Case Report **Pediatric orthopedics**

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DECODING RADIAL CLUB HAND

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ABSTRACT

6 month old case of radial club hand reported to us with upper limb deformity, absent thumb and difficulty to extend elbow since birth. Early genetic and systemic evaluation is very much essential because associated syndromes should be treated early, like fanconi'sanaemia. Fanconi anemia needs early diagnosis which is crucial for the child and family, because it needs bone marrow transplant which is the only cure. Early diagnosis will buy time to search for suitable donor.

KEY-WORDS: Radial club hand, Genetic analysis, Systemic evaluation.

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INTRODUCTION

Normal hand articulates with two bones of forearm, the radius and the ulna at the wrist. In radial club hand, radius is not formed properly in the womb, which in turn causes the wrist to be in a fixed bent position toward the thumb side of the hand. There may be a deformity or absence of the thumb as well. The condition is called "radial dysplasia". In most forms of radial club hand, not only is there bony abnormality but also of soft tissues. [1] Radial club hand is a defect seen at birth. As with the majority of other birth defects, we are still unaware about its cause, but what we know about radial club hand indicates that it does not result from the mother's lifestyle or conditions during her pregnancy.

Case History

Here is 6 months old male baby presented with upper limb deformity and difficulty to extend elbow since birth. Mother had a normal uneventful antenatal period. Baby was born by LSCS, and cried immediately after birth. This was the first born child and only baby. Baby presented with a short left upper limb (fig 1), with absence of thumb and a stiff elbow. The left shoulder was flexed at 30 degree and elbow at 100 degree . The left forearm length was shorter with a deep crease between first and second finger. A Single palmar crease was noted with thenar and hypothenar wasting. The wrist was radially deviated with ulnar head being more prominent. Radial pulse was felt but was feeble as compared to right. The other four metacarpals and MCP joint were normal. The right upper limb and both lower limbs were normal. The baby was otherwise active with Normal Developmental milestones till date. Plain Radiogram of the left upper limb showed hypoplastic and short radius and absent thumb ray bones. Humerus, ulna and four metacarpal rays were normal. X-rays of chest, spine and pelvis were normal. Hematological values showed hemoglobin with 12 g %, platelet count at 290000 cell/ µL. Peripheral smear showed normocytic normocromic picture. Electrocardiography & Echocardiography were normal. Ultrasound abdomen for both kidneys were found to be normal.The baby was further evaluated with Chromosomal analysis (Fig 2) in our genomic lab and found to be 46 XY (male), karyotype with no numerical or structural chromosomal abnormality. This helped us in reassuring and counseling the worried parents. The baby is being managed with splinting and stretching exercises by his mother as demonstrated. The parents have also been explained about the need for later surgery (pollicisation) after which the baby will be able to hold with his left hand.

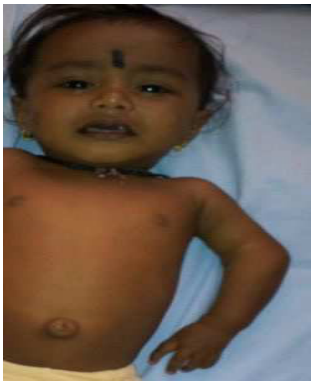

Figure 1
Showing anomaly of Left hand

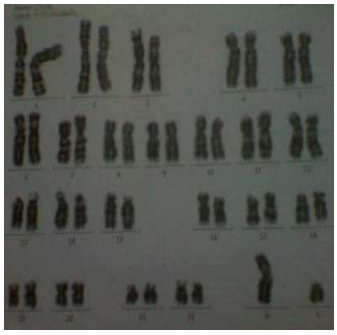

Figure 2
With normal karyotype

DISCUSSION

Radial club hand develops early in intrauterine life, sometime between the 28th and 56th day of gestation, when the bones of the hand and forearm are being formed. It is sometimes, but not always, detected on an antenatal ultrasound. Even if it is picked up antenatally, the condition cannot be treated until the baby is born. (PATHOLOGY)[2]. As the ulna grows in the mother's womb, the lack of growth in the radial anlage draws the hand into a deviated "club" position. This may also cause the ulna to bow. The degree, to which the hand and wrist deviating at birth, depends on two factors: the lack of support by radius, ulnar overgrowth to push the wrist further radially as it grows. [2, 3] The Degree of radius malformation dictates severity of function of child's radial club hand. In the most severe cases, the radius is completely absent, leading to very limited range of motion at the wrist. The elbow joint may also be disturbed or even fused with no motion which had similar findings in our case. There may also be underdevelopment or absence of the thumb, which interferes with hand function. Along with bony deformity there will be hypoplastic muscle and tendinous structure along the radial side. [4] Is it congenital and transmittable? Radial club hand occurs SPORODICALLY with no known cause. It occurs between 1 in 30,000 and 1 in 100,000 live births. Several theories have been raised, such as maternal drug exposure, compression of the uterus, and vascular injury, but none of these have been proven. Male incidence is more than female similar to other studies of comparison between sex ratio of congenital anomalies.[5] common among Caucasians and bilateral in 38-50%. When unilateral twice common on right side, some study shows that it is twice common in children born in summer than in winter. It is commonly associated with many congenital syndromes including those affecting cardiac, gastrointestinal and renal systems.

1. Holt Oram syndrome: heart anomaly (VSD, ASD, fibrillation and conduction defect) (6) - AD
2. Fanconi syndrome; associated with aplastic syndrome and fanconi anaemia– AR.
3. VATER syndrome: vertebral anamoly, anal atresia, trachea-oesophalangeal anamoly, renal Anamoly - sporadic
4. Robert syndrome associated malformation of the bones in the skull, face, arms, and legs.
5. Thrombocytopenia-Absent Radius (TAR) Syndrome -AR
6. Linked to chromosomal abnormalities - Trisomy 13, 18 and 21. [7]

What else should we keep in mind at this Diagnosis?

Radial club hand is diagnosed after inspection of the upper limb and an X-ray at birth. Check for other congenital deformities or syndromes that are associated with radial club hand, regarding the heart, kidneys, vertebral column, blood cells, and digestive system. Early genetic and systemic evaluation is very much essential because associated syndromes need to be planned

early, like fanconianaemia which will reveal after 3 years. Because bone marrow transplant is the only cure for Fanconi anemia, this prefatory diagnosis is crucial for the child and family. Early diagnosis provides ample time to search for a suitable bone marrow donor. [2, 3]

Goals

1. Correct the radial deviation of wrist.
2. Balance the wrist of forearm.
3. Maintain wrist and finger motion.
4. Promote growth of the forearm.
5. Improve the function of the extremity by following procedures.

Treatment

Splinting and exercise
lengthening of short bone
centralization of hand on single bone
replacement of other finger by rotation (pollicisation)[1,2]

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