

Double crushing weakness of the right hand: A case report with coexistence melorheostosis and cerebral palsy

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ABSTRACT

Herein, we reported a case of right median nerve injury due to concurrent melorheostosis in an 11-year-old male patient followed for right hemiplegic cerebral palsy (CP). The patient was diagnosed with CP at 17 months of age due to weakness on the right side. During follow-up, melorheostosis, a bone dysplasia, was detected on radiographs taken due to the progressive contractures in the right hand. When the patient was four years old, electromyography was performed due to complaints of progressive weakness and pain in the right hand, and total axonal damage was detected in the median nerve. The patient's follow-ups were interrupted due to the coronavirus disease 2019 (COVID-19) pandemic. Although the pain decreased with splint and exercise treatments, functional progress could not be achieved. The patient was referred to hand surgery. However, the patient, who was not considered for surgery, continued to be managed with splint and exercise. Diagnosis of nerve injury due to melorheostosis can be difficult in an extremity affected by CP, as the motor deficits associated with CP can mask or complicate the recognition of additional nerve damage. Electrodiagnostic and ultrasonographic evaluations may be required in addition to physical examination. In cases of CP where new symptoms and motor skill loss develop, close monitoring and appropriate differential diagnosis are important.

Keywords: Bone dysplasia, cerebral palsy, median nerve, melorheostosis, nerve injury.

Melorheostosis was first described by Leri and Joanny^[1] and is also known as Leri disease. It is a rare benign sclerosing bone dysplasia that usually affects the long bones of the extremities.^[1,2] Its prevalence is approximately 0.9 per 1,000,000.^[3] Melorheostosis typically presents with musculoskeletal symptoms, including pain, joint movement restriction, contractures, and bone deformities. In some cases, it can lead to limb length discrepancies, myositis, and muscle atrophy, particularly in childhood. It progresses rapidly in childhood and slowly in adulthood. It can occur in a single or multiple extremities, with single or multiple bones affected.^[4,5] It progresses rapidly in childhood and slowly in adulthood. It can occur in a single or multiple extremities, with single or multiple bones affected.^[4,5]

Diagnosis is based on typical radiological findings. The classic pattern is a dripping

wax/flowing candle appearance characterized by linear dense areas extending from the cortex to the cancellous bone and hyperostosis with increased thickness in the cortical bone. However, osteoma-like, myositis ossificans-like, osteopathia striata-like, and mixed patterns have also been reported in various cases.^[4,5]

In this case report, a patient with median nerve injury due to melorheostosis in an extremity affected by cerebral palsy (CP) was discussed, and the literature on melorheostosis was reviewed.

CASE REPORT

An 11-year-old male patient who was being followed for CP applied to the clinic due to increasing complaints of weakness in the right hand, difficulty in grasping, and pain. The patient's prenatal and

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postnatal history was questioned. The patient was born at term by cesarean section with normal birth weight. There was no postnatal intensive care history, and the parents were not consanguineous. Developmental milestones were within normal limits until the age of one. According to the family's statement, when the patient was 17 months old, he gradually began to use his right hand less and had balance problems while walking. The patient was diagnosed with CP based on clinical findings, which was also supported by cranial magnetic resonance imaging, revealing gliosis in the posterior horns of the thalamus and the third ventricle. Extension limitation in the right-hand fingers and positional anomalies in the left toes were detected. The patient was included in the rehabilitation program.

When the patient was three years old, sclerotic areas were observed in the epiphyses in the radiographs of both lower extremities and the right upper extremity (Figure 1). Magnetic resonance imaging of the right upper extremity revealed widespread subcortical and medullary sclerotic areas. In addition, the median nerve was significantly thin along the forearm, distal to the carpal tunnel. Considering bone dysplasia, genetic evaluation was recommended for osteopoikilocytosis and melorheostosis.

At the age of four, the patient developed complaints of weakness, difficulty in grasping, and pain in the right hand. While there was a

regression in the functions of the right upper extremity, no regression in the muscle strength of the lower extremities was detected. Therefore, an electrodiagnostic evaluation was performed for the differential diagnosis of the weakness in the right hand. Electromyography at age four revealed total axonal damage in the right median nerve, likely due to compression from the melorheostotic changes. In addition, equinus deformity due to melorheostosis began to develop in the patient's left lower extremity. When the patient was eight years old, botulinum toxin was applied to the plantar flexors due to limited dorsiflexion. Since relief could not be achieved with botulinum toxin, Achilles tenotomy was later performed. When the patient was 10 years old, distal femur osteotomy was performed due to length differences in both lower extremities (Figure 2). During this entire process, the follow-ups and the exercise program were disrupted due to the coronavirus disease 2019 (COVID-19) pandemic.

The patient applied to our outpatient clinic at the age of 11 due to difficulty in using his right hand (Figure 3). On physical examination, the range of motion (ROM) of the right elbow was full in flexion and extension. However, significant weakness was noted in the right hand, with diminished grip strength and inability to perform tasks requiring fine motor skills. Forearm supination was 20° and pronation was 70°. In wrist ROM evaluation, extension was 5°, flexion was 50°, and the wrist

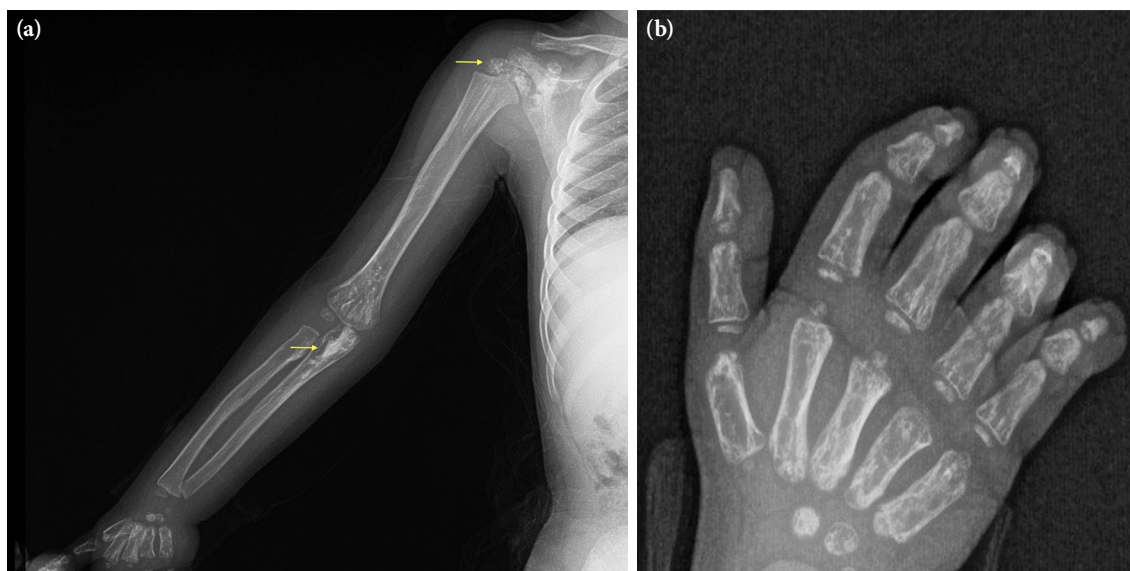


Figure 1. (a) Right upper extremity radiographs. Yellow arrows indicate lesions. (b) Right hand radiographs.



Figure 2. (a) Preoperative radiographs of the lower extremities. (b) Postoperative bilateral femur radiographs. (c) Postoperative bilateral cruris radiographs.

was ankylosed in 40° ulnar deviation. There was limitation in passive finger ROM. In addition, there were scleroderma-like stiff and tense skin changes around the right hand and wrist. In laboratory function tests, calcium, phosphorus, parathyroid hormone, 25-hydroxyvitamin D, kidney function, and thyroid function levels were within normal limits. Direct radiographs requested from the right wrist and hand showed melorheostosis-like changes, but it had an atypical appearance, as it was the hand with CP involvement. A splint was made for the right

wrist and fingers, and the patient was enrolled in a rehabilitation program. While there was a decrease in pain complaints, no improvement was achieved in functionality and deformities. Therefore, the patient was referred to hand surgery, but after further evaluation, surgery was not deemed appropriate at this time. At the last follow-up, the patient was managed with a splint and exercise program, with plans for reassessment in the future. Written informed consent was obtained from the parents of the patient.

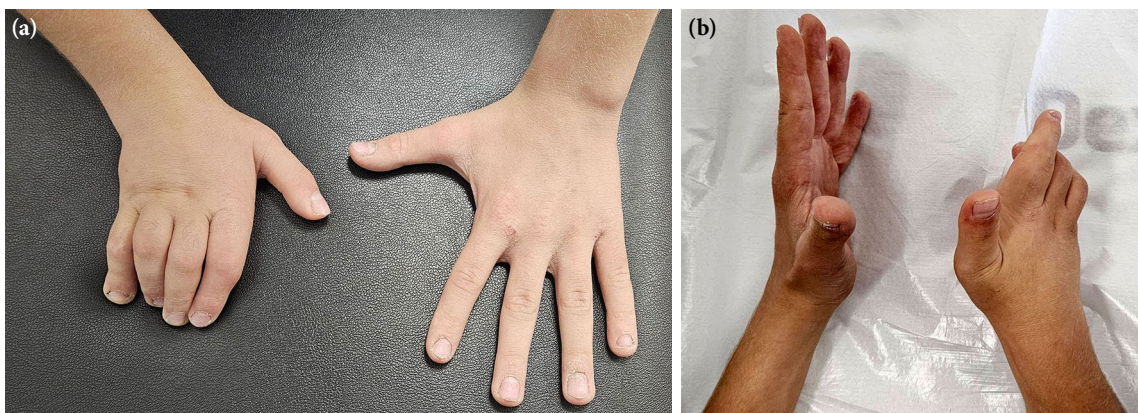


Figure 3. (a, b) Images demonstrating contracture and deformity in the right hand.

DISCUSSION

This case presented a rare form of melorheostosis, as it involved the right upper extremity, which was also affected by CP. This co-occurrence of melorheostosis and CP leading to a double-crush neuropathy has not been widely reported in the literature, making this case a valuable contribution to the understanding of these conditions. It is also quite difficult to notice the accompanying median nerve damage, particularly in the extremity with CP involvement. Therefore, our case report is a rare melorheostosis presentation.

More than 400 cases of melorheostosis have been described in the literature, but very few of them are related to the involvement of the hand bones.^[2] In a publication consisting of four cases with upper extremity involvement, Kumar et al.^[6] reported that one of the patients was followed for filariasis due to swelling, two cases presented with finger deformities, and in the remaining case, the condition was incidentally noticed in posttraumatic imaging. Therefore, melorheostosis cases may present with different clinical presentations. There are case reports describing hand involvement in melorheostosis in the pediatric population.^[4,6-8] This involvement may present with different clinical appearances, ranging from a near-normal appearance to total loss of function.^[4]

Melorheostosis usually affects the appendicular skeleton and the long bones of the lower extremities.^[2] There is a case report in the literature of idiopathic equinovarus concomitant with melorheostosis.^[9] In our case, an Achilles tenotomy was performed for equines deformity in the left foot. In addition, the patient had also undergone distal femur osteotomy surgery due to the involvement of the epiphyseal plate. Therefore, our case demonstrated different presentations of melorheostosis.

Melorheostosis affects the musculoskeletal system, and accompanying nerve damage may also be observed. Any bone lesion may cause peripheral nerve compression in the fibular, carpal, cubital, and tarsal tunnels.^[10] There are case reports indicating that the osteosclerotic structure around the hip affects the sciatic nerve and that the hyperostotic mass on the lateral side of the knee joint causes common peroneal nerve compression.^[11,12] In a case report published in 2023, bilateral carpal tunnel syndrome was detected in a pediatric case diagnosed with melorheostosis.

^[13] In our case, there was right median nerve involvement. In areas where the bone and nerve are in proximity, nerve damage due to melorheostosis should be taken into consideration.

In most cases, conservative treatments such as exercise, manipulation, and splint application are applied in the management of the disease. In necessary cases, surgical methods such as tenotomy and fasciotomies, nerve transfer, epiphysiodesis, arthrodesis, osteotomies, and even amputations can be used.^[14,15] In our case, during the follow-up after the left Achilles tenotomy, an osteotomy operation was performed from the left distal femur, as a length difference developed between the lower extremities due to growth asymmetry. The patient is also expected to undergo an operation on the median nerve in the future.

In conclusion, early diagnosis, multidisciplinary approach, timely application of conservative and surgical treatments, and regular follow-up are important in the diagnosis and treatment of melorheostosis to ensure that the patient continues their life with a higher quality of life. It is crucial for clinicians to monitor for new findings that may develop in the affected extremity, particularly in patients with CP. Early intervention and regular follow-up are essential to prevent complications and ensure optimal function.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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