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## *Full Length Research Article*

# MELORHEOSTOSIS AFFECTEDPAINFULL LIMB IMPROVED WITH ZOLODRONIC ACID – CASE REPORT OF A RARE DISEASE

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**ABSTRACT**

In 1922 Leri and Joanny, first described Melorheostosis (synonyms: candle bone disease, melting wax syndrome, Leri disease). Actually it's a metaplacia. Men and women are equally affected, dermatomal distribution, monomelic distribution. Onset is insidious, and most common symptom is pain. Classical radiological appearance of 'flowing hyperosteosis' resembling hardened wax that has dripped down the side of a candle. Recent literature shows relation with LEMD 3, encode inner nuclear protine,

**Case repot:** A 30 years old woman, residing at Tarakeswar, Hooghly, West Bengal, presented with left leg pain with mild swelling and limitation of knee movement. On examination non tender bony heard swelling and restriction of knee movement present. Plain radiographs showed extensive, dense, undulating or irregular cortical hyperostosis, resembling candle wax, extending along the length of bone. Zolidronicacid decrease her pain.

**Conclusion**: Melorheostosis, rare, benign, insidious, candle wax. Routine laboratory findings usually are normal. The exact cause remain unclear. There is nodefinite treatment available for this disease. Only symptomatic treatment improve the condition of the patients, more fruitful result obtain with zolodronicacid.



## INTRODUCTION

In 1922 Leri and Joanny, first described Melorheostosis (synonyms: candle bone disease, melting wax syndrome, Leri disease) is a rare chronic bone disorder actually it's a metaplacia. Men and women are equally affected, dermatomal distribution and no hereditary features have been discovered. Onset is insidious, and most common symptom is pain. Most common part of bone is diaphysis of the long bone of lower limb rarely the axial skeleton. Classical radiological appearance of 'flowing hyperosteosis' resembling hardened wax that has dripped down the side of a candle. Recent literature shows relation with LEMD3,encode inner nuclear protine, Its distribution often suggested a sclerotomal abnormality involment of one limb or monomelic distribution.Some times melorrheostosis shows leniar distribution through epiphysis, metaphysic, diaphysis,at times corossing contagious bone at joint. Though it is benign but its pain full. may cause deformity. Bisphosphonate has a role to decrease pain.there is a case with melorheostosis with monomelic distribution in a female and pain decreased by intravenous zolidronic acid. (12,13)

### Case report

A 30-year-old woman, residing at West Bengal, Tarakeshawar, presented with a history of mild swelling over shin border of tibia (Baer *et al*., 1994; Murphy *et al*., 2003) and pain over thigh and leg for last 15 years. and limitation of left knee. Pain is dull aching in nature, no improvement after prolong intake of analgesic and physiotherapy advised by local doctor.no history of any constitutional syndrome.On physical examination, the patient had hard swelling of the sclerosing bone dysplasia. Plain radiographs (Fig 2,3,4,5) of left leg showed extensive, the sclerotomes. The hypothesis ofdense, undulating or irregular cortical hyperostosis, candle wax, extending along the length of bone, Plain radiographs of the bones on the right lower limb was normal. Laboratory findings for CBC, secalcium, phosphorus, alkaline phosphatase within normal limit.

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**Fig. 1.**

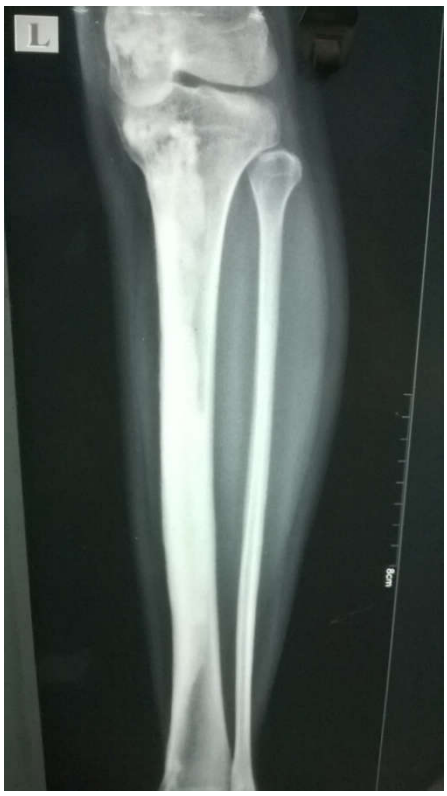


**Fig. 2.**

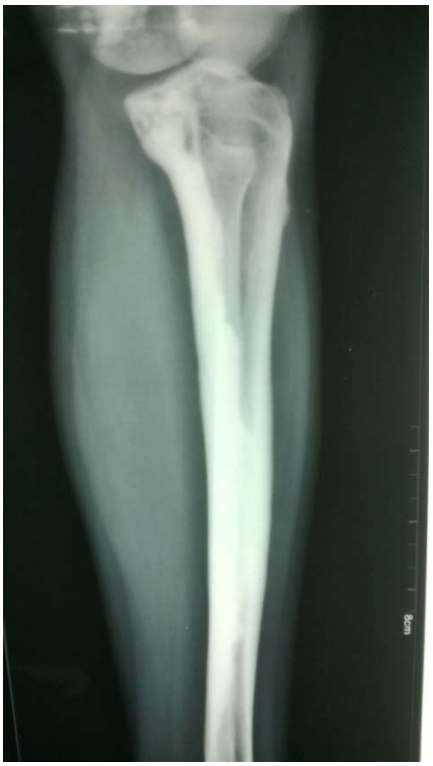

**Fig. 3.**

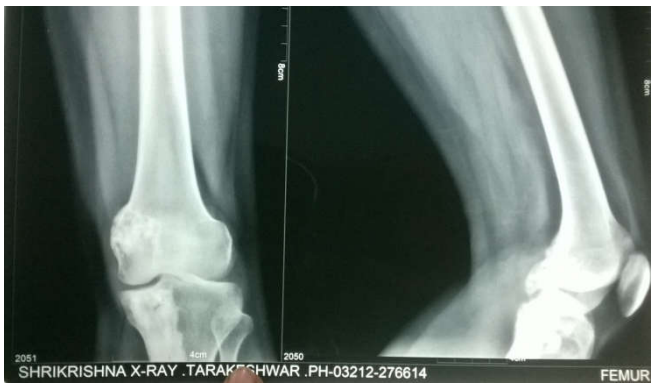


**Fig. 4.**

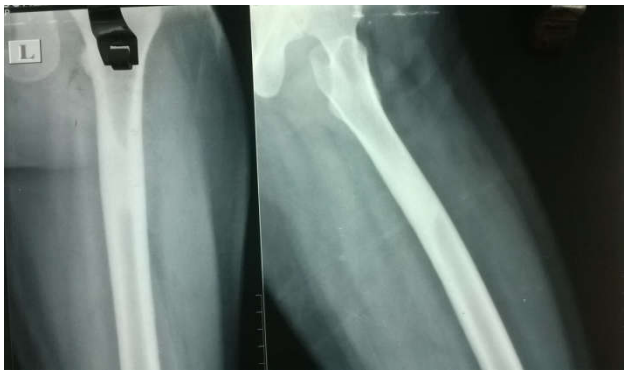


**Fig. 5.**

**Fig.1. Shows shin border thickening of tibia., Fig-2, 3 Shows tibial unilateral involvement, Fig. 4. showcongagious bone involvement across the joint. Fig-5 shows femoral involement**

After giving ivzolindronic acid 5mg dissolved in 100ml NS for over 30 min slow transfusion and oral supplement of calcium, and vitamine D3.After 6 month follow up pain decrease significantly.

## DISCUSSION

Melorheostosisis rare,the onset of this rare diseases isinsidious, and the first symptom isusually pain because of the subperiosteal bone formation. The monomelic variant is the most common presentation of this rare disease (Zeiller *et al*., 2005). In this patient, left leg was affected. Melorheostosis is often thought as a benign sclerosing bone dysplasia. The several hypothesis are given but the exact aetiology remain unclear. Murray and McCredie, 1979, hypothesis was that, embryonic infectionof nerve root causes neural scarring and segmental bone. Routine laboratory findings usually are normal. Histologic findings are usually nonspecific and often show dense bone formation, a mixture of mature and immature bone elements. Treatment is mainly symptomatic. Most patient receives nono perative treatment. Operative treatment consists of tendon lengthening, excision of hyperostoticbone, osteotomies, sympathectomy and lastly amputation is the last option (Freyschmidt, 2001).

Bisphosphonate are commonly use (Wood *et al*., 2002). Potential causes of the bone pain in melorheostosis include increased osteoclastic bone resorption and activation of painreceptors, raised intraosseous pressure and increased vascularity secondary to hyperosteosis and soft tissue involvement around joints. Thus, bisphosphonate treatment via a number of mechanisms would be expected to reduce inflammatory bone pain and symptoms in melorheostosisbisphosphonates in hibit osteoclastmediated bone resportion by direct and indirect actions onosteoblasts and macrophages and bone vascularity. They have been shown to decrease bone pain, slow progressionof bone lesion (Wood *et al*., 2002). Melorheostosis does not shorten life span, however, morbidity may be considerable.

### Conclusion

All routine laboratory findings usually normal. It is only diagnosed by characteristic candle wax type appearance in x-ray film. There is no definite treatment available for this disease. Only symptomatic treatment improve the condition of the patients, more fruitful result obtain with zolidronic acid.
But to establise definite role of zolidronic acid in melorheostosis need more case study.

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