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Successful Elbow Contracture Release Secondary to Melorheostosis

A Case Report

By Hyun Sik Gong, MD, PhD, Kyung Hak Lee, MD, Joo Han Oh, MD, PhD, Jin-Haeng Chung, MD, PhD, Goo Hyun Baek, MD, PhD, and Moon Sang Chung, MD, PhD

Investigation performed at the Departments of Orthopedic Surgery and Pathology, Seoul National University Bundang Hospital, Seongnam, South Korea

Melorheostosis is a rare, noninheritable developmental dysplasia of cortical bone that is often characterized by a classic radiographic pattern of flowing hyperostosis along the cortex of long bones. Patients with this condition may have joint pain, stiffness, deformity, and restricted range of motion due to soft-tissue contracture. Involvement usually follows a sclerotomal distribution and usually affects only one extremity. Joint contracture occurs when the abnormal ossification in the cortex of the long bone involves the soft tissues and extends into the joint, resulting in soft-tissue fibrosis and contracture. There is no specific treatment for this condition; options range from nonsurgical management (e.g.,...
splatning and early training in making optimal use of the un-
affected extremity) to various types of surgical management
(e.g., tendon lengthening, sympathectomy, implant arthro-
plasty, or even amputation). The literature indicates that sur-
gical release of a joint contracture may be difficult and that
recurrence of the deformity is frequent.

Melorheostosis of the elbow is rare compared with that
of the lower extremity, and a thorough search of the literature
revealed no reports on surgical release of a stiff elbow caused by
this condition. We report the case of a thirty-seven-year-old
man who had a fixed flexion contracture of the elbow as a
result of isolated melorheostotic involvement of the proximal
part of the ulna and who obtained a functional range of mo-
tion after open release of the elbow. The patient was informed
that data concerning the case would be submitted for publi-
cation, and he consented.

Case Report

A thirty-seven-year-old man presented with limitation of
motion of the left, nondominant elbow. He reported that
pain and limited elbow motion had developed after he played a
game of squash five years previously. There was no history of
elbow trauma or infection. The pain had decreased with time,
but limitation of elbow motion had continued to increase and
had resulted in a fixed flexion deformity of 90° two years be-
fore the patient presented to us. Physical examination revealed
no decrease in the range of supination and pronation of the left
forearm when compared with that of the right. The patient had
no ulnar nerve symptoms; sensibility in the ulnar nerve dis-
tribution was normal, and there was no intrinsic atrophy or
clawing of the hand.

Plain radiographs of the affected elbow showed cortical
hyperostosis of the proximal part of the ulna and speckled
calcification of the distal insertion of the triceps (Fig. 1). A
computed tomographic scan revealed nodularity of the peri-
osteal bone formation and the presence of a flowing candle-
wax pattern on a short segment of bone (Fig. 2). Magnetic
resonance imaging demonstrated low signal intensity in all
sequences (a classic sign of bone changes) and synovial in-
flammation without joint destruction or muscle atrophy.
Fig. 3
T2-weighted sagittal magnetic resonance image showing areas of low signal intensity, indicating bone changes (long arrows). The articular cartilage (arrowheads) is intact.

Fig. 4
Histologic examination revealed irregular, dense, hyperostotic, cortical bone trabeculae of varying thicknesses. The histologic features are consistent with melorheostosis (hematoxylin and eosin, ×100).
A diagnosis of melorheostosis of the proximal part of the ulna was made on the basis of the findings from these imaging studies.

Because splinting and physical therapy had proven to be ineffective and because the patient had severe limitation of functional use of the extremity, we recommended surgical release of the contracture.

The procedure was performed through a medial approach to the elbow. The ulnar nerve was identified and mobilized. There were no adhesions along the ulnar nerve. With care taken to preserve the anterior oblique bundle of the medial collateral ligament, we resected all medial fibrotic, contracted capsuloligamentous structures. At this point, passive range of motion of the elbow was from 20° of flexion to 110° of flexion. The dissection was extended posteriorly through the interval between the triceps and the posterior aspect of the humerus. Contracted fibrous tissue was removed along with the calcification in the area of the triceps insertion. There was synovial thickening in the olecranon fossa, but the osseous contour was intact. We obtained 140° of elbow flexion without further release of the triceps fascia or muscle. Because extension was still limited, an anterior release was performed. The flexor-pronator muscle mass was found to be contracted, therefore necessitating release of the common origin of these muscles by Z-plasty. To achieve full extension, the brachialis muscle was elevated from the anterior aspect of the humerus and the anterior part of the elbow capsule was resected under direct visualization. After hemostasis was obtained, the flexor-pronator muscle was repaired in a lengthened position and the ulnar nerve was transposed subcutaneously. The wound was closed over suction drains, and the elbow was immobilized in an extended position.

The hyperostotic cortical lesion of the proximal part of the ulna was partially excised for tissue diagnosis during the operation. Histologic examination of the tissue revealed dense bone, which was consistent with the diagnosis of melorheostosis (Fig. 4).

Three days postoperatively, the elbow was mobilized with use of a continuous-passive-motion machine. The anterior cubital area developed a large bulla, which stabilized and resolved. The patient was discharged seven days postoperatively with a passive range of motion from 20° of flexion to 100° of flexion. Under the supervision of a therapist, gentle range of motion was encouraged for the next six weeks. An extension splint was worn at night during this time. Two years postoperatively, the range of motion was 20° of flexion to 135° of flexion and there was no sign of recurrence of the contracture (Fig. 5).

**Discussion**

Melorheostosis involving the upper extremity is not as common as that involving the lower extremity. Most reported cases involving the upper extremity are focused on the hand or on the sclerotomal distribution pattern. This report describes the case of a patient who had melorheostosis with isolated involvement of the proximal part of the
ulna and an associated fixed 90° elbow contracture. Following contracture release, there was substantial improvement in the arc of elbow motion. The diagnosis of our patient was made on the basis of radiographic findings that showed hyperostosis along one side of the ulnar cortex. The nodularity of periosteal bone formation visualized on the three-dimensional computed tomography scan was unlike the structured laminated bone that is seen with myositis ossificans. Freyschmidt reviewed twenty-three cases of melorheostosis and described different radiographic patterns besides the classic one, which has been compared to dripping or flowing candle wax on the surface of the bone. The most frequent radiographic pattern described by Freyschmidt was “osteoma-like,” wherein the hyperostosis is located on the endosteal surface. Our patient had both endosteal hyperostosis on the posterior inner cortex of the ulna and nodular periosteal bone formation. A periosteal pattern of hyperostosis oriented in the long axis of the bone (as seen in our patient) is usually seen in adults, whereas an endosteal pattern of hyperostosis marked by streakiness of the long bones and spotting of the small bones prevails in children. The biopsy specimen obtained from our patient revealed dense benign bone that was consistent with melorheostosis and did not show features of myositis ossificans or parosteal osteosarcoma, both of which should be considered in the differential diagnosis.

Although bone scintigraphy was not acquired in the case of our patient, it may be useful in confirming the diagnosis when other tests are equivocal or for the purpose of ruling out other types of asymptomatic sclerosing dysplasia, such as pyknodysostosis, osteopoikilosis, and osteopathia striata. The magnetic resonance imaging scan did not demonstrate ligamentous or capsular calcification in our patient; however, it was effective in demonstrating a normal elbow joint and no atrophy of the muscles preoperatively. Judkiewicz et al. reported that intra-articular extension of melorheostosis occurred in 35% of patients and that this finding may be associated with mineralization of the articular cartilage or with mechanical cartilage damage. It is also recognized that the longer that intervention is delayed, the more contracted the muscles become, and that the articular cartilage may also degenerate.

Surgical treatment for this condition includes tendon lengthening, excision of fibrous and osseous tissue, fasciectomy, capsulotomy, sympathectomy, corrective osteotomy, Ilizarov lengthening, arthrodesis, implant arthroplasty, and even amputation of severely affected limbs with vascular ischemia. We were unable to find published reports of successful elbow contracture release in association with this condition. As the understanding of elbow anatomy and exposures improves, the results of the surgical treatment of heterotopic bone about the elbow are improving. We used the medial approach to release the ulnar nerve in our patient because, when flexion is limited to 90° prior to elbow release, ulnar neuritis frequently develops as patients regain flexion, which may limit further flexion following release of an elbow contracture. The medial approach is also helpful in addressing any fibrous contracture of the posterior oblique ligament of the medial collateral ligament; this approach was helpful in addressing the contracture found close to the hyperostotic lesion in our patient. It is notable that proximal release of the flexor-pronator muscle facilitated the approach to the anterior capsule and Z-plasty was effective in overcoming the long-standing muscle contracture.

Recurrence is a major concern following contracture release in patients with melorheostosis. Younge et al. noted that the soft-tissue contractures and periarticular fibrotic changes seen in patients with melorheostosis resemble those seen in patients with arthrogryposis multiplex congenita, in which the contractures are rigid and do not stretch with growth, thus causing recurrence of deformities. Only one of sixteen soft-tissue releases in their series was successful. Campbell et al. also reported that deformity recurred in five of eight joints. However, similar to the good result reported in the case of our patient, positive results have been reported anecdotally in adult patients who underwent surgical debulking of the hyperostotic cortex or who had total resection of the lesion.

References


