Hypoplasia of the Left Superior Ramus of the Pubis

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We present the case of an 11-year-old girl with hypoplasia of the left superior ramus of the pubis in isolation and without associated congenital anomalies.

Case Report

An 11-year-old girl presented to our clinic with a complaint of intoeing. Physical examination revealed increased femoral anteversion with a neutral tibial transmalleolar axis. The patient walked with an internal foot progression angle of 15°. Her elbows, hands, knees, and feet were normal in appearance and motion. The rest of the physical examination revealed no other abnormalities, including no palpable defect at the symphysis and an intact inguinal ligament. Pelvis radiograph showed hypoplasia of the superior ramus of the pubis on the left, along with the inferior ramus forming the left side of the symphysis pubis (Figure 1). Computed tomography (CT) scan showed a fully developed acetabulum, but there was only a very small projection of the pubic bone itself (Figures 2A, 2B). Subsequent renal ultrasound was normal.

Discussion

This is the first literature report of hypoplasia of the left superior ramus of the pubis in isolation and without associated congenital anomalies. Indeed, the conditions associated with this anomaly are themselves rare. Cortina and colleagues[1] reviewed 5 cases in which there was nonossification of the pubis. The cause in these cases included the following syndromes: Sjögren-Larsson syndrome, Wolf syndrome, cleidocranial dysostosis, spondyloepiphyseal dysplasia congenita. These syndromes have other distinctive morphologic features, which were absent from our patient's case. Schey and Levin[2] reported a case of familial maldevelopment of the pubic bone. A father and his 3 sons had delayed ossification of the medial portion of both pubic bones, and 2 of the brothers presented with transient hematuria. Although the cause of the hematuria was not discovered, the children were followed out to their teenage years, and their pubic bones continued to ossify. In the French literature, Slullitel[3] described the case of an infant who underwent a hernia repair and was found to be missing both pubic bones; the author discussed 3 similar cases. In a German-language case report, Plaue[4] described a patient who was missing one pubic bone, but this patient had gait abnormalities as well.

Other well-described syndromes are associated with pubic aplasia. Nail-patella syndrome, also known as osteo-onychodysplasia or Turner-Fong syndrome, has the distinguishing features of nail abnormalities and hypoplastic or absent patella. The elbow can also have problems, such as radial head hypoplasia, which leads to subluxation or dislocation. In addition to the musculoskeletal abnormalities, there can be renal problems, such as glomerulonephritis or nephrotic syndrome. Nail-patella syndrome is inherited in an autosomal-dominant pattern. Azouz and Kozlowski[5] and Habboub and Thneibat[6] separately described a less serious and rarer syndrome involving hypoplasia of the patella, pubic, and ischial bones, but their patients also had cleft palate and motor developmental delay. Scott and Taor[7] described 12 family members with patellar aplasia or hypoplasia. Of these, 7 had pelvic abnormalities, but the nature of these abnormalities was not discussed. Sferopoulos and Tsitouridis[8] described ischiopubic hypoplasia as a rare constituent of several congenital syndromes, including small patella syndrome and developmental dysplasia of the hip. Köhler and Zimmer[9] stated in their classic text, *Borderlands of Normal and Early Pathological Findings in Skeletal Radiography,* "The superior pubic ramus may be absent, associated with an underde-

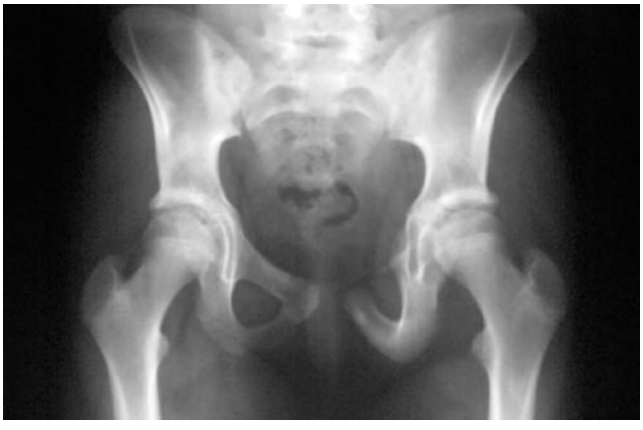

Figure 1. Upright anteroposterior radiograph of pelvis in 11-year-old girl.

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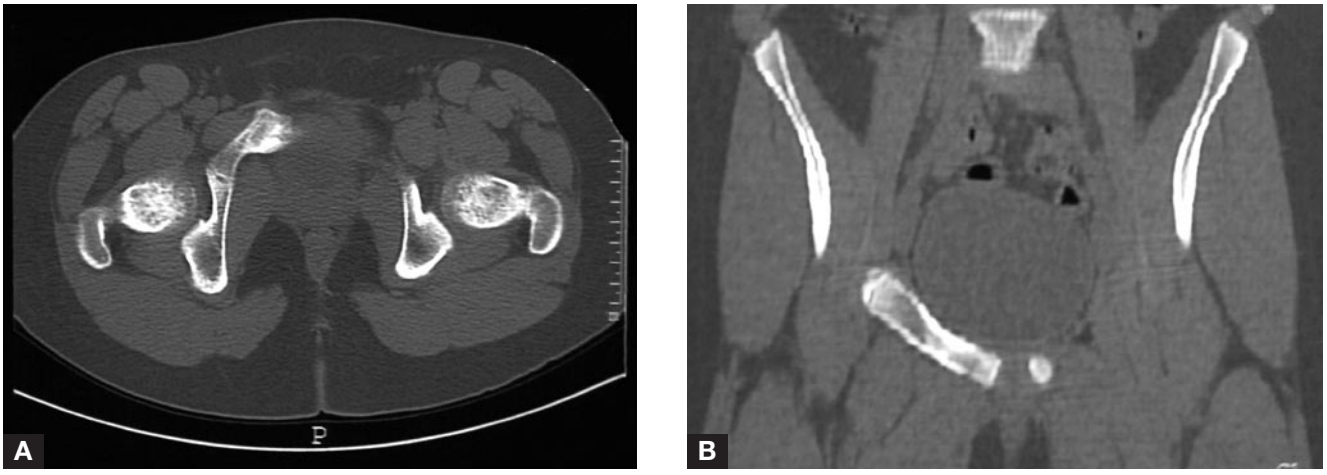

Figure 2. **(A)** Axial computed tomography (CT) scan shows lack of superior ramus. **(B)** Coronal CT scan shows tip of inferior pubic ramus on the left forming symphysis with the right pubic bone.

veloped ipsilateral leg." Our patient had well-developed legs with normal patellae and musculature.

Pubic bone ossification begins in the fetus between the fifth and sixth fetal month. The ossification center is in the superior ramus near the central and inferior margin of the acetabulum near the triradiate cartilage. The bone continues to grow by enchondral bone formation. By this stage, the fetal parts have differentiated, and the fetus undergoes growth and maturation for the rest of its gestation. An insult to the fetus during the differentiation stage likely results in multiple anomalies within multiple organ systems, whereas an insult during the growth-and-development stage could result in an isolated anomaly.

In our patient, CT scan showed a normal-appearing acetabulum, which could indicate that the os acetabuli, as described by Ponseti,[10] developed normally while the pubis bone proper failed to develop at all. It is also possible that further ossification could occur as this patient ages. Her CT scan did not show any evidence that a cartilaginous superior ramus had failed to ossify. Magnetic resonance imaging would have been helpful in this respect but was not performed; should the patient not experience further ossification during her growing years, a scan may be obtained then. The clinical significance of hypoplasia of the pubic bone in this patient is unknown but may be of concern during pregnancy or delivery. The soft tissues in this area may also be abnormal, which could lead to hernias as well.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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This paper will be judged for the Resident Writer's Award.