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Case Report

Symphalangism of the Digits - A Rare Congenital Condition

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ABSTRACT

Symphalangism is a rare congenital abnormality characterised by the ankylosis of the interphalangeal joints of the fingers and toes. The condition is inherited in an autosomal dominant pattern. It may involve both proximal and distal interphalangeal joints with involvement of proximal joint being most common. Compensatory hyperflexibility of the joints proximal and distal to the fused joints can occur which later can progress to arthritis. There may be other associated skeletal and non-skeletal abnormalities.

Here we present a 19 year old girl with symphalangism involving the distal interphalangeal joints of the middle and ring finger of the dominant (right) hand. She did not have any functional impairment and hence no medical or surgical intervention was performed.

As this is a very rare condition we would like to report it in our article.

Keywords: Symphalangism, fusion, interphalangeal joint.

INTRODUCTION

Symphalangism is a rare congenital abnormality characterised by the ankylosis of the interphalangeal joints of the fingers and toes. It was first reported by Benario [1] and by Sklodowski [2] in 1890. The condition of bony or fibrous ankylosis of the phalangeal joints was designated 'symphalangism' by Harvey Cushing [3] in 1916. The condition was first reported in India by Gemma Savarinathanand Willard R. Centerwall in 1966 [4]

The common associated skeletal abnormalities are brachydactyly, [5] camptodactyly, clinodactyly, syndactyly, trisomy, radio-humeral fusion, carpal and metacarpal anomalies, pes planus, bilateral hip dislocation, tarsal coalition, congenital fusion of cervical or thoracic spine, compensatory hyperflexibility of unaffected joints of the same digit. Non-skeletal anomalies include conductive hearing loss [6] and loss of skin creases over the affected joints. It may also be associated with syndromes like Poland's syndrome, Apert's syndrome, Herrmann's syndrome, Laurence-Mood-Biedel syndrome and Ellis-van Creveld syndrome.

CASE REPORT

A 19 year old lady presented to us with stiffness of the distal interphalangeal joints of the middle and ring fingers. She did not have any functional impairment and was able to write, perform her daily activities including buttoning her clothes. She had no such complaints in her family.

Her physical examination revealed that the affected fingers had loss of volar skin creases over the distal interphalangeal joints, complete loss of both active and

passive motion of the distal interphalangeal joints. There was no length discrepancy noted in the digits as compared to the contralateral hand except the affected fingers were distally tapered in appearance (Figures 1, 2).

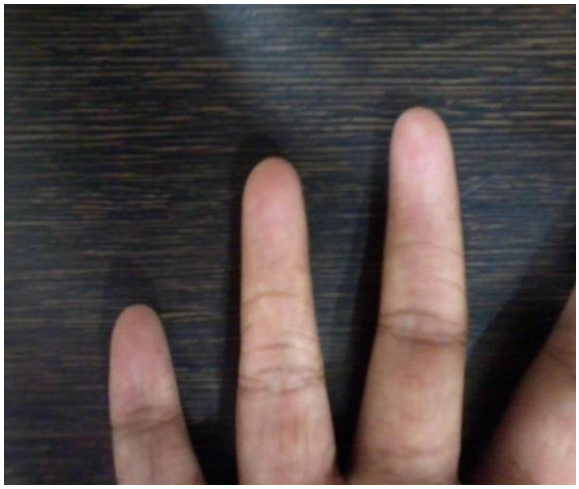

Figure 1: Absent volar crease over the distal interphalangeal joints of the middle and ring fingers

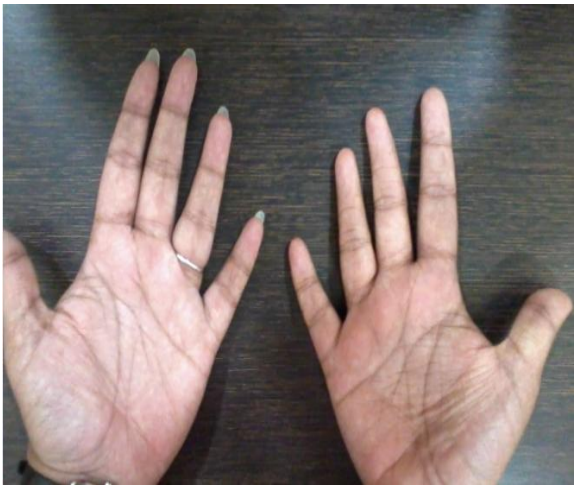

Figure 2: Comparative image showing absent volar crease in the distal interphalangeal joints of middle and ring fingers as compared to the left hand.

There was no increased flexibility of the metacarpophalangeal and proximal interphalangeal joints. All the nails were normal. The rest of the joints of all extremities including shoulder, elbow, wrist, hip, knee, ankle, and foot joints were normal. The spine was normal.

Examination of the rest of the body did not reveal any skeletal or non-skeletal abnormality.

Radiographs showed complete fusion of the distal interphalangeal joints of the middle and ring fingers. A diagnosis of Flat and Wood type of true symphalangism with Goo and Hyuk grade 3 bony symphalangism was made (Figures 3, 4).

She did not undergo any medical or surgical intervention as she did not have any deficits.

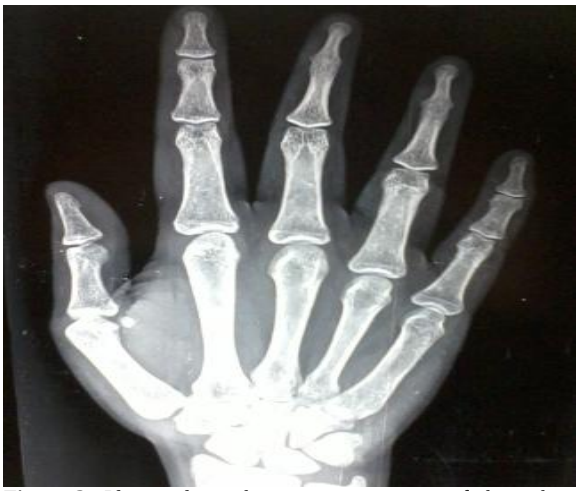

Figure 3: Plane radiograph anteroposteior view of the right hand showing bony fusion of distal interphalangeal joints of middle and ring fingers.

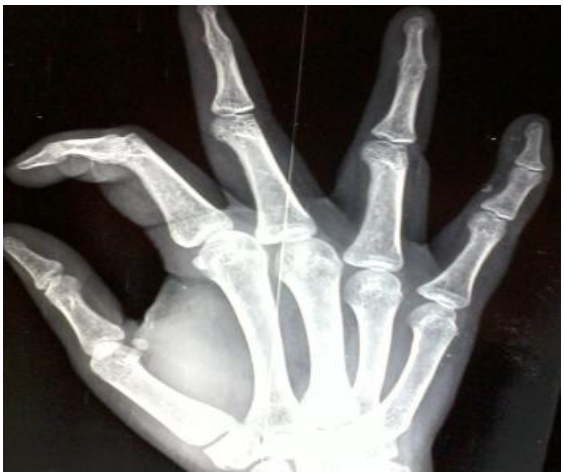

Figure 4: Plane radiograph oblique view of the right hand showing bony fusion of distal interphalangeal joints of middle and ring fingers.

DISCUSSION

Symphalangism is an autosomal dominant disorder. However drug induced symphalangism, such as thalidomide, was also reported. The non- hereditary symphalangism, often seen with symbrachydactyly, are reported as sporadic. [7]

Proximal interphalangeal joint symphalangism is more common than

distal interphalangeal joints. [8] Involvement of little finger is most common followed by ring, middle and index finger [8] Involvement of the thumb is rarer [9] However, in this patient the distal interphalangeal joints of middle and ring fingers were involved.

The longest record of transmission of symphalangism was in the Talbot family and was reported by Drinkwater. [10] Syndactyly, polydactyly with additional skeletal abnormalities may be associated as reported by Gemma Savarinathan and Willard R. Centerwall and Flatt and Wood. [11]

In 2006 Diganta Borah [12] and Sanjay Wadhwa reported proximal symphalangism in bilateral hands and feet. Flatt and Wood [11] have divided symphalangism into three main groups:

1) True symphalangism in which involved digits have normal length
2) Symphalangism in which digits are short as well as stiff
3) Symphalangism with associated anomalies such as Apert's syndrome or Poland's syndrome.

Goo Hyun Baek and Hyuk Jin Lee [7] graded symphalangism into three grades:

Grade 1: Fibrous symphalangism-mild joint space narrowing in distal interphalangeal joint.

Grade 2: Cartilaginous symphalangism-only slit of joint space is observed.

Grade 3: Bony symphalangism.

In adults, the attempts to restore mobility in the stiff joints resulted in poor outcomes. Our patient had absence of volar skin creases over the distal interphalangeal joints but did not have any evidence of osteoarthritis in the proximal joints. Also since the condition presents since birth most patients adapt to perform tasks and so do not have any functional impairment. Our patient did not have any functional impairment and so did seek any surgical intervention for restoration of movement in the affected joints.

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