A rare case of symphalangism of bilateral interphalangial joint of thumbs: a case report

Anil Kumar*

Department of Orthopaedics, Pandit Madan Mohan Malviya Hospital, Malviya Nagar, New Delhi, India

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*Correspondence:
Dr. Anil Kumar,
E-mail: dranil10051992@gmail.com

ABSTRACT

Symphalangism is rare congenital disorder characterised by ankylosis of interphalangeal joints of hands and feet. The fusion can involve the proximal or the distal joints; however, involvement of the proximal interphalangeal joints is more common. There may other associated skeletal and non-skeletal abnormalities. Here the author reports a 14 year old girl with symphalangism involving the interphalangeal joints of the thumbs of bilateral hand. She did not have any functional impairment and hence no medical or surgical intervention was performed. As involvement of thumbs is very rare condition, author would like to report it.

Keywords: Ankylosis, Interphalangeal joint, Symphalangism

INTRODUCTION

Symphalangism is a rare entity, Harvy Cushing in 1916, reported a family with hereditary fusion of the proximal interphalangeal joints and he designated this condition of bony or fibrous ankylosis of the phalangeal joints as 'symphalangism'. Inman in 1924, reported four generations of a family who had ankylosis of the distal interphalangeal joints and described two types of symphalangism i.e. distal and proximal, proximal type is more common. Both types of symphalangism have an autosomal dominant inheritance pattern. The condition was first reported in India by Gemma Savarinathan and Centerwall in 1966.

Symphalangism have been associated with other skeletal abnormalities including brachydactyly, syndactyly; camptodactyly, clinodactyly, radiohumeral fusion, carpal and metacarpal anomalies, in particular carpal synostosis, bilateral hip dislocation; tarsal coalitions, predominantly talonavicular synostosis; congenital fusion of the cervical or thoracic spine; and compensatory hyper flexibility of the unaffected joints in the affected phalanx. Non skeletal anomalies have included conductive hearing loss and absence of cutaneous creases over the affected joint.

The most frequently affected digit is the little finger, followed by the ring, long, and index fingers, respectively. Involvement of thumb and hallux is very rare. Symphalangism also has been found to be associated with Apert syndrome, Poland syndrome, Herrmann’s syndrome.

Flatt and Wood classified symphalangism into 3 syndromes: true symphalangism in which involved digits have normal length; symbrachydactylism in which digits are short as well as stiff; and symphalangism with associated anomalies, such as Apert's syndrome or Poland's syndrome.

Grading of symphalangism given by Baek et al.

- Grade I: fibrous symphalangism - mild joint space narrowing in distal interphalangeal joint.
Grade II: cartilaginous symphalangism - only slit of joint space is observed. Grade III: bony symphalangism.

CASE REPORT

A 14 year old girl presented to us with stiffness of the interphalangeal joints of the thumbs of both hands. She did not had any functional impairment and was able to write, perform her daily activities. There wasn’t any history if similar complaints in the family. On physical examination, there was loss of volar and dorsal skin creases at the interphalangeal joint of the thumb of both hands; there was no passive or active movement of the interphalangeal joint, there was no increased flexibility of the metacarpophalangeal 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 examination was normal. Examination of the rest of the body did not reveal any skeletal or non-skeletal abnormality.

Radiographs showed complete fusion of the interphalangeal joints of both thumbs and diagnosis of Flat and Wood type of true symphalangism with Goo and Hyuk grade 3 bony symphalangism was made.

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. Strasburger et al proposed that the genes for the disorder act at 8 weeks of gestation and prevent the normal development of the affected joint. Involvement of little finger is most common followed by ring, middle and index finger and involvement of thumb is rare, so here author reports a 14 year old girl with with involvement of interphalangeal joints of bilateral thumb.

One reported long-term complication associated with symphalangism is osteoarthritis in the joints proximal and distal to the fused joint because of the increased mechanical stress of the joints proximal and distal to the ankylosed joint. Most individuals with symphalangism have been observed to function well and consequently do not require treatment. The treatment options discussed in the literature included repositioning of the affected joint in a more flexed position to increase its functional ability and arthroplasty. Most treatment results have been unsatisfactory because of the poor cosmetic appearance of a finger fused in flexion or the failure of surgery to significantly increase range of motion.

Patient in thi case report did not undergo any surgical intervention and was able to do her daily activities like writing, buttoning and pinch grasp.

The functional ability of affected individuals is good, and most learn to adapt to the lack of interphalangeal flexion.

CONCLUSION

As this entity is very rare, and involvement of bilateral thumb in this condition only reported by a single author, reporting this condition would help in exploring the genetic basics of the condition and to find out new ways of treatment and rehabilitation for the patients.

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