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# MELORHEOSTOSIS OF PROXIMAL FEMUR- A RARE CASE REPORT

Orthopaedics

**Dr. Sudhir Shyam Kushwaha** Associate Professor, Dept. of Orthopedics, AIIMS, Gorakhpur-U.P.
**Dr. Atil Kumar Lal** Assistant Professor, Dept. of Orthopedics, B.R.D. Medical College, Gorakhpur-U.P.
**Dr. Asad Khan** Senior Resident, Dept. of Orthopedics, B.R.D. Medical College, Gorakhpur-U.P.

KEYWORDS

## INTRODUCTION

Melorheostosis is an uncommon benign tumour of mesodermal origin belonging to group of sclerotic bone dysplasia. Leri and joanny in 1922 described it for the first time as candle wax hyperostosis (1). Melorheostosis is derives from the Greek word melos = limb and rheos = flow due to the classic radiological appearance of 'flowing hyperosteosis' resembling hardened wax that has dripped down the side of a candle. Its incidence is as low as 0.9 per million population (2) No hereditary predisposition has been seen. Both men and women are equally affected (3). The disease tends to be monmelic with both monostotic and polyostotic variants. The disease is usually silent and sometimes may be found incidently. If symptoms appear they are usually pain and limitation of joint movement other complains can be limb length discrepancy, progressive deformity and ossification of surrounding soft tissue.

## CASE REPORT

We report the case of 19 year old female with complains of dull aching pain left hip, knee and leg region since 4 years. There was no relevant family or any history of trauma. On physical examination a Genu Valgum (Fig.-1) deformity of Q angle of 15 degree at left knee joint was measured. Left hip internal rotation was slightly restricted and painful. No quadriceps hypertrophy, no fever, no swelling and no limb length discrepancy were found.

Plain radiograph of proximal femur and shaft showed irregular cortical hyperostosis, classical candle wax appearance resulting in narrowing of the medullary cavity. (Fig.2) At knee joint hyperostosis was seen on the medial aspect resulting into valgum deformity. In leg similar to femur entire medial aspect of tibia showed hyperostosis.

A CT scan was done which showed hypercondensation in medial cortex of femur and tibia. Laboratory investigations such as complete hemogram, serum calcium, phosphorus, alkaline phosphatase, erythrocyte sedimentation rate, C-reactive protein, α-fetoprotein, carcino-embryonic antigen were all found to be within normal limits. We started alendronate 70mg twice a week for 4 weeks. Analgesics were given for symptomatic relief. Alleviation of pain was seen after three months follow up.

## DISCUSSION

Melorheostosis etiology is still not well known, although role of genetic predisposition has been suggested but still no familial cases have been reported. Associations with melorheostosis include mainly skin changes in form of hyperpigmentation of overlying skin and scleroderma(5,6,7). Its association is also seen with tumours such as fibrous histiocytoma, osteogenic sarcoma and dermoid cysts (3, 4).

Melorheostosis mainly present in monomelic form with monostotic and polyostotic variant (5). Both upper and lower limbs can be affected in addition to the axial skeleton. The lower limbs are affected more often than the upper limbs. Plain Radiographs show a very characteristic candle wax or melting wax appearance and is sufficient to make the diagnosis. MRI can be used to differentiate other diagnosis and soft tissue extension (8,9). Bone Scintigraphy is recommended to detect early changes and for monitoring progress of disease.

Treatment is not well established. It is symptomatic in most cases, in order to control bone pain. Some of the major causes for bone pain in melorheostosis are thought to be increased osteoclastic bone resorption, increases intraosseous pressure, activation of pain receptors and soft tissue involvement around joints. Hence, bisphosphonates are mainly used as a treatment as they reduce inflammatory bone pain , inhibit osteoclast-mediated bone resportion by direct and indirect actions on osteoblasts and macrophages and bone vascularity(10). Pamidronate 30mg daily for 6 days is generally used and is seen to relieve symptoms but it does not decrease the disease progress(12). Surgical treatments include osteotomies, tendon lengthening, excision of hyperostotic bones and amputation(11).

## CONCLUSION

Melorheostosis is rare, chronic benign tumor. Plain radiograph with typical candle wax appearance is enough to make the diagnosis. However MRI and Bone Scintigraphy can be used to determine other differential diagnosis, subclinical detection and to monitor disease progress. Treatment is mainly conservative and symptomatic with Bisphosphonates and NSAIDs.


**Fig-1 :** Shows genu valgum deformity in right knee. Q-Angle was measured clinically and was found to be- 15 degrees.

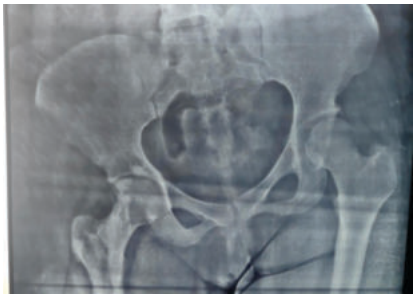

**Fig-2 :** Shows irregular cortical hyperostosis, classical candle wax appearance resulting in narrowing of the medullary cavity on right side.

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