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Bilateral Camptodactyly in Twin Sisters

CASE REPORT

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ABSTRACT

Camptodactyly is defined as a permanent flexion contracture at the proximal interphalangeal (PIF) joints with mostly the fifth finger involvement. The incidence of camptodactyly is approximately 1%–2% in the general population. Camptodactyly is usually overlooked unless it causes discomfort either cosmetically or functionally. Camptodactyly is generally isolated; however, it may also accompany other anomalies or a number of very rare syndromes. Because the condition can also be mistaken for some rheumatologic diseases, such as rheumatoid arthritis, early diagnosis with a detailed musculoskeletal examination is vital for both prevention of deformity and unnecessary anxiety.

Keywords: Camptodactyly, twins, contracture

INTRODUCTION

Camptodactyly is defined as the non-traumatic, permanent flexion contracture of the proximal interphalangeal (PIF) joints of the fingers. It can be unilateral or bilateral. Fifth finger is affected the most, and other fingers and toes can also be affected (1). More than half the total number of cases are bilateral. Two types of camptodactyly are defined. The first one draws attention in the first year and affects boys and girls at an equal rate. The second type becomes prominent after the age of 10 and mostly affects girls. Both types increase in rapid growth periods and do not progress after the twenties. It displays autosomal dominant inheritance and rarely occurs sporadically; therefore, it is referred as familial camptodactyly (2, 3). Here we present a case of camptodactyly in twin sisters.

CASE REPORT

Twin sisters aged 15 years were admitted to our outpatient clinic with a complaint of deformity in the fifth finger of both the hands for approximately one year (Figure 1). The deformity was not accompanied by pain, swelling, or rash. There was no history of oral or genital ulcer, photosensitivity, Raynaud's phenomenon, or spinal pain. Systemic examinations were normal. As per the physical examination, an approximately 30-degree flexion contracture was present in the proximal interphalangeal joints of the fifth finger of both hands. In the locomotor system examination, no arthritis was detected; other joint movements were painless and unrestricted, including the spine and toes. Clinodactyly, syndactily, scoliosis, or other spinal deformities were not presented. Neurological examination was normal. Laboratory tests (erythrocyte sedimentation rate [ESR], CRP level, complete blood count, biochemical and serological tests) were within the normal range. ANA and RF were negative. On the plain radiographs, PIF joints of the fifth fingers displayed narrowing and flexion contracture; however soft tissue swelling, erosion, or cyst were not detected (Figure 2). When family history was questioned, it was found that no similar deformities existed in their families or close relatives. With these findings, stretching exercises and static splint were recommended to the patients with diagnosis of camptodactyly.

DISCUSSION

Although isolated camptodactyly is not a rare condition, it can be overlooked if it is confined to the fifth finger and does not affect other functions. Camptodactyly was first defined by Landouzy as the flexion deformity of the PIF joints of one or more fingers (4). Its prevalence in the general population is approximately 1–2% (5). It can be observed in all fingers even though the fifth finger is affected the most; however, its causes are not clear. The proximal joints of the finger and the skin, subcutaneous tissue, pulley system, tendons, and joint capsules are affected. It is assumed that it occurs when palmar soft tissues do not keep up with the growth rate of the neighboring bone tissue; therefore, it displays bimodal presentation, particularly in infancy and adolescence periods where rapid skeletal growth is observed (4-6). Two clinical types were defined: early and late onset. Early onset is present

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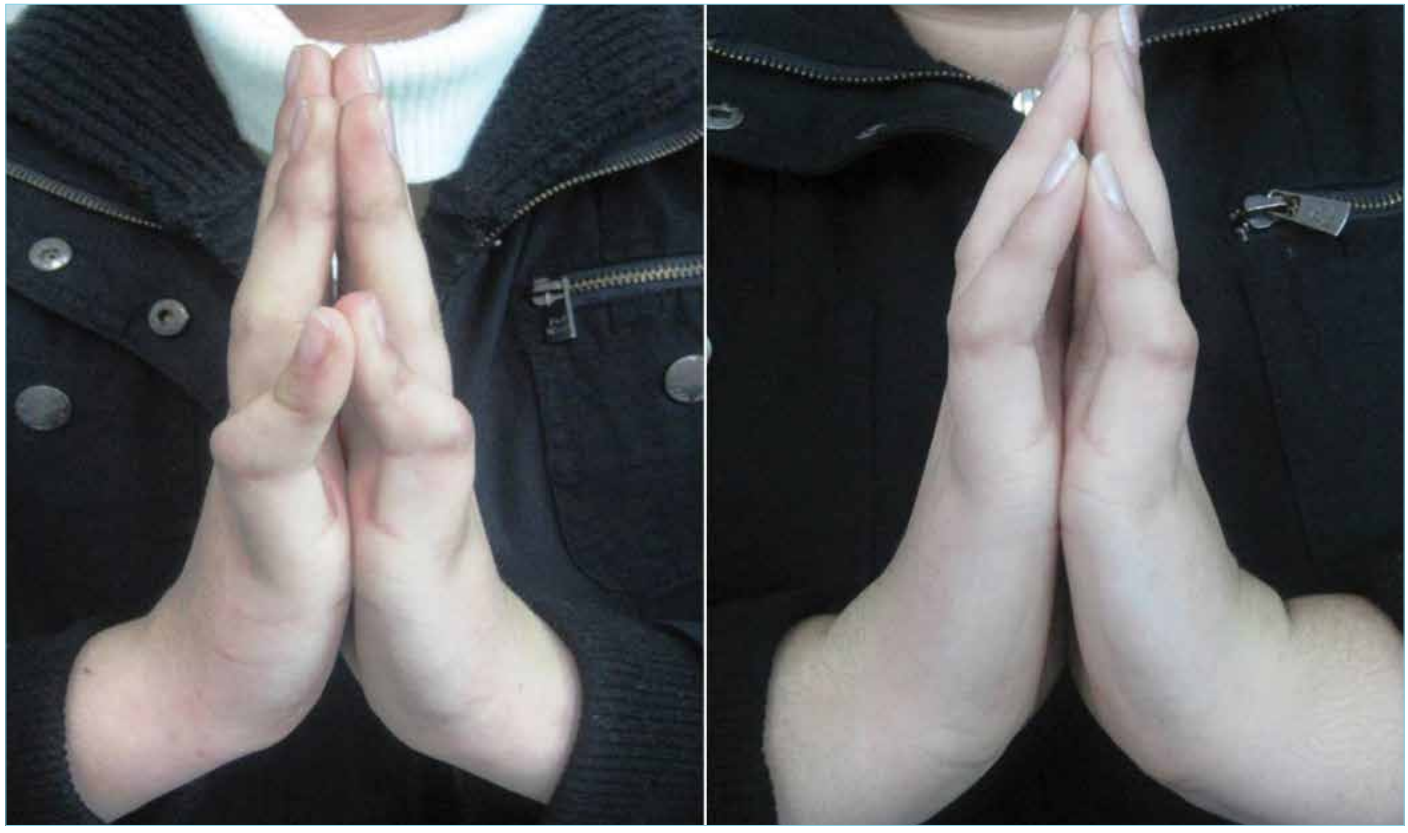

Figure 1. Extension restriction in the PIF joint of the fifth finger of both the hands

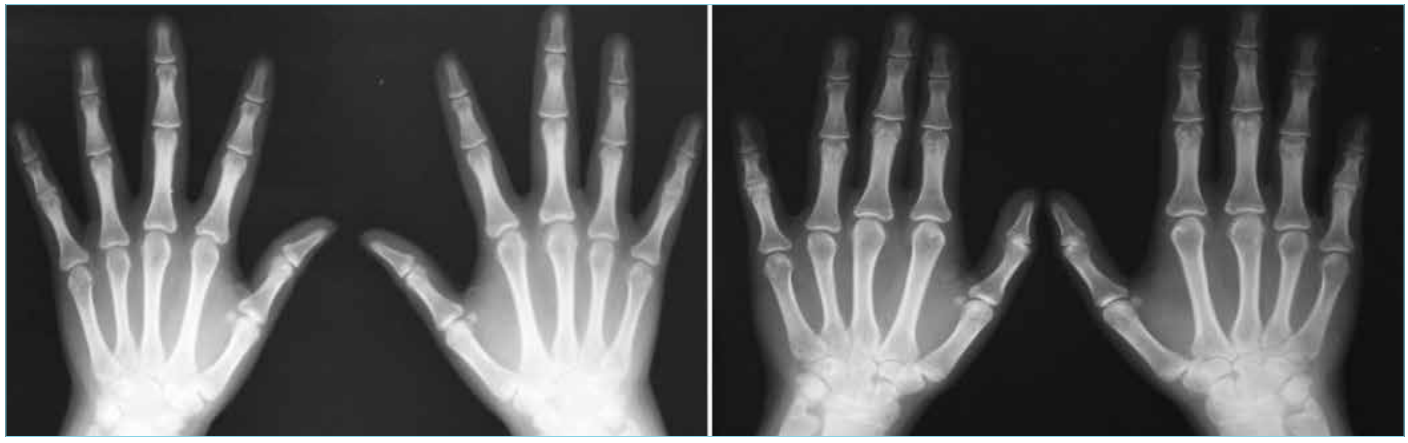

Figure 2. No pathological findings were detected in the plain radiographs of the cases apart from narrowing in the PIF joint of the fifth finger

at birth or occurs during the first year of life and usually affects both hands. It can be isolated or part of another syndrome. Late onset is observed mostly in girls after 10 years of age. In our case, the fifth finger was bilatrally affected and complaints arose in the adolescence period.

Furthermore, rheumatic and genetic diseases that may cause contracture in childhood must be excluded. Joint contractures can be observed in the oligoarticular type of juvenile chronic arthritis; however, it can easily be differentiated by the accompanying ESR, CRP levels, RF or ANA positivity, contracture, pain morning stiffness, radiological findings, and the existence of extraarticular findings such as uveitis. In our case laboratory findings were completely within the normal range and no finding of arthritis was detected. Scleroderma is an inflammatory disease that can also be observed in childhood and causes deformity in hands. This diagnosis was withdrawn owing to the absence of sclerodactyly, digital ulcer, stiffness in the skin, dysphagia, and Raynaud's phenomenon in our patients. Systemic Lupus Erythematosus (SLE) diagnosis was excluded because of deformity that was not easily reductable,, ANA negativity and absence of neurological, hematological, or nephrological findings. Contracture can be observed in the fifth finger in Dupuytren's contracture; however, this is usually accompanied by nodules or fibrous bands in the palm. No pathology was detected in the palm or other fingers of our patients. Although it usually mani-

fests sporadically, it displays autosomal dominant inheritance. It can accompany rare events such as Marfan syndyome, cranio–carpo–tarsal dystrophy, Blau syndrome, Freeman–Sheldon syndrome, Jacobsen syndrome, cerebrohepatorenal syndrome, Weaver syndrome, Jacob arthropathy–camptodactyly syndrome. It can also be observed along with anomalies such as high-arched palate, scapula anomalies, scoliosis, ptosis, hemihypertrophy. No systemic, mental, or neurological disease findings belonging to these syndromes were detected in our patients apart from camptodactyly.

In early periods, prophylactic dynamic and static splints are significantly effective in its treatment. Surgical treatment is recommended for patients with more than 60 degrees and that do not respond to the conservative treatment. Stretching exercises must accompany both treatment methods. Our patients are recommended static night splint for the fifth finger and stretching exercises.

CONCLUSION

Camptodactyly is a condition that may affect all fingers, primarily the fifth finger. Given that it causes deformity in fingers, it may be confused with various rheumatic diseases and may cause unnecessary anxiety. Exercise and methods such as splinting and/or surgical are found to be effective in its treatment. In this respect, it is important to identify the deformity, rule out other diseases, and arrange proper treatment.

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