

Case Report

Melorheostosis: A Rare Cause of Limb Pain

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ABSTRACT

Melorheostosis is a rare skeletal dysplasia, and to date there are no reports of this condition in Australia. This is a case of a lady who presented with dull arm pain with a pathognomonic radiological findings. The plain radiographs obtained of the limb demonstrated a characteristic candle wax appearance in a monomelic distribution. The natural history, presentation, and management are discussed in detail.

Keywords

Melorheostosis; Skeletal dysplasia; Central Australia.

CASE PRESENTATION

An indigenous lady in her fifth decade was referred by her general practitioner to the orthopaedic clinic with 5-year history of a dull left upper limb pain. She had no significant medical history, nor history of trauma or infection. Physical examination revealed full range of motion of her affected limb wasting of the thenar eminence, and positive Tinel's test at carpal and cubital tunnels. There were no other significant findings of her upper or lower

limbs. Plain radiographs revealed diffuse cortical thickening and sclerosis, resembling candle wax, along the radial border of the hand and wrist diagnostic of melorheostosis (Figure 1). Similar findings were noted in the proximal humerus (Figure 2). Results of biochemical investigations for infection and metabolic bone screen were normal. The information with imaging was sufficient for a diagnosis of melorheostosis. Further investigations were organized to address the nerve compression symptoms, however, the patient defaulted follow-up.

Figure 1. Classic Candle Wax Appearance Seen along the Radial Border of the Wrist and Hand



Figure 2. Similar Candle Wax Appearance of Melorheostosis Seen in the Proximal Humerus



DISCUSSION

Melorheostosis, first described in 1922, also known as Leri's disease, is a rare mesodermal bony dysplasia that may involve surrounding soft tissues.¹ The global incidence is approximately 0.9 per million population.² It is known for its classical appearance of periosteal cortical thickening in a "dripping wax" or "candle wax" form on plain radiograph. There are other known radiological patterns of this disease including osteoma like myositis ossificans like and osteopathia striata like.³ These features have a sclerotomal distribution and maybe monostotic, polyostotic or monomelic.⁴ It was monomelic (one limb) in our patient. Melorheostosis presents in adulthood with a gradual onset of non-specific symptoms including pain and stiffness. When adjacent soft tissues are involved scleroderma-like skin changes, stiffness, deformity, and nerve compressive symptoms may manifest, as were present in our case above.⁵

Plain radiographs are sufficient for diagnosis. The differential diagnosis of infection or musculoskeletal neoplasm should be considered, and a biopsy is recommended if there is any doubt. Further imaging is generally of little value for diagnosis.⁴ Magnetic resonance imaging (MRI) can be helpful to evaluate the soft tissues and to exclude other pathologies.⁶ With nuclear medicine scans, the lesions have an increased uptake and hence is useful for surveillance and identifying lesions at various sites.⁷ A link between the intensity of uptake and symptoms have yet to be established. Biochemical tests such as alkaline phosphatase and serum calcium levels are inconclusive but may serve in the work-up to exclude other pathology.⁴

There is no definite cure established for melorheostosis. Hence, there is no specific treatment and management is largely towards symptom relief. Multidisciplinary care from orthopaedic surgeons, metabolic bone and pain physicians are important in the long-term management for these patients. Non-operative options include analgesia, physiotherapy, and use of appropriate orthosis for immobilization. There are reports in the literature on use of bisphosphonates to decrease bone pain, as it reduces osteoclastic activity.⁸ Surgical management poses a unique set of challenges as operations are performed on pathological bone. A variety of procedures have been described to relief symptoms such as, but not limited to nerve decompression, excision of fibrous tissue, debulking of exostosis, corrective osteotomy, and arthrodesis.⁹

We present a case of a rare pathology in our local setting, with no previous reports in literature on its prevalence in Australia. Knowing the global incidence, we extrapolate that there are potentially up to 24 cases per year, given the Australian population of 25 million.¹⁰ It is also an example of the varying pathology that is prevalent in our local population, and more so Central Australia. We hope readers will have an increased awareness of this rare skeletal dysplasia and unique pathology in Australia. Early diagnosis and timely counselling of patient can avoid invasive diagnostic

investigations and procedures. Misdiagnosis could lead to surgical treatment with bone debridement if mistaken for osteomyelitis. This could result in more harm instead of improving a patient's outcome.

DISCLOSURE STATEMENT

Nothing to disclose.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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