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Case Report

Congenital absence of lunate and triquetrum with hypoplastic scaphoid: a case report and review of literature

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ABSTRACT

The absence or hypoplasia of bones or deformities associated with various bones have been documented in the past by various authors. These absent or hypoplastic carpal bones have mostly been associated with various congenital syndromes and anomalies. Here, a case is reported with absent lunate and triquetrum with hypoplastic scaphoid bone without any congenital anomaly or syndrome.

Keywords: Carpal absence, Hypoplastic scaphoid, Lunate absence, Triquetrum absence, Congenital

INTRODUCTION

The absence or hypoplasia of bones or deformities associated with various bones have been documented in the past by various authors. These absent or hypoplastic carpal bones have mostly been associated with various congenital syndromes and anomalies. Few of such documented syndromes are VACTERL syndrome and also similarly in Fanconi's Anaemia.[1-5] Aplasia or hypoplasia have also been demarcated according to the longitudinal defects of radial elements and limb buds. It is a rare occurrence to find out any isolated aplasia, hypoplasia or complete absence of carpal bone/s without any associated syndromes or congenital anomalies.

Here, a case is reported with absent lunate and triquetrum with hypoplastic scaphoid bone without any congenital anomaly or syndrome. To the best of our knowledge so far, this combination of absence of carpal bones has not been reported.

CASE REPORT

A 20 year old male, patient presented in outpatient department clinic, with complains of pain at right wrist since 3 months. Pain was insidious in onset, dull aching, intermittent and was increased when lifting heavy weights; relieved on rest, medication and use of supports. There was no history of trauma, infection or fever. There was no history of childhood illness. He was delivered normally at term gestation. He had no history of any Intensive Care Unit visit after birth. There was no history of any deformity/defect at birth. There was no history of any prior treatment taken for wrist pain anywhere else. This was his first hospital visit regarding this wrist pain. His parents were non consanguineous. On general appearance, he looked normal built, well nourished, head looked normocephalic without abnormal facies, local examination of right wrist revealed no scar, sinus, erythema, swelling or induration. There was no visible deformity. On deep palpation there was mild tenderness near distal lateral wrist radial margin along with mild tenderness at anatomical snuff box. Thenar and hypothenar eminence of both hands were almost equal to each other. There was no upper limb length discrepancy. There was no distal neurovascular deficit. Radiographs of bilateral wrist revealed the absence of lunate, triquetrum with hypoplasia of scaphoid and radial styloid on the right wrist while all the carpal bones were normal on the left side. Furthermore, to look for any suspicious infection or soft tissue abnormality, Magnetic Resonance Imaging of the right wrist was advised which confirmed the radiological finding and there was no

evidence of infection. All the structures appear normal apart from the absence of the carpals and a hypoplastic scaphoid and radial styloid.

Patient was advised and counselled regarding imaging findings and diagnosis. He was managed with Non-Steroidal Anti inflammatory drugs, muscle strengthening exercises and was advised using wrist support while lifting weights. Active range of motion was advised to prevent development of any stiffness.

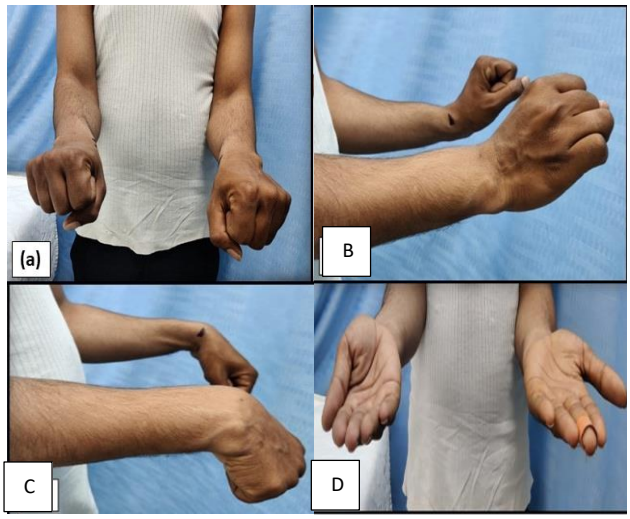

Figure 1 (A-D): Clinical images of the patient. Front view showing full pronation of B/L forearm and wrist. View showing full extension of B/L wrist. View showing full flexion of B/L wrist. View showing full supination of B/L forearm and wrist.

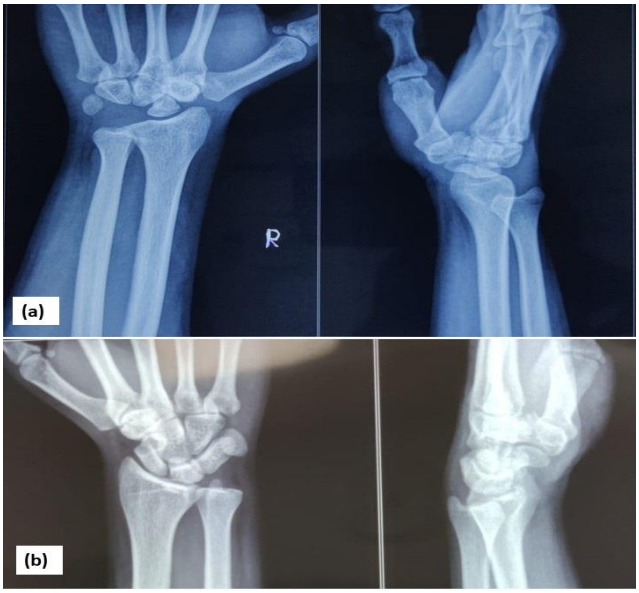

Figure 2 (A and B): Radiological images of the patient, hypoplastic scaphoid with absent lunate and triquetrum in affected right wrist alongwith radial styloid hypoplasia, normal carpal distibution of unaffected left wrist).

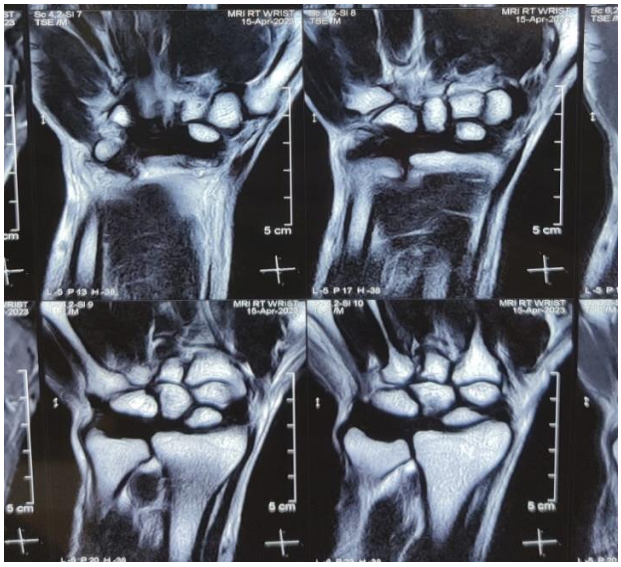

Figure 3: MRI T1 images showing hypoplastic scaphoid with absent lunate and triquetrum.

DISCUSSION

The wrist and hand are part of a complex functional unit comprising of a complex anatomy and is made up with the small, muscular ligamentous complexes held along with various bony structures to provide mechanical stability. This arrangement is helpful in providing wrist joint versatile range of motion and functions that it performs. The upper limb starts developing at around 4 weeks of life. At around 8 weeks complete outline is formed.[6,7] Thus, even before pregnancy is diagnosed, almost all the abnormalities, if any in limb are already established. Cartilaginous precursors appear at around 6 weeks of intrauterine life which undergo separation and cavitation. The first carpal bone to formed is capitate followed by scaphoid, lunate, hamate and triquetrum. The congenital abnormalities have been known to occur in less than 2% of all new-borns and out of which nearly 9-10% affect upper limb.[7]

Various upper limb deformities such as club hand, radial hemimelia, carpal arrest deformities etc. that are already documented have been classified in various classification systems. The classification system of international federation of societies for surgeries of the hand, classifies these deformities into seven groups namely failure of formation; failure of differentiation; polydactyly; overgrowth; under growth; amniotic band syndrome; generalised skeletal syndromes.[2-9]

Congenital malformations of upper limb vary from a great variety of small to large skeletal abnormalities which may affect little bit of functional status of the limb to a completely disabling malformation as well as deformity of the upper limb. Carpal absence is also seen in congenital extremity malformations such as intercalary radial hemimelia.[10,11]

Absence of carpal bones is also seen in hereditary multicentric Idiopathic osteolysis along with tarsal osteolysis which starts between the age of 2 to 7 years. It starts spontaneously with pain and swelling in hands and feet. The progression of the disease ceases in adolescence. Non-hereditary multicentric osteolysis with nephropathy is also associated with absence of carpal and tarsal bones along with malignant hypertension and renal failure.[12]

In this case report, it is proposed that structural anomaly is due to combination of failure of formation (for lunate and triquetrum) and undergrowth (for the hypoplastic scaphoid).

Majority of the authors have reported (Table 1) absence of scaphoid, as the most common carpal bone absence with or without any other bony involvement.[1,13-23] Only one report has documented bilateral scaphoid absence.[19] Most of them managed conservatively with satisfactory outcomes. Ubeda et al have reported a case of hypoplasia of scaphoid with carpal instability in which they performed arthroscopic removal of scaphoid remnant. Patient became asymptomatic after surgery.1[7] Panciera et al have reported a case of degenerative intercarpal arthritis with absent scaphoid and lunotriquetral coalition in 45 year old male manual worker. Capitate-lunate-hamate-triquetral arthrodesis performed using staples and Herbert screws.[4]

Table 1: Manuscripts published regarding absence or hypoplasia of various carpal bones.

Authors	Age (In years)/ sex	Bone involved	Symptoms/ inciting event	Treatment	Results
Kuz et al[1]	18/M	Absent scaphoid right + hypoplastic scaphoid left	Mild wrist pain after sprain	Conservative	Symptomatic with heavy activities
Radford et al[13]	15/M	B/l hypoplastic scaphoid	Paraesthesia, weakness	Surgical (Carpal tunnel release)	Good
Newcomb et al[14]	45/M	Absent lunate+ hypoplastic sacphoid left	Chronic wrist pain	Conservative	Satisfactory
Roche[15]	12/F	Absent lunate U/L	No symptoms	Conservative	Satisfactory
Mavi et al[16]	9/M	Absent scaphoid +trapezium + thumb + radius right	Short forearm and hand	Conservative	N/A
Ubeda et al[17]	53/F	Hypoplastic scaphoid bilateral	Chronic wrist pain left	Arthroscopic removal of scaphoid remnant	Satisfactory
Postacchini et al[18]	38/M	Absent scaphoid + lunate + triquetral right	Pain at wrist + shortening	Conservative	Satisfactory
Patankar[19]	28/M	Absent scaphoid+ radial styloid B/L	Post traumatic Pain Left wrist	Conservative	Satisfactory
Srivastava et al[20]	26/M	Absent scaphoid + radial styloid right	Post traumatic Pain right wrist	Conservative	Satisfactory
Schick[21]	Adult/M	Absent Scaphoid U/L	Post traumatic wrist pain	Conservative	Satisfactory
Papanikolaou et al[22]	22/M	Absent scaphoid left	Post traumatic wrist pain	Conservative	Satisfactory
Gurav et al[23]	14/M	Absent all carpals with fused metacarpals left+ absent trapezium with short 2nd metacarpal	Deformity left hand	N/A	N/A
Panciera et al[4]	45/M	Absent scaphoid	Degenerative arthritis	Capitate-lunate-hamate-triquetral arthrodesis	Satisfactory
Kobayaschi et al[24]	20 /M	Absent lunate B/L	Wrist pain	N/A	N/A
Eraltug[5]	N/A	Absent scaphoid+ capitate U/L	N/A	N/A	N/A
De Smet[26]	N/A	Absent scaphoid + lunate	N/A	N/A	N/A

It has been observed from the published literature that patients are more symptomatic when scaphoid is hypoplastic as compared to its absence. Hypoplastic scaphoid is generally associated with radial styloid hypoplasia. This makes carpal tunnel shallow. Radford et al reported a case of chronic median nerve compression in a 15 year old boy with hypoplastic scaphoid and radial styloid. Carpal tunnel release was performed and intraoperatively, they found carpal tunnel to be shallower than normal.[13] Onset of intercarpal arthritis or carpal instability seems to depend on the age and extent of manual work performed although because of very small number of patients, no conclusive evidence can be drawn. Few authors have reported isolated absence of lunate.[14,15,24]

CONCLUSION

It is concluded that this combination of absence of lunate and triquetrum with hypoplastic scaphoid without any syndrome is distinct entity and this is first documented report.

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