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Case Report: A 25-year-old female with right leg pain for a two-year duration. She has been seen by a lot of physicians and could not have been managed properly. Her radiographs were showing hyperostotic left ulna and right tibia. Sinister pathologies were excluded using MRI and biopsy. Her symptoms settled with intravenous Zoledronate. Pain recurred after two years and she responded well after repeated Zoledronate injection.

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Conclusion: Though there are no key clinical features, diagnostic tests, or cause of this clinical condition, symptomatic management is adequate to manage patients with this clinical condition. Intravenous Zoledronic acid is an option for patients who have severe and resistant symptoms.

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I. INTRODUCTION

Polyostotic melorheostosis is a rare congenital non-hereditary benign sclerosing mesenchymal dysplasia involving multiple bones(1–4). Melorheostosis is presented in the second decade of life. The male and female ratio is the same. The classical description of appearance in the radiographs is “flowing candle wax”(1). The axial skeleton is rarely involved. The treatment is mainly non-operative. Surgery is reserved for the lesions complicated with fractures.

We present a case report on a patient with symptomatic polyostotic melorheostosis successfully treated with intravenous Zoledronate injection.

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II. CASE REPORT

A 25-year-old female presented to the orthopaedic unit with pain over her right leg left forearm and back for two years duration. She has been seen by a lot of physicians for the same ailment and they could not help her. The pain gradually worsened over time and made her visit us. On examination, the right leg and the left forearm were swollen diffusely. There is no history of back pain.

Radiographs show hyperostosis of the right tibia and the left ulna (Figure 1). Technetium 99 bone scan revealed hyperactivity seen on the right tibia, left ulna, left fibula, left femur and the spine (Figure 2). The bone profile including serum calcium, phosphate and alkaline phosphate was normal. Bone biopsy revealed normal bony architecture. She was given an intravenous Zoledronic acid injection and became asymptomatic. She was followed up at the

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Orthopaedic clinic for two years. Again, she developed pain over the same region and she was treated again with the same medication. Now she is completely pain-free and her activity of daily living has improved dramatically.

Figure 1: Shows radiographs of symptomatic anatomical regions show polyostotic sclerotic bony lesions involving the left proximal ulna (Figure 1A), right tibia (Figure 1B), left fibula (Figure 1C) and left proximal ulna (Figure 1D). All lesions are identical and have scalloping edges compatible with the classical description of the disorder.

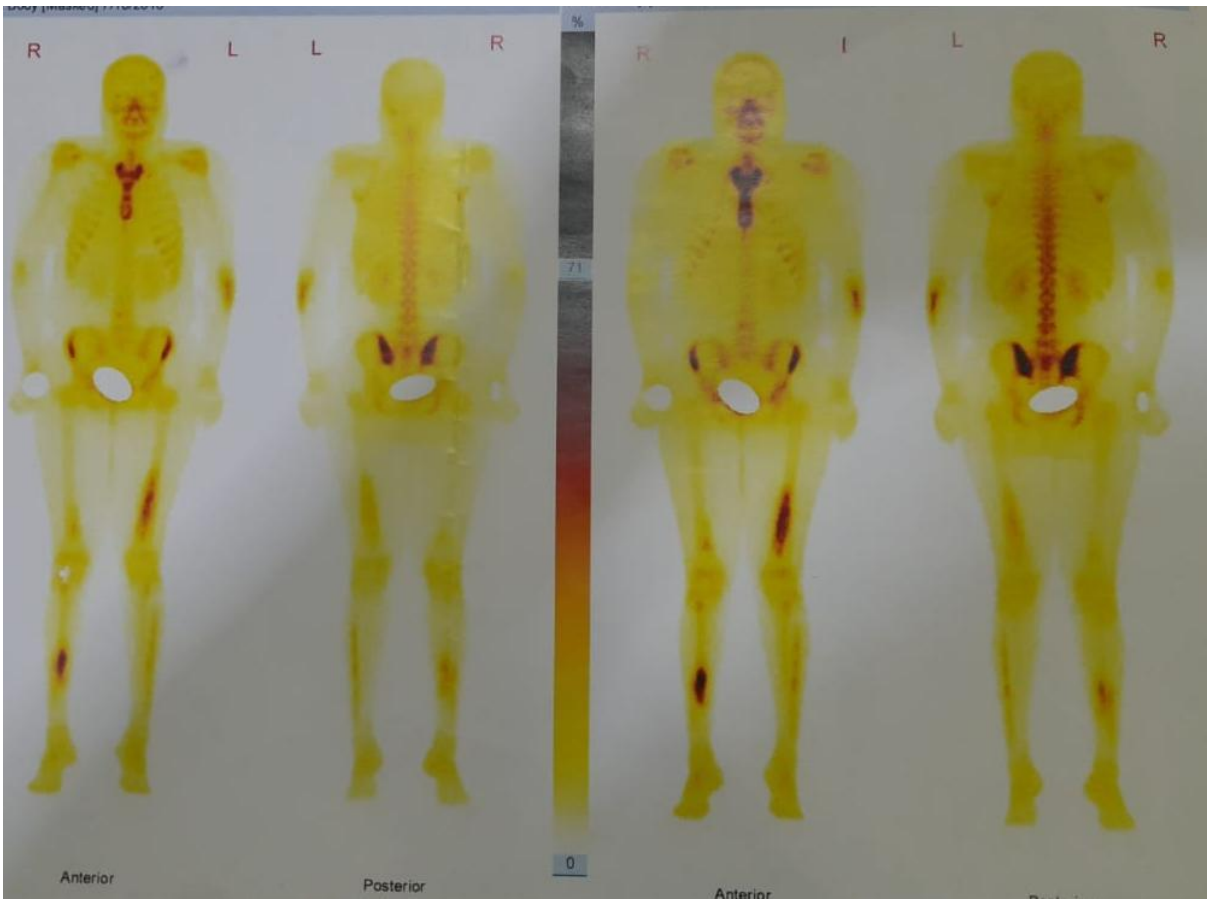

Figure 2: Shows the whole body Technetium bone scan of the same patient which revealed hyperactivity seen in the right tibia, left ulna, left femur and left fibula. In addition, the spine and pelvis also show hyperactivity.

III. DISCUSSION

Melorheostosis is a rare disorder in which sclerosing dysplasia with derangement in endochondral and intramembranous ossification of bones. The summary of demographical details is in Table 1.

Demographic Details	
1. Incidence	0.9/Million
2. Male: Female	1:1
3. Age of presentation	2-64 years
4. Majority of presentations at the age of	Around 20 years

Figure 3: Summary of demographic details of patients with melorheostosis (1, 2, 4, 5)

These lesions can be on one bone (Monostotic), many bones (Polyostotic), involving one limb (Monomelic) or generalized. Axial skeleton involvement is rare. According to the anatomical involvement, severity and the deformity caused by the lesions symptoms and signs will be varied.

Commonest symptoms are painful limb swelling, restricted range of motion and contractures of joints (usually asymmetric). Axial skeleton involvement may present with back, and neck pain,radicularpain,scoliosis,stiffness,giddiness,symptomsofvertebrobasilarinsufficiencyandevidenceofcord compression(1).

Melorheostosis is not a fatal condition. But it impacts significantly the activity of daily living of patients with this ailment. The management of melorheostosis varies from simple analgesics to surgery. The management depends on the

anatomical site, the extent of lesions, the severity of symptoms and the degree of deformity. The primary goal of management is to relieve symptoms and achieve mobility. Pain relief, nerve blocks, braces, and physiotherapy play a vital role. Patients who are not responded to these options will undergo nerve blocks. Surgery deserves for patients with lesions complicated with fractures.

There are few case reports on the usage of intravenouszoledronatefor melorheostosis(6–8). They reported successful outcomes following treatment after the injection. On our patient single dose of intravenous zoledronic acid (5mg over 30 minutes) has provided remission for two years and it worked for relapse as well after two years. Intravenous zoledronate is an effective treatment for symptomatic polyostotic melorheostosis. Zoledronate is a bisphosphonate which inhibits osteoclasts and reduces bone pain, prevents pathological fracture and reduces blood supply(6–8). Bone resorption by osteoclasts, stimulation of pain receptors and increased intraosseous pressure are major reasons for bone pain among patients with melorheostosis (6–8). Thus, zoledronate is a viable option for the ents with melorheostosis causing bone pain. Anyway, careful selection of patients and preparation is necessary before the zoledronate infusion to avoid complications (Avascular necrosis of the jaw, fever, allergic reactions etc).

IV. CONCLUSION

Symptomatic polyostotic melorheostosis may be resistant to symptomatic treatment. Intravenous zoledronate is a viable option for such patients.

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