalition in our patient was unique, especially because it was bilateral. The coalition pattern differed between feet but included both tarsal and tarsometatarsal

bones, and it occurred in the absence of any syndrome. An accumulation of similar cases is needed to develop clinical pathways for the identification, diagnosis, and treatment, whether conservative or surgical, so that we can easily approach this type of deformity in the future.

Written informed consent was obtained from the parents of the patient for publication of this case report and the accompanying images. Considering that we have reported a single case and not a clinical study, there was no requirement for ethical approval.

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