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Debridement arthroplasty of a rare case of elbow stiffness. A case report and literature review

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ABSTRACT

INTRODUCTION: Elbow stiffness is a common condition that affects the quality of life of patients. Melorheostosis of the elbow associated with elbow stiffness is extremely rare.

PRESENTATION OF CASE: We report the case of a 28 yr old male who presented with elbow stiffness which occurred within one year without prior history of trauma or infection. The patient had decrease in range of motion together with progressive worsening pain that forced him to seek medical attention.

DISCUSSION: There is no standard treatment for melorheostosis, and management plans must be made on an individual patient basis. The aims of treatment are pain relief and maintaining function.

CONCLUSION: Debridement arthroplasty is safe and effective in treating elbow stiffness associated with Melorheostosis.

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1. Introduction

Melorheostosis is a rare sclerosing bone dysplasia that affects both cortical bone and adjacent soft tissue structures in a sclerotomal distribution [1]. In the standard radiology or orthopedics literature Melorheostosis (from the Greek: melos = limb, rheos = flow) is described monotonously as a “flowing hyperostosis” resembling dripping candle wax [2]. Patients with this condition may have joint pain, stiffness, deformity, and restricted range of motion due to bony and soft-tissue contracture. Involvement usually follows a sclerotomal distribution and usually affects only one extremity [3]. 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. Up to date there is no specific treatment for this condition; options range from nonsurgical management (e.g. splinting and early training in making optimal use of the unaffected extremity) to various types of surgical management (e.g. tendon lengthening, sympathectomy, implant arthroplasty, or even amputation [4,5]). Melorheostosis of the elbow is extremely rare compared with that of the lower extremity, and a thorough search of the literature revealed only one case reported with a stiff elbow caused by this condition [3].

We report another case of a twenty eight-year-old man who had limited motion of the elbow as a result of isolated melorheostotic involvement of the radio-capitellar and ulnohumeral joint, who obtained a functional range of motion after debridement arthroplasty of the elbow using a combined medial and lateral approach. This study has been reported in line with the SCARE criteria [6].

2. Case report

A twenty- eight year-old male office worker presented with ankylosis of the right, dominant elbow. He reported limited elbow motion had developed gradually within 1 year. Initially there was no pain, but as the disease progressed the patient started feeling pain together with loss of motion. There was no history of elbow trauma or infection and the patient's daily work did not involve any strenuous exercise involving the arm. The pain and limitation of elbow motion had continued to increase and had resulted in a stiff elbow with a flexion deformity of 90° one year before the first clinic visit. The patient did not seek medical attention early because initially there was only loss of motion but 4 months before the patient presented to us, he mentioned that it was the progressive worsening pain that force him to look for medical help. Physical examination revealed decrease in the range of flexion, extension, supination and pronation of the right elbow when compared with that of the left. The patient had slight ulnar nerve symptoms; sensibility in the ulnar nerve distribution was present, but 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 osteophytes on the anterior aspect of the elbow

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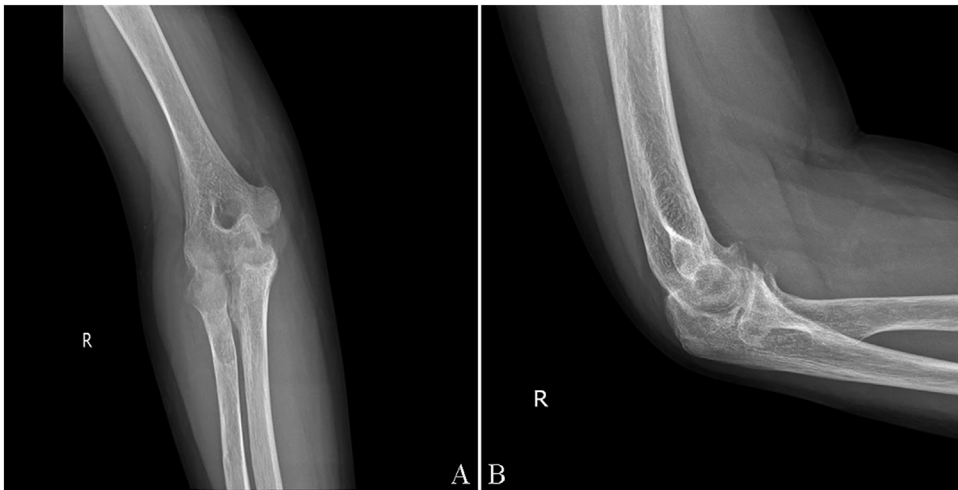


Fig. 1. Antero-posterior and lateral plain radiographs of right elbow.

(Fig. 1). A computed tomographic scan revealed nodularity of the periosteal bone formation and the presence of a flowing candle wax pattern on a short segment of bone (Fig. 2).

A diagnosis of melorheostosis of the radio-capitellar and ulno-humeral joint was made on the basis of the findings from these imaging studies. We did not perform splinting and physical therapy

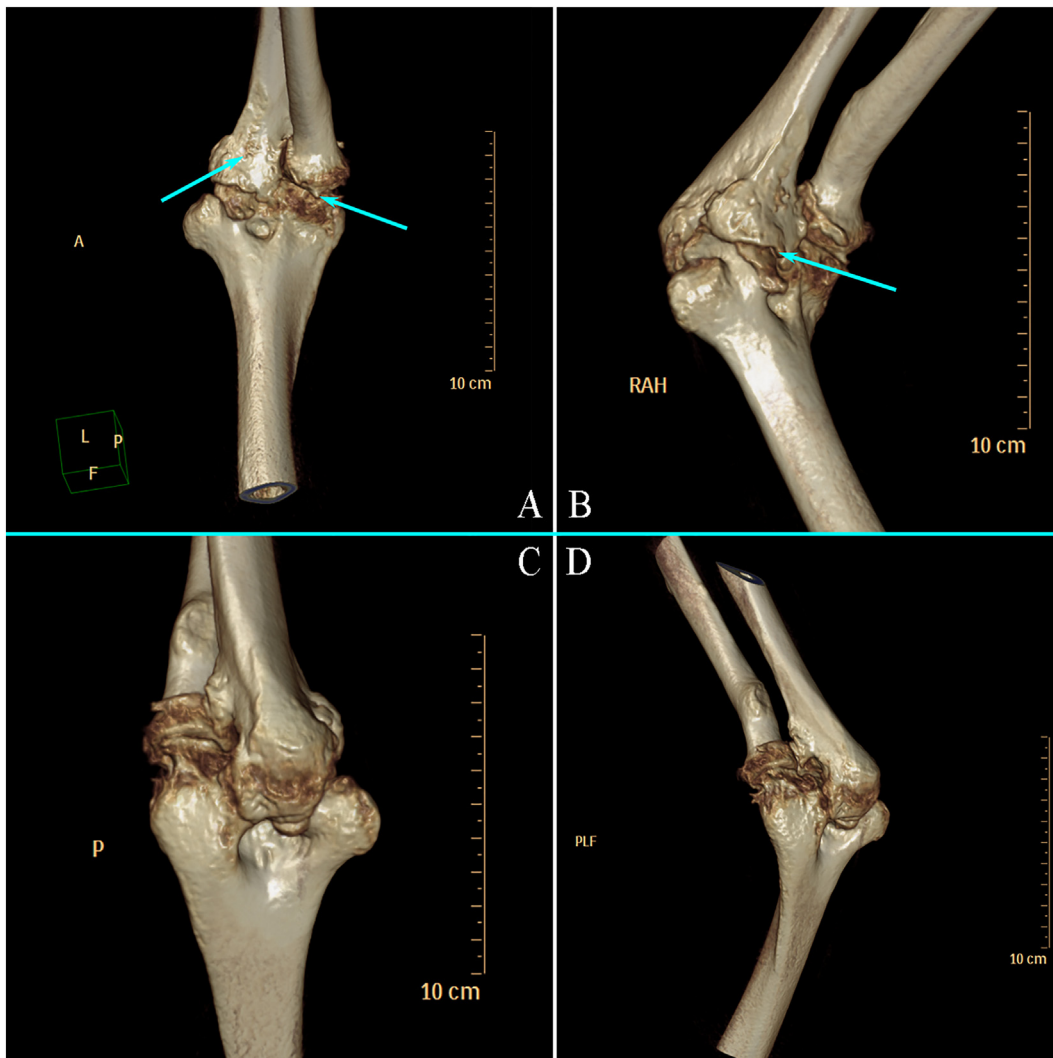


Fig. 2. showing 3D reconstruction computed tomography showing the classic appearance of dripping candle wax (arrows).



Fig. 3. Range of motion under regional block Flexion (80°) and Extension (60°).

because the patient had severe pain and limitation of functional use of the extremity; we immediately recommended surgical release of the contracture.

3. Surgical approach

The operation was performed with a sterile tourniquet and a brachial plexus block. The surgical approach was chosen based on the location of pathology. The medial approach was used to perform medial and posterior arthrolysis. The ulnar nerve was found to be compressed slightly at the groove of the ulnar nerve so it was released, and gently transposed anteriorly. To release the anterior and lateral parts, a lateral column procedure via an extended Kocher approach was used. The contracted capsule, scar tissue, and osteophytes were excised. For Pronation-supination limitation, the annular ligament and humeroradial joint were also released including osteophytes around the proximal radioulnar joint. The release was considered to be sufficient when the mobility passively achieved 0° to 130° . We also performed synovectomy to further relieve pain. Synovectomy was carried out until the lateral and medial compartments were cleaned of proliferative synovial tissue involving the capsule. Once all the osteophytes and synovial tissue had been excised, we released the tourniquet and with the utmost care we cauterized all the bleeding sites with a Bovie cauterizer. We hone in on this step as it helps reduce bleeding and swelling after surgery. The wound was closed over two drains, one anterior and one posterior. The elbow was splinted for 3 days, and then therapy was begun.

Before surgery under regional block the patient had a Flexion of 80° and extension of 60° as shown in (Fig. 3) and during surgery after contracture release the range of motion was 130° of flexion and 0° of extension. Peri-operatively we collected the excised hyperostotic cortical lesion and sent it for tissue diagnosis (Fig. 4). Histologic examination of the tissue revealed dense bone, which was consistent with the diagnosis of melorheostosis (Fig. 5). Three days postoperatively, Cryo/Cuff therapy was applied for at least 3 sessions of 20 min per day for 1 week and the patient was guided by a trained physiotherapist from our hospital to perform active and passive motion of the elbow. The patient was discharged twelve days postoperatively with a passive range of motion from 30° of flexion to 110° of flexion. Under the supervision of our physiotherapist, gentle range of motion was encouraged for the next six weeks. An extension splint was worn at night during this time. One year after surgery the patient returned for a follow up with a ROM of 110° , Flexion 120° , extension 10° , pronation 50° and supination 50° . The detailed pre-operative and post-operative data are shown in Table 1.



Fig. 4. showing the excised osteophytes after debridement.

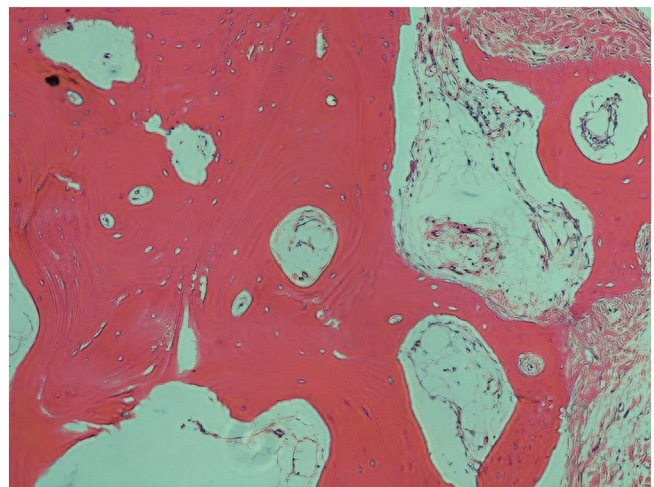


Fig. 5. Histologic examination of the hyperostotic cortical lesion showing irregular, dense, hyperostotic, cortical bone trabeculae of varying thicknesses.

4. Discussion

Melorheostosis was described for the first time in 1922 by Leri and Joanny [7]. Melorheostosis typically begins in an insidious manner, with slow progression over time. In some cases, it may demonstrate a waxing and waning course, with both active and quiescent periods [4]. This disorder may be diagnosed at any age, but usually presents in childhood or adolescence. Almost one-half

Table 1
Detailed Pre operative and Post operative data.

Patient data				before surgery						after surgery						
Case	Symptoms/duration	Side R/L	Approach	Pro/°	Sup/°	F/E°	ARC/°	Pain (VAS)	MEPS	PRo/°	Sup/°	F/E°	ARC/°	Pain	MEPS	Follow up
1	Loss of rom, Pain, numbness	R	Medial & Lateral	0	0	90/90	0	8	30	50	50	120/10	110	1	85	12 mo

Abbreviations: Range of motion (ROM), Months (mo), Flexion and Extension (F/E), Pronation (pro), Supination (Sup), Right and left (R/L).

of all cases are diagnosed by 20 years of age. The osseous lesions appear to progress most rapidly in childhood, with variable progression in adulthood. Both sexes appear to be affected equally by this non-hereditary condition. The disorder is usually recognized by limb deformity, contracture, bone or joint pain, joint stiffness, and/or limited range of motion, with pain and stiffness often the major symptoms.(1)

Elsheikh et al. [7] described the case of a 51 yr old male that presented with a painful swelling on the right shoulder. The patient had no history of trauma and the pain was progressive in nature. On examination, a swelling was identified in the antero-superior aspect of the left shoulder with redness of the skin overlying it. The swelling was firm, localized, and mildly tender. Radiographs demonstrated calcification around the greater tuberosity and sub-acromial space as well as hyperostotic lesions in the scapula blade and CT scan revealed extensive yet well-defined ossification within the proximal deltoid muscle and hyperostotic masses dribbling from the scapular blade. The patient was treated mainly for pain and was followed up for 6 months and the findings were stationary.

In 1968, Campbell et al [8], suggested that the mutation causing melorheostosis must be congenital and specifically that it must cause an early disturbance in embryonic development of neural crest structures. Murray and McCreadie [9] hypothesized that a neural infection, such as herpes zoster, might result in lesions distributed along the tissues supplied by the affected nerve root. In their series of 30 patients, 19 patients had abnormalities confined to one sclerotome and 11 had involvement of multiple sclerotomes, with the latter cases correspondingly associated with more severe clinical manifestations. Kim et al. [10] suggested that another causal mechanism might be downregulation of adhesion proteins involved in osteoblastic regulation, specifically of the TGF-β-induced gene product, which might contribute to the development of hyperostosis and associated soft tissue abnormalities. Papadakis et al. [11] have recently performed whole exome sequencing of affected and unaffected bone from 23 patients with melorheostosis, with the results soon to be reported.

Melorheostosis of the upper extremity is extremely rare compared to that of the lower extremity. Most reported cases involving the upper extremity are focused on the hand or on the sclerotomal distribution pattern [5,9,12–20]. Our case report describes the case of an office worker who had stiffness of the elbow without strenuous use of the elbow and without trauma or infection. In literature up to date there is only one case report, Gong et al [3], mentioned melorheostosis of the non-dominant elbow of a thirty- seven year old man that presented with pain and limited elbow motion that had developed after he played a game of squash five years previously. The patient had pain that decreased with time, but limitation of elbow motion had continued to increase and had resulted in a fixed flexion deformity of 90° which was different compared to our patient that had limitation of motion without pain initially and as the disease progressed our patient had severe pain with decreasing range of motion forcing him to seek medical attention. The other differences were that their patient had no limitation in pronation, supination and no ulnar nerve symptoms pre-operatively.

There is no standard treatment for melorheostosis, and management plans must be made on an individual patient basis. The aims of treatment are pain relief and maintaining function [21]. Some

Surgical treatment for this condition includes tendon lengthening, excision of fibrous and osseous tissue, fasciotomy, capsulotomy, sympathectomy, corrective osteotomy, Ilizarov lengthening, arthrodesis, implant arthroplasty, and even amputation of severely affected limbs with vascular ischemia. We believe it is of great importance to raise awareness among orthopedic surgeons and researchers to be familiar with this extremely rare disease that cause stiffness of the elbow in young people. Further research and more studies reporting about melorheostosis of the elbow will help us gain insight about this complex disease.

This study has been approved by the Ethics committee of our institution: Reference number: L-2017-11

Disclaimer

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Conflict of interest

The Authors declare that there is no conflict of interest.

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Ethical approval

Ethical approval has been obtained from The second affiliated hospital and Yuying children’s hospital of Wenzhou Medical University Research Ethics Committee.

Ethics reference number: L-2017-11.

Consent

The patient was informed that data concerning the case would be submitted for publication, and they consented.

Author contribution

Author’s contributions: KP and HY conducted the conception and design of the study, acquisitioned interpretation of data, drafting the article; CLY, XZ, ZW, DL for English editing. All authors have reviewed the manuscript.

Registration of research studies

Not applicable

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