Numerical variants of carpal bones have been extensively documented in the literature. More than 20 accessory ossicles scattered across the carpus have been described by O’Rahilly (1) and later by Senecal et al (2). Their incidence in the general population can vary from 0.4% to 1.6%, as demonstrated by O’Rahilly (1) and Bogart (3), respectively. The most frequently observed accessory ossicles are the os centrale, located between the scaphoid, capitae and trapezoid, and the os triangular, found in the triangular cartilage just distal to the ulnar styloid (4). They present as cartilaginous nuclei in the developing embryo and occasionally persist into adult life as normal anatomical variants (5). Supernumerary carpal bones can also arise from failure of fusion of ossification centres resulting in congenital anomalies such as a bipartite scaphoid (6). The entirety of these congenital variations may be found in isolation or occur in the context of a genetic disorder such as Larsen syndrome and otopalatodigital syndrome type 1. Other syndromes associated with accessory carpal bones include brachydactyly type A1 (Farabee-type brachydactyly; mainly shortening of middle phalanges), Ellis-van Creveld syndrome and Holt-Oram syndrome (4).

First described by Larsen et al (7) in 1950, Larsen syndrome is a rare congenital disorder occurring in one in 100,000 live births. It is characterized by a wide variety of craniofacial and musculoskeletal features such as hypertelorism, prominent forehead, depressed nasal bridge, flattened midface, cleft palate, short stature, equinovarus or equinovalgus foot deformities, and various hand anomalies, including supernumerary carpal bones. The authors present a case of Larsen syndrome with bilateral supernumerary carpal bones as the sole clinical manifestation. A literature review investigating skeletal anomalies in patients with Larsen syndrome was performed and revealed that the present case represents a unique presentation of this disorder, lacking all of the major clinical features previously described in the literature. An approach to patients with supernumerary carpal bones is discussed.

Key Words: Carpal bones; Larsen syndrome; Supernumerary
On questioning, the patient mentioned that he experienced knee pain, more pronounced on the right. Both ankle joints were hypermobile and fell into pronated position when standing. There was mention of similar findings in the patient’s father. No large joint dislocations were noted.

A skeletal survey was performed to detect any underlying radiological findings. X-rays revealed several anomalies, which were most notable at the level of the cervical spine. There was evidence of dysmorphic

TABLE 1
Summary of skeletal survey findings

| Area                  | Findings                                                                 |
|-----------------------|--------------------------------------------------------------------------|
| Cervical spine        | Abnormal morphology of the cervical vertebrae, particularly posterior elements. Nonfusion of the posterior elements of C6 and C7 and incomplete fusion at C2 Abnormal articular processes of all cervical levels with hypertrophy at C2-C5 The facet joints within the cervical spine appear dysplastic Mild increase in anteroposterior width of C2-C4 vertebral bodies Inferior end plate depression is also noted at multiple levels Anterolisthesis of C2-C3 |
| Thoracic spine and ribs | Unremarkable Alignment is anatomic                                         |
| Lumbar spine          | Possible rudimentary ribs at L1 and L2 Alignment is anatomic               |
| Pelvis                | Sacroiliac joints and pubic symphysis are congruent Hips are unremarkable |
| Bilateral humeri      | Unremarkable                                                               |
| Bilateral forearms    | Normal alignment at the elbow Mild remodelling of distal articular surface or the right radius Bilateral femora Mild lateral bowing of the distal femoral metaphysis bilaterally Bilateral tibia and fibula Rounded appearance of the talar domes bilaterally (ball-and-socket) |
| Feet                  | Abnormal configuration around the Lisfranc joints Evidence of medial angulation of the little toes bilaterally Tiny left calcaneal bony spur is noted |
| Full-length weight-bearing legs | The right leg is shorter by 7 mm compared with the left                         |
TABLE 2
Comparison of reported skeletal anomalies in patients with Larsen syndrome

| Characteristic | Babat and Ehrlich (11) | Bicknell et al (8) | Percin et al (12) | Rahalkar et al (13) | Stanley and Seymour (14) | Steel and Kohl (10) | Total, n/n (%) |
|---------------|------------------------|-------------------|-------------------|-------------------|-------------------------|------------------|----------------|
| Total patients, n | 3                      | 52                | 1                 | 1                 | 8                       | 3                | 68 (–)         |
| Patients with accessory carpal bones, n | 2                      | 48                | 1                 | 1                 | 6                       | 2                | 60 (–)         |
| Short stature (<10th percentile) | 2 of 2                 | 44 of 48          | 1 of 1            | 1 of 1            | 0 of 0                  | 0 of 0           | 48/52 (92.3) |
| Characteristic facies | 0 of 2                 | 47 of 48          | 1 of 1            | 1 of 1            | 6 of 6                  | 2 of 2           | 57/60 (95.0) |
| Hip dislocations | 1 of 1                 | 46 of 48          | 1 of 1            | 0 of 0            | 3 of 6                  | 2 of 2           | 53/58 (91.4) |
| Knee dislocations | 2 of 2                 | 45 of 48          | 0 of 0            | 0 of 0            | 2 of 6                  | 2 of 2           | 51/59 (86.4) |
| Elbow dislocations | 0 of 0                 | 42 of 48          | 1 of 1            | 0 of 0            | 6 of 6                  | 2 of 2           | 50/57 (87.7) |
| Foot anomalies | 1 of 1                 | 43 of 48          | 1 of 1            | 0 of 1            | 6 of 6                  | 2 of 2           | 53/58 (90.6) |
| Spatulate thumbs | 0 of 0                 | 47 of 47          | 1 of 1            | 0 of 0            | 6 of 6                  | 2 of 2           | 56/56 (100)   |
| Scoliosis | 0 of 0                 | 41 of 48          | 1 of 1            | 1 of 1            | 2 of 6                  | 1 of 2           | 46/58 (79.3) |
| Cervical spine anomalies | 2 of 2                 | 41 of 47          | 1 of 1            | 0 of 0            | 0 of 0                  | 0 of 0           | 44/50 (88.0) |

Posterior elements, abnormal articular processes, dysplastic facet joints, inferior end plate depressions and anterolisthesis at the level of C2-C3. The remaining findings are summarized in Table 1.

DISCUSSION

We present a case of bilateral supernumerary carpal bones as the main clinical finding in a patient with Larsen syndrome. To our knowledge, this is a unique presentation of this disorder, lacking all of the major diagnostic features previously described in the literature.

Supernumerary carpal bones were not emphasized in the original description of Larsen syndrome in 1950. Rather, other features, such as multiple large joint dislocations, characteristic facies and equinovarus foot deformities were considered to be the cardinal features of this disorder (8). Accessory carpal bones were first reported in the context of Larsen syndrome by Steel and Kohl (10) in 1972. Since then, only a few other articles reported supernumerary carpal bones in Larsen syndrome. Despite being an infrequently reported finding, Bicknell et al (8) suggested that supernumerary carpal bones are an invariant feature of the syndrome because it was found in 47 of their 48 patients. In their series, accessory carpal bones were described more frequently than large joint dislocations. Although the pathogenesis of this skeletal dysplasia remains unknown, supernumerary carpal bones scattered in a random fashion with bizarre deformations of their shape is a feature highly characteristic of Larsen syndrome (3,4).

We performed a literature review of studies describing cases of Larsen syndrome with supernumerary carpal bones. Six studies with a total of 68 patients were identified. Hand radiographs were unavailable for six patients. A total of 60 patients were found to have supernumerary carpal bones. This subgroup was further analyzed and the following characteristics were noted: 95.0% (57 of 60) had dysmorphic facies, 100% had at least one large joint dislocation, 89.8% (53 of 59) had clubfeet or other foot deformities, 100% (55 of 55) had spatulate thumbs, 92.3% (48 of 52) were of short stature, 79.3% (46 of 58) had scoliosis and 88% (44 of 50) had cervical spine anomalies (Table 2). From the above-mentioned features, only cervical spine anomalies and supernumerary carpal bones were found in our patient.

Accessory carpal bones are generally asymptomatic; however, they may be associated with wrist subluxations (10). All cases reviewed were managed conservatively and no surgical intervention was required. Despite not having frank joint dislocations, our patient manifested mild joint laxity at the levels of the ankles, knees and patellas bilaterally. Although not a commonly used defining feature, ligamentous joint laxity has been described by some authors as characteristic of Larsen syndrome (9). In this generalized meniscal joint disorder, ligamentous laxity may be related to large joint dislocations on a spectrum of connective tissue involvement, ranging from laxity to subluxation to dislocation.

Despite the rarity of this condition, it is important to recognize supernumerary carpal bones as an entity rarely occurring in isolation. This finding should prompt a careful evaluation and appropriate investigations for potentially serious underlying manifestations such as cervical spine abnormalities. We recommend imaging of the wrists bilaterally as well as a full skeletal survey. Referral for genetic counseling and molecular analysis is crucial for diagnosis because Larsen syndrome may present with supernumerary carpal bones as a sole clinical finding.

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