True Symphalangism of All Bilateral Proximal Interphalangeal Joints with Multi-carpal Coalition and Hypoplasia of the Ulnar Styloid Process

Hao Yu 1 • Chongjie Li 2

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Abstract
Symphalangism is a rare genetic condition characterized by ankylosis of the proximal interphalangeal (PIP) or/and distal interphalangeal (DIP) joints. The patient presented with fused bilateral PIP joints and poor flexion, and an unsatisfactory range of motion (ROM) in the metacarpophalangeal (MP) and DIP joints. Concomitantly, multi-carpal coalition, proximal carpal malalignment, and ulnar styloid process abnormality were also observed in radiographs obtained at diagnosis. Rehabilitation training of the MP and DIP joints and a wrist supporter were recommended to achieve MP and DIP functional motion and restrict dramatic wrist motion. This is the first case report of symphalangism with multi-carpal coalition and abnormality of the ulnar styloid process to the best of our knowledge.

Keywords
Symphalangism • Ankylosis • Proximal interphalangeal • Multi-coalition • Proximal carpal malalignment • Ulnar styloid process abnormality

Case Report
A 16-year-old girl presented with a complaint of stiffness in all proximal interphalangeal (PIP) joints on both hands. The affected fingers lacked skin creases over the PIP joints. The bilateral PIP joints were locked in extension, with no active or passive movement. Flexion was abnormal for other joints in the fingers, except the thumbs. Specifically, the ranges of motion (ROMs) for both the metacarpophalangeal (MP) joints in digits 2–5 and the distal interphalangeal (DIP) joints were about 0–60° (Fig. 1). The flexion in the MP and DIP joints was M4 at most. The ROMs for other joints, including the wrist, elbow, and shoulder, were normal.

The radiographs of bilateral hands showed bony arthrosis/fusion of the PIP joints in all digits (Fig. 2). Multiple coalitions of the carpal bones, specifically coalitions of the scaphoid-trapezium, lunate-triquetrum, and trapezoid-capitate-hamate, were also observed but were asymptomatic (Fig. 3). The misalignment of the proximal carpal bone and the abnormal scapholunate joint was asymptomatic (Fig. 3). Moreover, hypoplasia of the ulnar styloid process was visible, although no ulnar-side wrist symptoms were observed. However, the patient declined further MRI examination. Systematic rehabilitation training of the MP and DIP joints was suggested to the patient to avoid dramatic wrist movement. Regular follow-up was also recommended.

Discussion
Symphalangism, which accounts for 0.03–0.04% of all congenital anomalies of the upper extremities, is a rare congenital malformation characterized by ankylosis of the interphalangeal joints of the fingers or toes [1]. Symphalangism was named by Cushing in 1916 and later described in the Talbot family by Drinkwater in 1917 [2]. Flatt [1] classified symphalangism into three syndromes: true symphalangism, in which the involved digits have a normal length; symbrachydactyly, in which the digits are short and stiff; and symphalangism with associated anomalies, such as Apert’s syndrome or Poland’s...
In 2012, Goo [3] introduced three grades for symphalangism: grade I: fibrous symphalangism, showing mild joint space narrowing in the DIP joint; grade II: cartilaginous symphalangism, in which only a slit of the joint space is observed; grade III: bony symphalangism. However, ankylosis of all bilateral PIP joints along with multi-carpal abnormality is rarely seen in individuals with symphalangism. Our literature review revealed only one other report [4] of symphalangism in all PIP joints with carpal coalition. However, our case was more complicated due to the limited ROM and powerless flexion of the MP and DIP joints accompanied by multi-coalition of the carpal bones and hypoplasia of the ulnar styloid process. Given the unusual presentation of this case, the discussion of potential interventions is essential.

The age of the patient at the time of surgery is very important. Upton [5] suggested that soft tissue release for grades I and II symphalangism of the interphalangeal joints of the hand is useful for children younger than 2 or 3 years. Several other
surgical techniques have also been reported, such as interposing arthroplasties with silicone caps [1] and silicone implant arthroplasties [6]. The outcomes of these operations are poor, and these types of operations are not applicable to growing children. Shaving of articular surfaces can fail, resulting in re-ankylosis [7]. In this case, which presented relatively late, there was limited guidance because all bilateral PIP joints showed grade III symphalangism. We also noticed that the patient preferred to put her hands in her pocket rather than showing them to the doctors and others. Accordingly, we speculated that the lack of use may be the reason for the unsatisfactory ROM and weak flexion of the DIP and MP joints. Consequently, the surgical interventions mentioned above may not be the optimal choice for this case. Instead, we suggested that systematic rehabilitation training and proper psychological counseling may be more favorable for this patient.

Asymptomatic multi-coalition and hypoplasia of the ulnar styloid process were observed in our case. The carpal condition may become symptomatic by altering the wrist’s normal biomechanics and soft tissue support through increased compensatory motion in the surrounding joints [8]. We could not visualize the internal condition of the intercarpal ligaments and triangular fibrocartilage complex since the patient refused an MRI. Given the non-clinical symptoms in our patient’s wrist, proper wrist protection and avoiding frequent load-bearing were preferable.

Fig. 2 The fused PIP joints (arrows)

Fig. 3 The coalition of the scaphoid-trapezium (small arrow). The coalition of the lunate-triquetrum (big arrow). The coalition of the trapezoid-capitate-hamate (hollow arrow). The abnormality of the ulnar styloid process (arrowhead). The misalignment of the proximal carpal bone and abnormal scapholunate joint (circle)
Declarations  The present study is a case report with ethical clearance and informed consent provided by the patient before the study.

Conflict of Interest  The authors declare no competing interests.

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