Bilateral Carpal Tunnel Syndrome in a 9-year-old Boy With Acromicric Dysplasia

LEVENT BULUC, MD; OZGUR SELEK, MD; YASEMIN ARANAY, MD

abstract

Full article available online at Healio.com/Orthopedics. Search: 20120919-29

Acromicric dysplasia is a skeletal dysplasia that is characterized by short stature, short hands and feet, typical facial dysmorphism, normal mental development, and characteristic hand radiology. Carpal tunnel syndrome may be seen in adults with acromicric dysplasia; however, to the authors' knowledge, it has not been reported in pediatric patients. This article describes a 9-year old boy with bilateral carpal tunnel syndrome and acromicric dysplasia treated operatively. No recurrences occurred during 1 year of postoperative follow-up.

Carpal tunnel syndrome is a rare disease in childhood. The etiologic factors of carpal tunnel syndrome include trauma (especially distal radius epiphysealis), overuse, genetic or metabolic disorders, space-occupying lesions in the carpal tunnel, hemophilia, congenital anomalies, adverse effect of growth hormone replacement therapy, and idiopathic carpal tunnel syndrome. Acromicric dysplasia should be considered in the etiology of childhood carpal tunnel syndrome.

The surgical outcome of carpal tunnel syndrome is good with early diagnosis and treatment. However, in the case of skeletal dysplasia, the diagnosis of carpal tunnel syndrome may be delayed due to anomalies of the hand and due to the child's difficulty in expressing symptoms. Because of the delay in diagnosis of carpal tunnel syndrome in patients with skeletal dysplasia, the treatment outcomes may not be promising. Electrophysiologic studies should be performed early when the clinical signs are positive.

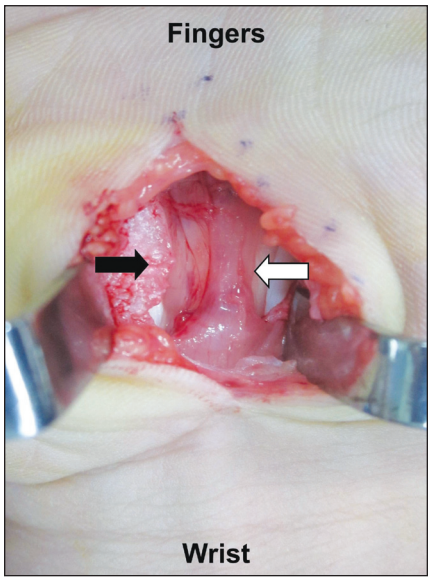


Figure: Photograph showing the color change and reduction in diameter of the right median nerve (white arrow). The black arrow shows the transverse carpal ligament.

Dr Buluc is from the Department of Orthopaedics and Traumatology, Kocaeli University School of Medicine, Dr Selek is from the Department of Orthopaedia and Traumatology, Izmit Seka State Hospital, Kocaeli; and Dr Aranay is from the Department of Pediatric Genetic Istanbul Acibadem Maslak Hospital, Istanbul, Turkey.

Drs Buluc, Selek, and Aranay have no relevant financial relationships to disclose.

Correspondence should be addressed to: Ozgur Selek, MD, Department of Orthopaedia and Traumatology, Izmit Seka State Hospital, Cukurbag Mahallesi Yusuf Sen Sokak No: 8/3, Iznit, Kocaeli, Turkey, 41300 (ozgurselek@yahoo.com).

doi: 10.3928/01477447-20120919-29

Carpal tunnel syndrome is rarely present in childhood. Carpal tunnel syndrome in childhood was initially reported by Martin and Masse.[1] They reported 3 cases with severe hand pain but could not explain the reason for this syndrome. Since then, clinical studies and case reports have shown that the most common cause of carpal tunnel syndrome in childhood is lysosomal storage disease. Skeletal dysplasia is another etiologic factor.[1]

Acromicric dysplasia is a skeletal dysplasia that is characterized by short stature, short hands and feet, typical facial dysmorphism, normal mental development, and characteristic hand radiology.[2] Maroteaux et al[3] reported 6 patients with acromicric dysplasia. Faivre et al[2] reported 22 patients with the disease from different countries. The diagnosis of acromicric dysplasia is based on the observation of short stature, short hands and feet, short and stubby metacarpals and phalanges with an internal notch on the second metacarpal and an external notch on the fifth metacarpal, and mild deformity of the femoral heads with markedly delayed bone age. Carpal tunnel syndrome may be seen in patients with acromicric dysplasia, although it is not a characteristic feature. Faivre et al[2] reported 3 patients with carpal tunnel syndrome who were aged 33, 48, and 53 years. To the current authors' knowledge, carpal tunnel syndrome accompanying acromicric displasia diagnosed in pediatric patients has not been reported in the literature.

This article describes a case of a 9-year-old boy with bilateral carpal tunnel syndrome treated operatively with no recurrence during 1 year of postoperative follow-up.

Case Report

A 9-year-old boy presented with nocturnal pain in both hands. He was delivered by caesarean section at 39.5 weeks, with a birth weight of 3700 g and length of 50 cm. No postnatal complications occurred. However, he was unable to make a fist with either hand in infancy.

At age 8 years, his height was 114 cm (less than 3 percentile) with disproportionate short stature, and his estimated bone age was 5 years. He had normal motor and mental development and no cardiac anomalies. Pubertal examination demonstrated Tanner stage 1. He had typical facial dysmorphism, and ophthalmologic evaluation revealed the eye refraction defect myopia, which was treated with eye glasses. The diagnosis of acromicric dysplasia was confirmed after endocrinologic and genetic investigation. Bilateral upper-extremity electromyography showed reduction in median nerve sensory response, and the patient was treated with wrist bracing. After that, nocturnal pain and tingling increased. Physical examination revealed thenar muscle atrophy and mild restriction of flexion in the finger proximal and distal interphalangeal joints. Tinel's and Phalen's signs could not be evaluated because the patient could not cooperate. Hand radiographs showed symphalangism and an internal notch on the second metacarpal and an external notch on the fifth metacarpal, which are typical signs of acromicric dysplasia (Figures 1, 2). Repeat electromyography revealed loss of sensory response and reduced compound muscle action potential of the bilateral median nerve.

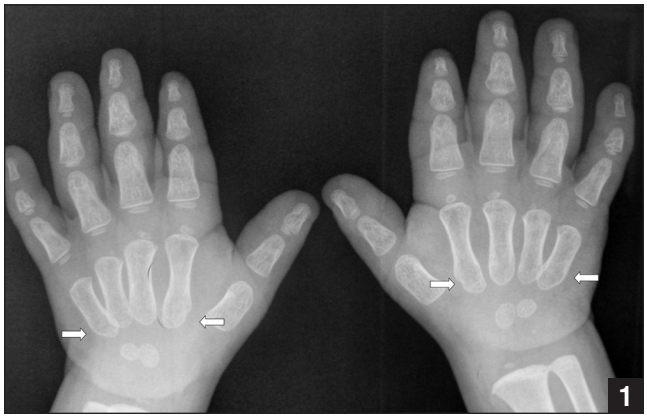


Figure 1: Anteroposterior hand radiograph of the patient at age 4 years showing metacarpal notching (arrows).

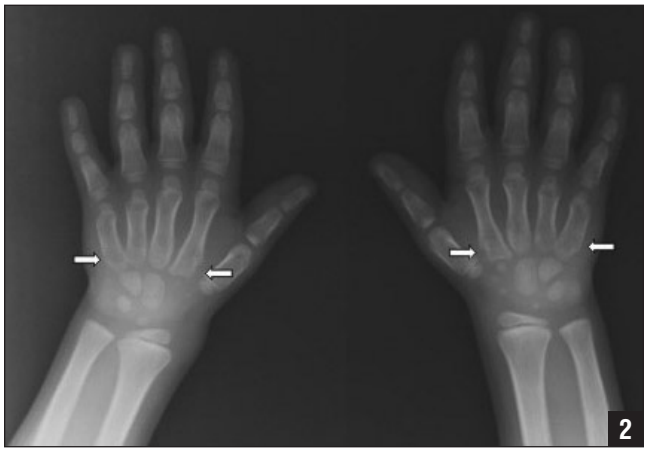


Figure 2: Anteroposterior hand radiograph of the patient at age 9 years showing metacarpal notching (arrows).

Bilateral carpal tunnel decompression was performed under general anesthesia. A classic mini-open incision was made. Exploration showed thickening of the transverse carpal ligament and change in the color of the median nerve. The diameter of the nerve was significantly decreased bilaterally (Figures 3, 4). No postoperative complications occurred. Nocturnal pain disappeared immediately postoperatively. The patient was followed with no bracing or limitation. At the end of the first postoperative year, the patient was asymptomatic.

Discussion

Carpal tunnel syndrome is a rare disease in childhood. The etiologic factors of carpal tunnel syndrome include trauma (especially distal radius epiphysealis),

overuse, genetic or metabolic disorders, space-occupying lesions in the carpal tunnel, hemophilia, congenital anomalies, adverse effect of growth hormone replacement therapy, and idiopathic carpal tunnel syndrome.[4,5]

Carpal tunnel syndrome has an unusual clinical presentation in childhood. According to Haddad et al,[6] the classic symptoms of numbness, tingling, and nocturnal pain are rarely present in children. However, signs like decreased sweating, thenar muscle atrophy, and manual clumsiness are more common. Tinel's and Phalen's signs are negative in childhood but are important physical examination signs in adults.[6] Although the gold standard diagnostic test of carpal tunnel syndrome is electrophysiological studies, pediatric patients may not comply with tests.[7]

The treatment options of carpal tunnel syndrome in childhood are the same as in adults. Initial treatment includes nonsteroidal anti-inflammatory drugs, limitation of activity, and wrist splints. If symptoms persist, surgery is indicated.[8]

Some childhood carpal tunnel syndrome cases have been reported in the literature with mucopolysaccharidosis.[6] To the current authors' knowledge, childhood carpal tunnel syndrome accompanying acromicric dysplasia or similar diseases have not been reported in literature. Acromicric dysplasia has similar features with Moore-Federman syndrome and geleophysic dysplasia; however, it has been reported as a distinct entity because it has some differences; thickened skin, hepatomegaly and cardiac valve dysplasia are not found in acromicric dysplasia. The prognosis of acromicric dysplasia is better than that of geleophysic dysplasia.[2]

Carpal tunnel syndrome accompanying acromicric dysplasia has been described by Faivre et al,[2] who reported the largest number of patients (22) with acromicric dysplasia.[2] Faivre et al[2] diagnosed 3 patients with carpal tunnel syndrome accompanying acromicric dysplasia, but none were pediatric patients. Acromicric dysplasia should be considered in the etiology of childhood carpal tunnel syndrome.

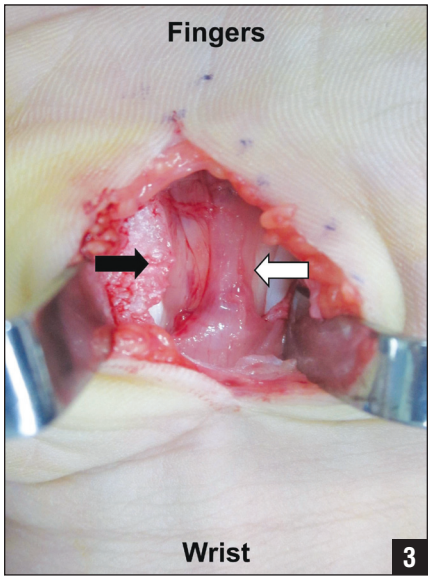


Figure 3: Photograph showing the color change and reduction in diameter of the right median nerve (white arrow). The black arrow shows the transverse carpal ligament.

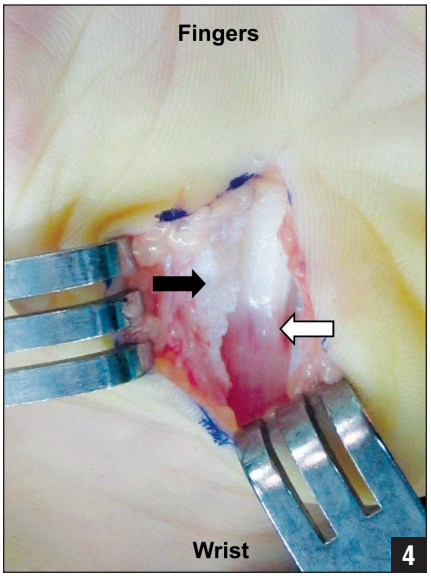


Figure 4: Photograph showing the color change and reduction in diameter of left median nerve (white arrow). The black arrow shows the transverse carpal ligament.

Intraoperatively in the current case, thickening of the transverse carpal ligament was seen. In patients with skeletal dysplasia, the pathophysiologic mechanism underlying carpal tunnel syndrome may be narrowing and shallowing of the carpal tunnel in addition to thickening of the transverse carpal ligament.[4]

CONCLUSION

In the presence of skeletal dysplasia, the diagnosis of carpal tunnel syndrome may be delayed due to anomalies of the bones and joints in the hands. Another difficulty may be the child's inadequate expression of the symptoms. The surgical outcome of carpal tunnel syndrome is good with early diagnosis and treatment. Because of the delay in the diagnosis of carpal tunnel syndrome in patients with skeletal dysplasia, the treatment outcomes may not be promising. Electrophysiologic studies should be performed early when the clinical signs are positive.

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