Congenital Complete Entire Carpal Fusion with Massive Carpometacarpal Coalition

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Congenital Complete Entire Carpal Fusion

Carpal coalition, sometimes referred to as carpal fusion, is a rare entity that occurs as an isolated conjunction. Many different combinations of carpal coalitions have been reported, often associated with other congenital anomalies and hereditary syndromes (1,2).

We describe an unusual case of congenital complete entire carpal coalition with massive carpometacarpal coalition except the first carpometacarpal joint and abnormal distal radioulnar joint bilaterally. According to our knowledge, this condition has not been previously reported in the literature.

Case report

A 19-year-old right-handed white man, whose both wrists had slight ulnar deviation, was admitted to emergency unit for another cause. A symmetrical range of motion was present in both wrists; 60 degrees of flexion, 60 degrees of extension, 0 degree of radial deviation, 40 degrees of ulnar deviation, 90 degrees of pronation and 70 degrees of supination. The pain, tenderness and swelling were absent at the wrist region. Family history was unremarkable.

The case was revealed on radiographic examination incidentally. Radiographs showed both congenital complete entire carpal coalition and massive carpometacarpal coalition except the first metacarpophalangeal joint bilaterally. In addition, distal radiocarpal joint surfaces were slightly irregular but compatible. There were abnormal distal radioulnar joints and ulnar styloids articulated with the ulnar sides of the lunate fossae of both wrists (Figure 1).

No other systemic or local pathology was found in the physical and radiological skeletal survey. Erythrocyte sedimentation rate, C-reactive protein and rheumatological parameters were in normal ranges.

Discussion

Congenital coalition has been described between radius and ulna, tibia and fibula, spine, skull, and the bones of the feet and hand (1). Carpal coalitions are usually asymptomatic. There are no functional motion deficits with a coalition. Recently, there have been reports of symptomatic pisohamate coalitions, scapholunatricquetral coalition, and scaphoidtrapezium synchondrosis (1,3,4).

Carpal fusion is told to be a result of failure of separation of the cartilagenous lining of the bones as early as the fifth week of intrauterin life, resulting in continuity of adjacent bones (4). The term carpal fusion is a misnomer since the anomaly represents failure of segmentation of the primitive carpal mesenchyme with absence of joint formation. The cartilage carpal elements were joined of ossific centers (5).

Coalition between most adjacent carpal bones has been documented. Less commonly, there is coalition between the carpus and the radius or between the distal carpal row and the metacarpals (6). Carpal coalition involves more than one carpal bone, which is more common when coalition occurs as part of a syndrome of congenital malformations. Carpal coalition has been reported in almost every possible combinations (1). Isolated fusion has been described in almost all of the carpal bones with triquetralunate fusion being the most frequent site (4,7,8). Next frequency is a coalition between the capitite and hamate, whereas other combinations are quite uncommon (8). Isolated coalitions usually involve two carpal bones in the same row. In contrast, multiple coalitions, which usually extend across carpal rows are associated with rare syndromes such as arthrogryposis, synphalangea, diastrophic dwarfism, Turner’s syndrome, Ellis-van Creveld syndrome (1,4).

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Carpal coalitions may be complete or incomplete between carpal bones. Incomplete coalition probably relates to abnormal differentiation of the interzone, allowing cartilaginous continuity to occur without intervening synovial tissue (7).

Massive carpal coalition is common in the Ellis-van Creveld syndrome. It is also seen in Nievrgelt’s syndrome. Such a massive carpal fusion is very rare as an isolated anomaly (4). However, it is seen bilaterally and associated with musculoskeletal anomalies especially arthrogryposis patients (9). Combined coalitions in the presence of wide spread non-skeletal abnormalities are described with the hand-foot-uterus syndrome (4). We are not aware of a previous report of massive complete carpometacarpal fusion, but capitometacarpal coalitions have been presented with other skeletal anomalies in the literature (1,2).

Although, our case was not associated with any other syndrome, it had entire carpal complete coalition, massive complete carpometacarpal fusions except the first carpometacarpal joint, irregular but compatible radiocarpal joint surfaces, and ulnar styloid jointed with incisura ulnaris of distal radius, bilaterally. Also, he had slight functional and aesthetic deficiency but no complaint of this condition. This patient’s pattern of coalition represents a unique case, being especially in a healthy individual and not as part of a syndrome of other anomalies.

References


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