ABSTRACT

The objective of present study is to report a case of synostosis seen in a girl aged seven years who came to outpatient department of orthopaedics at Velammal Medical College Teaching Hospital, Madurai, Tamilnadu with a complaint of inability to move the left forearm in clockwise and anticlockwise direction since birth. Her daily routine activities such as toilet habit and locking the door were very difficult to carry out. She had restriction of the elbow joint in mid prone position associated with pain. She underwent surgery for her right superior radioulnar joint last year. Post operative period was eventful. There was no relevant history in the neither past except surgery of right elbow nor similar complaints in the family.

Keywords: Synostosis, Superior Radio Ulnar Joints, Interrosseous Membrane, Proximal Radio Ulnar Synostosis

INTRODUCTION

It was Sandi fort in 1793 who described congenital Radio ulnar synostosis (Lescault et al., 2000; Fakoor, 2006). Congenital radio ulnar synostosis occurs between age group of 2-5 years of age (Lescault et al., 2000). According to literature, about 350 cases of Congenital Radio ulnar synostosis have been reported (Fakoor, 2006; Farzan et al., 2002; Wurapa). It said to be a rare congenital anomaly causing very limited rotational movements of the forearm leading to difficulty in carrying out normal activities (Masuko et al., 2004; Kao et al., 2005). In 60-80% of cases it is seen bilaterally (Lescault et al., 2000; Wurapa). It runs in the family in 9% of cases (Fakoor, 2006). It is believed that this anomaly is connected with the aberrations of X chromosomes (Elliot et al., 2010). There are also reports on the cases of congenital radio ulnar synostosis due to aberrations of Y chromosomes (Syed and Quinton, 2008).

CASES

A seven old girl came to the outpatient Department of orthopaedics of Velammal Medical College Hospital & Research Institute, Madurai, Tamil Nadu with history of absence of pronation and supination movements of left elbow since birth. Elbow was fixed in midprone position. She was second child to her parents. There was first degree of consanguinity. She underwent surgery for her right superior ulnar joint last year. Later she underwent Chandra Prakasham Osteotomy procedure here at our teaching hospital. Past history- Post operative period was uneventful. Now, she is able to carry out her normal activities by her supination and pronations movements.

X-Ray of left elbow

The plain radiograph of left elbow showed bony fusion of proximal aspect of Radius and Ulna. Elbow joint appears to be normal. Rest of the visualized radio ulnar shaft & distal aspects appears normal. Visualized wrist joint, metacarpal bones & soft tissues appear to be normal.
DISCUSSION
Embryo logically the upper limb arises from the unsegmented body wall which occurs between 25 to 28 days. On 34\textsuperscript{th} day elbow appears & on 37\textsuperscript{th} day all the three bones of the upper limbs i.e., humerus, radius & ulna appear. Initially these three bones are connected to each other through their cartilaginous analogue before segmentation. Radius and ulna share common perichondrium. Any abnormal events occurs during this period may result in failure of segmentation. It is the duration and severity that determines the degree of failure of segmentation. This is followed by endochondral ossification followed by cartilaginous synostosis. This may occur either partially, or completely, in the longitudinal or transverse planes. Failure of segmentation between radius and ulna is the aetiology of synostosis (Lewis, 1901).

The congenital radio ulnar synostosis occurs between proximal radius & ulna this condition is present at birth. There is absence of pronation and supination movements Incidences in both sexes are equal and occurs bilaterally in more than 50%. Congenital radio ulnar synostosis have been classified into two types, namely Type-1 & Type-2.

\textit{In type-1}, there is proximal fusion of radius and ulna. This is complete type variety.

\textit{In type-2}, it is of less severity, where there may be partial union where fusion occurs between radius and ulna beyond proximal radial epiphysis. This condition is associated with dislocation of radial head (Wilkie, 1914). This congenital anomaly can give rise a spectra of anomalies. It can also show fusion in varying length and with or without movement of head of the radius (Simons \textit{et al.}, 1963). Congenital radio ulnar synostosis occurs due to uterine insult in association with skeletal deformity such as dislocation of hip, club foot, polydactyl syndactyly & laxity of ligaments etc (Jaffer \textit{et al.}, 1981). Very rarely there may be severe associated syndromes like Apart Syndromes, Carpenter Syndrome, Fetal Alcohol Syndrome (Jaffer \textit{et al.}, 1981).

**Present Study:** This is unilateral case of superior radio-ulnar joint synostosis found in a seven years old girl. She was second child to her parents and was daughter of first degree consanguinity. There was absence of movements of supination and pronations movements at left elbow. Movements in the other joints were present. There was no involvement of radial head. There was no other anomaly in this subject. There was no associated skeletal deformity such as dislocation of hip, club foot, polydactyl syndactyly & laxity of ligaments, thumb hypoplasia and No associated syndromes like Apart Syndromes, Fetal Alcohol Syndrome Carpenter Syndrome. Now she can carry out her normal activities.
Case Report

Take Home Message: This congenital deformity has to be managed surgically to remove bony attachments, soft tissues and for the purpose of regaining the position of forearm in order to perform her normal activities & make the proximal radio ulnar joint more functional (pronation & supination).

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