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CASE REPORT

A Case Arthrogryposis Multiplex Congenita with Renal Affection at Older Age

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Abstract

Report on a known case of female patient, 58-years-old, deaf since birth with skeletal deformity presenting with acute kidney injury.

These features are consistent with diagnosis of arthrogryposis multiplex congenita. A congenital disease presenting with multiple joint contractures and deformity with deafness and may be renal affection. The second point is that, up to our knowledge, we are the first authors to take into consideration that the arthrogryposis multiplex congenital may be presented with renal affection also.

Keywords

Skeletal deformity, Deafness, Acute kidney injury, Arthrogryposis multiplex congenita

Introduction

Arthrogryposis multiplex congenita (AMC) is a congenital disease characterized mainly by joint contractures. It is diagnosed mainly in prenatal period [1]. Large percent of arthrogryposis cases are caused by neurological abnormalities [2].

Treatment mainly supportive including physiotherapy and surgical orthopaedic management.

So, the aim of this case is to present a case of Arthrogryposis multiplex congenita with multiple joint contractures and renal affection.

Case Presentation

58-years-old neglected female patient, deaf since birth presented to us with confusion, vomiting and oliguria for which she was admitted to our department at Kasr Al-Ainy School of Medicine.

On presentation

She looked confused with normal blood pressure and temperature. Height = 150 cms, weight = 60 kgs, BMI = 18.7.

Musculoskeletal examination revealed non-correctable flexion deformities of distal metacarpophalangeal and metatarsophalangeal joints without swelling or tenderness. Cardiac, chest, abdominal and neurological examinations were apparently free.

Laboratory investigations revealed

Hemoglobin 8.5 g/dL (microcytic hypochromic anemia), white blood cell count 10,000/mm^3, platelet count 529/mm^3, creatinine 16 urea 280, Na 138, K 5.7, Ca 9.5 mg, arterial blood gases revealed metabolic acidosis (pH: 7.34, HCO_3: 17, PCO_2: 23), with normal liver functions. A diagnosis of acute kidney injury was made for which the patient received 3 sessions of dialysis with declining in kidney functions and improvement of her conscious level.

Radiological investigations of the patient revealed osseous ankylosis of distal metacarpal and metatarsal joints with sever angulation, diffuse osteopenia, right femoral head superior dislocation and sacroiliac joint ankylosis and marked reduction of intervertebral disc spaces as shown in Figure 1, Figure 2 and Figure 3.

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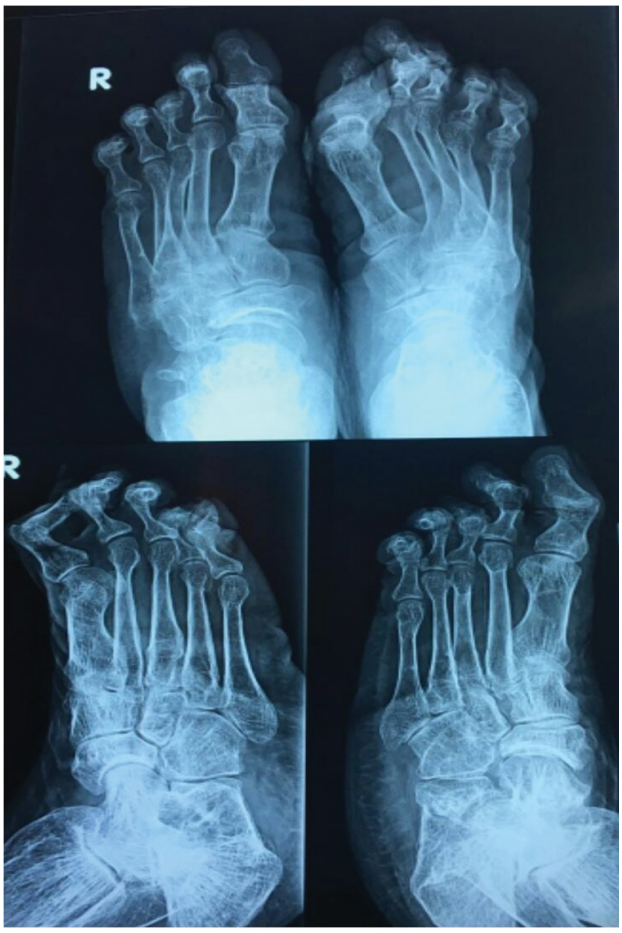

Figure 1: Plain X-ray feet, A/P view.

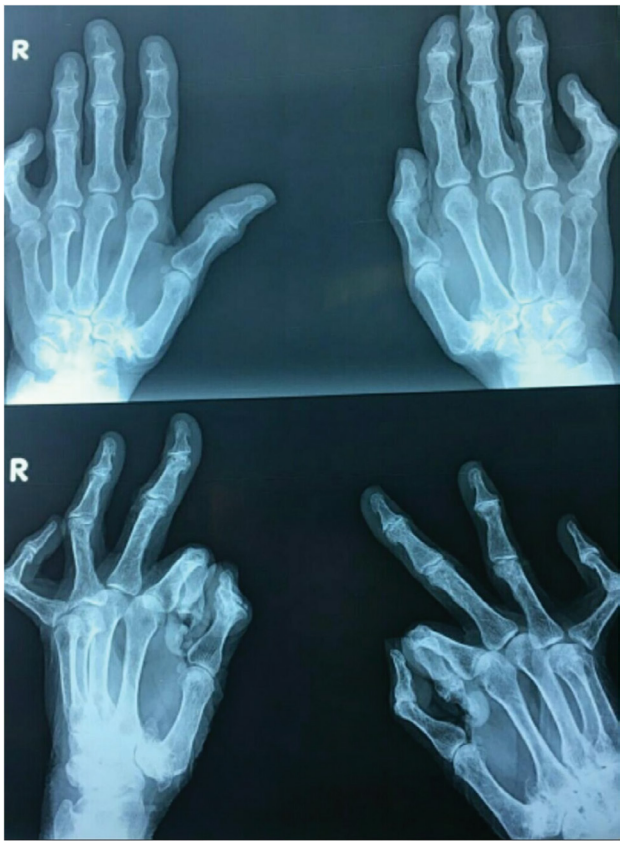

Figure 2: Plain X-ray hand, A/P view.

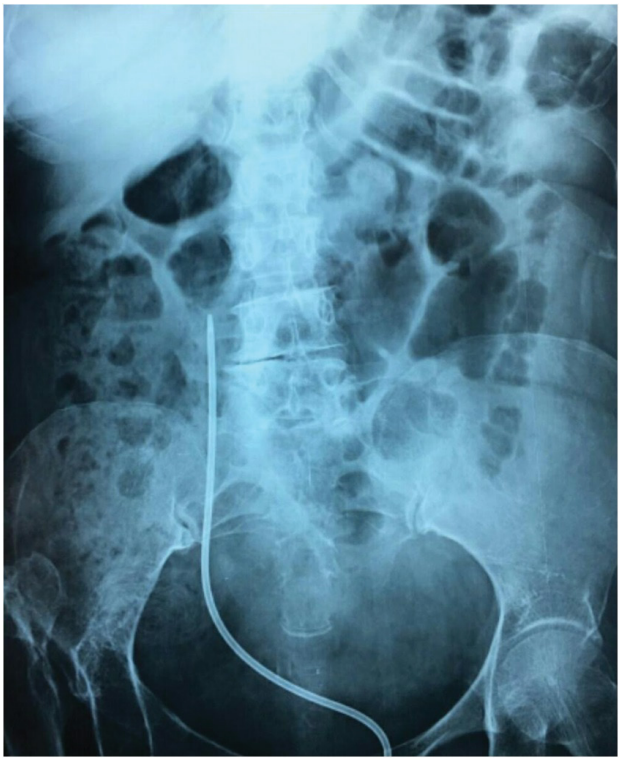

Figure 3: Plain X-ray abdomen and pelvis, A/P view.

Abdominal us revealed Left atrophic kidney while right kidney showed moderate back pressure changes for which double J catheter was inserted with improvement of the kidney functions.

The patient run a conservative course regarding her renal impairement and was sent for orthopedic correction of her contracture.

By these features we assumed that the patient has deafness arthrogryposis multiplex congenita with renal affection.

Discussion

Since 1841, the syndrome known as arthrogryposis multiplex congenita was first described by Otto which has been discussed in the orthopedic, neurologic, and pediatric literature [3].

Arthrogryposis multiplex congenital is considered as a complex of symptoms of congenital joint contractures associated with both neurogenic and myopathic disorders [4].

There are several etiological factors that can result in development of AMC; collectively any process which can limit the fetal mobility in utero can result in development of AMC [5].

The association between AMC and renal affection was described in many ways as reported by Lutz-Richner and Landolt' in 1973 who report the association between arthrogryposis multiplex congenita, cholestatic liver disease, renal tubular acidosis, and death in infancy (ARC syndrome). Since the first, four other similarly af-

fected families have been reported [6]. As regarding our case there was no cholestasis or renal tubular acidosis also our patient is presented with renal affection at old age so we can assumed our case either as atypical variant of ARC syndrome or a coincidence to presence of renal affection in an arthrogryposis multiplex congenital patient.

AMC is a description of clinical findings present rather than a specific diagnosis [7].

Conclusion

We present a case of AMC with multiple joint contractures presenting with renal affection.

Declarations

Ethics approval and consent to participate

The study was approved by institution ethical committee and form review board of Kasr Al-Ainy hospital. Oral and written informed consents were obtained from the patient or from his eligible relatives.

Consent for publication

Oral and written informed consents were obtained from the patient or from his eligible relatives.

Availability of data and material

Not applicable.

Competing interests

The authors declare no potential competing interests.

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Authors' contributions

The authors alone are responsible for the content and writing of the paper.

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