



## Bilateral congenital dislocation of the patella associated with synostosis of proximal tibiofibular and proximal radioulnar joints: A case report

Proksimal tibiofibular ve proksimal radioulnar eklemlerin sinositozunun eşlik ettiği iki taraflı doğuştan patella çıkığı: Olgu Sunumu

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### ABSTRACT

Congenital dislocation of the patella is a rare and difficult pathology to treat. We present a case of bilateral congenital dislocation of the patella with synostosis of proximal tibiofibular and proximal radioulnar joints without genu valgum deformity of both knees in a 30-year-old man. To our knowledge, congenital dislocation of the patella associated with synostosis of proximal tibiofibular and proximal radioulnar joints has not been reported in the literature yet.

**Keywords:** Congenital patella dislocation; radioulnar synostosis; tibiofibular synostosis.

### ÖZ

Doğuştan patella çıkığı nadir ve tedavi edilmesi zor bir patolojidir. Bu yazıda, 30 yaşında bir erkek hastada her iki dizde, genu valgum deformitesinin eşlik etmediği iki taraflı doğuştan patella çıkığı, proksimal tibiofibular sinositoz ve proksimal radioulnar sinositozun bulunduğu bir olgu sunuldu. Bizim bilgimize göre, proksimal tibiofibular sinositoz ve proksimal radioulnar sinositozun eşlik ettiği doğuştan patella çıkığı henüz literatürde bildirilmemiştir.

**Anahtar sözcükler:** Doğuştan patella çıkığı; radioulnar sinositoz; tibiofibular sinositoz.

Congenital dislocation of the patella (CDP) is often familial and bilateral and may be followed by other abnormalities such as arthrogryposis multiplex congenita, and Down syndrome. This deficiency typically occurs around the 8<sup>th</sup> to 10<sup>th</sup> week of embryonic development.<sup>[1]</sup> It is permanent, irreducible, and generally followed by abnormalities of the quadriceps mechanism. Absence or severe contraction of the vastus lateralis may occur, and the patella may be dislocated laterally and attached to the anterior aspect of the iliotibial band. The capsule on the medial side of the knee may be stretched, the lateral femoral condyle flattened, or the insertion of the patellar tendon located more laterally than normal.

Congenital dislocation of the patella is uncommon but is a well-known orthopedic condition, and can manifest as distinctive clinical presentations. It is believed that failure of internal rotation of the myotome which originates the femur, the quadriceps muscle, and the extensor mechanism causes CDP. Congenital dislocation of the patella generally presents as genu valgum, flexion contracture, and external rotation of the tibia after birth.<sup>[2]</sup> The congenital form is permanent, irreducible, and presents at birth. It is characterized by short quadriceps, a major patellofemoral dysplasia, and short height.<sup>[3]</sup> Diagnosis is often made by clinical examination and observation of an abnormal gait. Once the diagnosis is made, early surgery can be performed to prevent valgus

Received: May 04, 2018 Accepted: May 23, 2018

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### Citation:

İlyas G, Eren TK, Kaptan AY, Ulucaköy C, Kanatlı U. Bilateral congenital dislocation of the patella associated with synostosis of proximal tibiofibular and proximal radioulnar joints: A case report. Eklem Hastalıkları Cerrahisi 2018;29(2):123-127.



**Figure 1.** Clinical appearance of the lower extremity (anteroposterior).

deformity, flexion contracture, or external rotation deformity of the knee.

In this article, we report a male patient with bilateral CDP associated with synostosis of proximal tibiofibular and proximal radioulnar joints, a rare and difficult pathology to treat.

### CASE REPORT

A 30-year-old man was admitted to our hospital with pain in his patellofemoral joint region which



**Figure 2.** Clinical appearance of the elbow extension.

increased with movement and recurrent dislocation of his left patella during flexion. The patient's pain was intermittent and did not affect his daily life.

Extensive lateral retinacular release was applied for both knees in a different hospital after diagnosis when he was 13 years old. His parents were cousins, he had a healthy brother, and one sibling of his grandfather had scoliosis.

Physical examination of lower limbs showed that active and passive ranges of motion (ROM) of both knees were between  $0^\circ$  extension and  $120^\circ$  flexion. Quadriceps angles were  $10^\circ$  valgus when the knees were in extension (Figure 1). In extension, both patellae were palpable on the anterior aspect of the knee; however, in flexion the left patella dislocated laterally while the right patella remained reduced.

Physical examination of upper limbs showed that ROM of both elbows were between  $-30^\circ$  extension (Figure 2) and  $145^\circ$  flexion. Ranges of motion of the right forearm was between  $30^\circ$  pronation and  $45^\circ$  supination and the left forearm was between  $50^\circ$  pronation and  $35^\circ$  supination (Figure 3). Both wrists had normal ROM.

Merchant's view showed that the left patella was located at the lateral aspect of the femoral condyle, the right patella remained reduced and deviated laterally and both femoral trochlea were dysplastic (Figure 4). Sulcus angles were  $150^\circ$  in the right knee and  $154^\circ$  in the left knee. There were no degenerative changes according to radiographic assessment. Both patellae were hypoplastic. Anatomical tibiofemoral angles of the right and left knees were  $8^\circ$  and  $10^\circ$  of valgus and mechanical tibiofemoral angles were  $2^\circ$  and  $3^\circ$  valgus, respectively, on long-leg standing radiographs (Figure 5).

Radiographic views also showed bilateral O'Dwyer type II proximal tibiofibular synostosis which is described as complete bone bridging between the bilateral proximal tibia and fibula (Figure 6). There



**Figure 3.** Clinical appearance of the supination and pronation.



**Figure 4.** Merchant's view of the bilateral knees.

was also Cleary type III bilateral synostosis of the proximal radioulnar joint, which is defined as osseous synostosis with a hypoplastic and posteriorly dislocated radial head (Figure 7).

The patient did not have any systemic or metabolic disorder and he had a healthy mental state. Treatment options were presented to the patient but he did not accept either conservative or surgical treatment as his pain was not affecting his daily life.

#### DISCUSSION

Patients with CDP have distinctive symptoms and it should be recognized that CDP is clinically different from other patellar dislocations.<sup>[4,5]</sup> It is portrayed as a relatively rigid joint, short quadriceps, and major patellofemoral dysplasia. Congenital dislocation is permanent and irreducible, and it presents as flexion contracture of the knee, genu valgum, and external tibial torsion just after birth.



**Figure 5.** Long-leg standing radiography of the lower extremity.



**Figure 6.** Anteroposterior view of the bilateral knees.



**Figure 7.** Anteroposterior and lateral views of the bilateral elbows.

Every patient with arthrogyrosis, skeletal dysplasia, or other related syndromes should be suspected to have CDP. It should be particularly examined in patients who have knee flexion contracture, genu valgus, external tibial torsion, foot deformity, and delayed walking.<sup>[6]</sup> Congenital dislocation of the patella is frequently related to other congenital disorders such as dislocation of the radial head, congenital absence of the fibula, congenital cardiac anomalies, talipes equinovarus, and carpal fusion.<sup>[7]</sup> This case report, on the other hand, is related to synostosis of the proximal tibiofibular joint which has been described in only a few reports.<sup>[8-10]</sup> Careful physical examination of the patella, which is small, relatively fixed, and lies laterally on the lateral condyle, is required. Ultrasonographic examination helps to locate the

patella and verify the diagnosis.<sup>[11]</sup> Visualization of anatomical details and its relationship with the extensor mechanism is possible via magnetic resonance imaging (MRI) and should be used in preoperative planning.<sup>[12,13]</sup>

Congenital dislocation of the patella can only be treated surgically. Serial casting and bracing are effective in the treatment of flexion contracture of the affected knee, but genu valgum, external tibial torsion, and subluxation of the tibia inevitably develop without surgical intervention.<sup>[14]</sup>

Ghanem et al.<sup>[5]</sup> reported that all of the eight knees with CDP had genu valgum deformity. In our case, our patient did not have genu valgum despite his diagnosis of CDP.

Isolated synostosis of the proximal tibiofibular joint is uncommon and is usually presented in multiple hereditary exostosis, especially kissing osteochondromata,<sup>[8-10,15]</sup> or it may be related to other general syndromes and knee valgus.<sup>[16]</sup>

O'Dwyer<sup>[16]</sup> reported three distinct patterns of proximal tibiofibular synostosis. Leg length discrepancy, bowing of the fibula with increased interosseous distance, and valgus deformity of the knee were components of all three types.<sup>[16]</sup> According to O'Dwyer, the deformities present at birth or before closure of the epiphyseal plate. In the present report, our patient did not have the deformities mentioned above after the closure of epiphyseal plate, despite of having type II proximal tibiofibular synostosis according to O'Dwyer classification.

Proximal radioulnar synostosis is a rare congenital pathology and is often a component of syndromes such as Crouzon, Apert's, and Poland's syndrome. Radio-ulnar synostosis is considered a failure of longitudinal segmentation. Synostosis is mostly seen in the proximal one-third of the forearm. Approximately 40 percent of the cases are unilateral whereas the rest is bilateral. It is more common in male patients.<sup>[17]</sup> Genitourinary, gastrointestinal, cardiovascular, central nervous, and musculoskeletal system pathologies have been identified in almost one-third of patients with radio-ulnar synostosis.

Cleary and Omer<sup>[18]</sup> reported four distinct radiological patterns of radio-ulnar synostosis. Our patient had type III radio-ulnar synostosis according to Cleary's classification.

Congenital dislocation of the patella should be differentiated from other patellar dislocations which result from congenital or genetic syndromes. The tempting pathogenetic theory is described as failure of internal rotation of the quadriceps myotome

during embryonic development. We believe that genu valgum, which develops in CDP as reported in the literature, may not be developed in the presence of proximal tibiofibular synostosis and this may be the result of lateral mechanical block effect. We consider this hypothesis may be critical in new treatment methods if supported by other case series.

#### Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### Funding

The authors received no financial support for the research and/or authorship of this article.

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