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RESEARCH ARTICLE

CONGENITAL SWAN NECK DEFORMITY: A CASE REPORT AND REVIEW OF LITERATURE

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Abstract

Many theories have been suggested for the etiology of congenital swan neck deformity. The one main theory is that there is a lack of anatomical balance of the extrinsic and intrinsic muscles of the finger that is affected. Also, due to the imbalance between the flexors and extensors, flexion at the proximal interphalangeal joint results. As the patient gets older, there is increased lag in the extensors leading to worse flexion deformity, and this is mainly an aesthetic rather than a purely functional problem.A seven-month-old female infant presented with a hand deformity (congenital swan neck deformity on the fifth finger) that started appearing spontaneously and insidiouslywhen the infant was six months old. Operations for this type of deformity can range from skin plasties, artholysis, tenotomies, tendon transfers, and osteotomies to arthrodesis.Postoperative care consists of a splint, in which the duration in which it is kept is based on the technique used.

Introduction:-

Congenital swan neck deformity is a flexion deformity of the proximal interphalangeal joint that is not associated with trauma and is usually irreversible. It most commonly involves the ring finger. Sometimes Congenital swan neck deformity involves flexion of the metacarpal and distal interphalangeal joints, but this is usually associated with trauma. There are two types of congenital swan neck deformity, simple and complex. In the simple type, there is a deformity that involves flexion of the proximal interphalangeal joint, whereas the complex is generally associated with other deformities like polydactyl and symbrachydactyly. Congenital swan neck deformity is a rare condition that is seen in less than 1% of the population. It presents usually in two periods: infancy and puberty. The early type is congenital and affects the ring finger of both males and females. The latetype is more common and affects females who are in their adolescent years.The etiology of Congenital swan neck deformity is not well known. Since it is usually seen in ages of fast growth (infancy and adolescence) there are some theories that it is because bone growth is faster and out of proportion to palmar tissues, leading to abnormalities in skin and ultimately causing tethering of the finger. Congenital swan neck deformity may also be due to atypical lumbricals or an atypical or sometimes even absent flexor digitorum superficialis. In this case report, we shall discuss our experience with a seven month baby with congenital swan neck deformity.

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Case report:

Here we describe a case of a seven-month-old female infant that presented on 4th April 2019 to the plastic surgery clinic as an out-patient at King Fahad University hospital. The infant is medically free with normal developmental history. The only presenting complaint was her hand deformity. Her pasthistory is not significant. No past surgeries or illness. No history of communicable diseases.Regarding family history, she has a second-degree relative also diagnosed with congenital swan neck deformity. On physical examination, there was a fixed flexion deformity of her fifth proximal interphalangeal joint on her left hand with no pain or paresthesia on either hands. These changes began when the infant was six months and appeared spontaneously and insidiously. There were no other visible lesions on either hands. No indication of other hand deformities such as syndactyly or polydactyly. We could not appreciate any facial or cranial anomalies or any abnormalities of the feet.Her temperature was 37.3 degrees Celsius, respiratory rate was 16 breaths per minute, blood pressure was 132/92, heart rate was 110 beats per minute. She has taken all vaccines as required and was delivered by normal vaginal delivery.The patient's cousin developed congenital swan neck deformity on her fourth and fifth proximal interphalangeal joints on her left hand. She developed them when she was nine months old and still has them now at the age of 2 years.The laboratory evaluation showed a normal complete blood count with no immunologic antibodies. Anti-smooth muscle, anti-mitochondrial,anti-nuclear, anti-double-stranded DNA, Anti-Scl 70, anti-centromere antibodies were all negative. A muscle biopsy was done but did not show any changes. Serum creatinine, liver function tests, and urinalysis were all within normal ranges. Rheumatoid factor and HLA-B27 were both negative.A radiograph for both hands (PA and oblique) was ordered for the patient on 13th May 2019 after she had sustained a superficial injury of the wrist and hand while playing at home. There was no displaced fracture or dislocation. There was early ossification of the left distal radial epiphyses, but no soft tissue swelling.

Discussion:-

Congenital swan neck deformity is a contracture of the proximal interphalangeal joint in the anteroposterior direction. The fifth finger is affected in the majority of cases, after that comes the fourth finger. Other fingers are rarely affected (less than 10% of cases). There are different theories regarding the etiology, but the one main theory is that there is a lack of anatomical balance of the extrinsic and intrinsic muscles of the finger that is affected. Congenital swan neck deformity could be sporadic or inherited. If it is inherited (in 30% of cases) it will be autosomal dominant. It could be seen in different syndromes such as Poland syndrome and Holt-Oram syndrome.

Due to the imbalance between the flexors and extensors, flexion at the proximal interphalangeal joint results. As the patient gets older, there is increased lag in the extensors leading to worse flexion deformity. The main problem patients face isn't purely functional, it is more of an aesthetic problem that many patients complain about due to the deformity in their hands.

Congenital swan neck deformity can be seen in three different patient populations. It could be seen as a congenital anomaly in a newborn patient affecting the fifth and/or fourth fingers. Sometimes in the case of newborn infants, it could affect all fingers. The second common scenario observed is a young female between the ages of 10-19 with congenital swan neck deformity of the fifth finger that started suddenly and progresses as the patient grows. Finally, congenital swan neck deformity can be observed as part of other syndromes.

Foucher has recognized four different conditions for congenital swan neck deformity (1A) early stiff (1B) early correctable (2A) late and stiff and finally (2B) late and correctable. Several tests can be done to assess the degree of severity of congenital swan neck deformity. The first test is to assess is while the wrist is in the neutral position, an active extension of the proximal interphalangeal joint can be done? The second test involves maximum extension of the finger at the metacarpophalangeal and proximal interphalangeal joints and assessing for skin blanching. The third test involves extending both the wrist and metacarpophalangeal joints and attempting to flex the proximal interphalangeal joint. This test is also known as the superficial flexor tenodesis test. Another test done is called Smith test. This test is done while the wrist and the metacarpophalangeal joint are flexed and the proximal interphalangeal joint is passively extended. The Bouvier maneuver is done by applying pressure over the proximal pressure over the proximal phalanx resulting in the passive flexion of the MP joint causing the straightening of the distal joints and temporary correction of claw deformity. The optimal patient to be chosen for operation is one with flexible PIPJ but can not actively extend the PIPJ. Also, those based on expected compliance to hand therapy should be selected. For treatment, a splint should be applied to all patients with congenital swan neck deformity in which the PIPJ is not flexible. The splint works by stressing on the PIPJ gradually to as much extension as possible. The MCPJ is in flexion in a forearm splint. A reduction to < 30° extension lag is considered good enough to refrain from surgery. The duration

of splinting therapy before further surgical treatment varies from 3 to 12 months. If after the splinting the patient demonstratesdifficulty in active extension or movement but has improved passively then surgery can be considered.

Operations can range from skin plasties, artholysis, tenotomies, tendon transfers, and osteotomies to arthrodesis. In the correctable PIPJ, mostly skin and fibrous band release are performed, with the exploration of the lumbrical muscle, interosseous muscles, FDS, and flexor digitorum profundus. In most cases, the lumbrical is anomalous together with the FDS. If the FDS is usable and pulling on the lateral band gives extension to the PIPJ, FDS tendon transfer to the lateral band will be performed. In the non-correctable PIPJ, which improves after splinting, an extensive skin release is performed with a Malek flap which is a proximally based palmar homodigital flap. Afterward, the flexor sheath just proximal from the PIPJ is opened transversely and the checkreins, accessory ligament, and if required the palmar plate is released resulting in passive extension. The intrinsic and extrinsic muscles of the operated fingers are explored and released if an anomaly is present. An FDS tendon transfer is used from the same or adjacent finger to restore active extension. The transfer is attached to the lateral band or the central band according to which one is sufficient.

Postoperative care consists of a splint, in which the duration in which it is kept is based on the technique used. A K-wire for 2 weeks can be used for the PIPJ following the release of the joint. Compliance with the therapy is essential. In the young child, long-term splinting during the night of the stiff PIPJ results in a good outcome, with an 80-92% improvement rate after long-term splinting. On the Caucasian hand, these resulting in a good outcome can not be achieved using a similar way. Children with stiff joints do react well with splinting, resulting in an average improvement of 40° following 19 months of splinting. Splinting is usually relied on due to the variable improvement associated with the different surgeries. However, if no satisfactory improvements are achieved, surgery is performed resulting in improvement rates that range from 68 to 88%, depending upon whether or not the joint was passively correctable. Complications related to the prognosis include insufficient possibility to extend the PIPJ after surgery, osteoarthritis, recurrence of the contracture, and stiffness and pain. Secondary procedures include the re-release of capsular contracture, corrective osteotomy, or formal PIPJ arthrodesis.

Conclusion:-

Congenital swan neck deformity is a rare disease caused by flexion deformity of the proximal interphalangeal joint, most commonly affecting the fifth and/or fourth fingers, it’s seen in less than 1% of the populations, congenital swan neck deformity could be sporadic or inherited, it could be seen also in different syndromes such as Poland syndrome, and Holt-Oram syndrome, there are several testsis done to determine the severity of the deformity, the patient who suffers from congenital swan neck deformity should go for splinting therapy for 3-12 months before further surgical treatment, operations can vary, most commonly performed is skin and fibrous bands release with explorations, postoperatively patient is kept on long term splinting for a good outcome.

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