MELORHEOSTOSIS in an adolescent with limb length discrepancy. Management with epiphysiodesis using 8 –plates.

N Laliotis, C Chrysanthou, P Konstantinidis, Lizeta Papadopoulou

Inter Balkan Medical Center Thessaloniki

MELORHEOSTOSIS

- Melorheostosis is a rare benign sclerosing bone dysplasia. It is described as flowing hyperostosis resembling the flow of wax at the side of a candle. The term is derived from the Greek words melos that means limb, rheos that means flow and oston that means bone.

MELORHEOSTOSIS

- The disorder begins in childhood. It affects mainly the long bones of the upper or lower limb and is rare in the axial skeleton. Main symptom is pain, contractures and deformity of the limb. Limb length discrepancy (LLD) has been reported as a clinical sign of the disease.

MELORHEOSTOSIS INITIAL CLINICAL PRESENTATION

MELORHEOSTOSIS x ray

MELORHEOSTOSIS x ray

- On x-ray examination of his legs, femur and tibia on the right leg had **longitudinal diffuse hyperostosis**, affecting the medullary canal. There was **endosteal and cortical irregular thickening** mainly of the tibia. There were irregular areas of ossification in the epiphysis of the femur and tibia. Endosteal ossification was present on the talus of the same foot. Growth plates were open and normal both for femur and tibia. Soft tissue was normal, with no signs of heterotopic ossification

MELORHEOSTOSIS scanogram

- discrepancy as 2 cm in the femur and 1, 6 cm in the tibia.

MELORHEOSTOSIS mri

MELORHEOSTOSIS mri b

- On T1 and T2 images the areas of ossification appeared as heterogeneous intensities that had uptake on gadolinium. Surrounding soft tissues appeared normal

MELORHEOSTOSIS bone scan

- Technetium -99 bone scan showed increased uptake of the femur and tibia and osteoblastic activity, corresponding to the radiological extension of the hyperostosis.

MELORHEOSTOSIS epiphysiodesis

- We proceeded in epiphysiodesis, using 8 plates on both sides of the growth plate, in the distal femur and proximal tibia.

MELORHEOSTOSIS final result

- We removed his 8 plates with the completion of growth, 2 years after initial surgery, having complete equalization of the femur and 0,8 cm discrepancy of the tibia.

MELORHEOSTOSIS

- Melorheostosis is a mesodermal bone sclerosing dysplasia. It is a rare condition with an incidence of 0.9 in 1,000,000.
- Several series have been reported, trying to define this rare entity with unknown etiology.
- In contrast to other bone dysplasia as osteopetrosis or osteopoikilosis it is not a hereditary disease, having a sporadic presentation. There is a possible genetic basis with loss of function of LEMD 3 gene but this is not always found.

MELORHEOSTOSIS

- It affects the long bones rather the axial skeleton. Usually it is a monomelic disease. The lower limbs are more often affected. The skull, spine, ribs and facial bones are exceptions for been affected. It can be monostotic or polyostotic. Bilateral involvement is rare. In polyostotic cases, ossification may cross the joints.

MELORHEOSTOSIS

Melorheostosis is usually diagnosed in childhood, adolescent or early adult life. It can be an incidental finding in an x-ray examination. Usually it presents with pain, contractures and deformity that may be initially confused with arthrogryposis, if they affect the hand or feet.

A variety of skin and vascular lesions may be associated. Artner et al [1] reviewing 313 cases reported the presence of naevi, scleroderma, haemangiomas and fibrosis. These lesions have usually a sclerotome distribution, similar to the radiological appearance spelling

MELORHEOSTOSIS xray patterns

- Melorheostosis is mainly diagnosed from radiographic examination.
- Freyschmidt has described different radiographic patterns for the disease. He reviewed 23 cases over a 16 year period.
- The patients reported had a mean age of 34yrs (range 7 – 70 years).
- cortical thickening with the appearance of flowing wax.
- osteoma like appearance with endosteal hyperostosis that was eccentrically located.
- A myositis ossificans pattern with soft tissue ossification around the hip and knee
- An osteopathia striata like pattern, with eccentric dense hyperostosis striatations.
- a mixed pattern for 4 patients.
- Younge et al [10]underline that **children have endosteal sclerosis**, in contrast to adults with the **extraosseous flowing outside** the diaphysis of the long bone.

MELORHEOSTOSIS LLD

- Limb length discrepancy (LLD) was found in 49 of 182 cases from Artner (27%). Morris described 35 cases of LLD with similar percentage. The short limb was the affected one and the exception was the elongation of the affected one. [1,8]
- In the series of Smith et al from Mayo Clinic, in 24 patients, LLD was found in 4 patients. They report improvement of gait with shoe raiser. They further report good result with epiphysiodesis. [2]

MELORHEOSTOSIS LLD

- Limb length discrepancy (LLD) was found in 49 of 182 cases from Artner (27%). Morris described 35 cases of LLD with similar percentage. The short limb was the affected one and the exception was the elongation of the affected one. [1,8]
- In the series of Smith et al from Mayo Clinic, in 24 patients, LLD was found in 4 patients. They report improvement of gait with shoe raiser. They further report good result with epiphysiodesis. [2]

MELORHEOSTOSIS Ilizarov treatment

Marshal and Bradish[14] report a 27 year old man with melorheostosis, with shortening of the affected limb of 4 cm that was equally divided to femur and tibia. He was initially diagnosed at the age of 11. They treated him with lengthening of the tibia, with 4 cm elongation, with a monolateral external fixator. The regenerated tibia had the same appearance as the melorheostosis. Since callus formation in lengthening is mainly intramembranous, they conclude that the disorder is an intramembranosous ossification disturbance.

MELORHEOSTOSIS MESSAGE

We treated our patient with epiphysiodesis with 8 plates, since his active growth plate was normal and not affected from the melorhostosis. We achieved an almost complete equalization of the discrepancy. Our patient was very satisfied with the final result. We continue to review our patient, even though melorheostosis has a benign course. No further increase in his radiographic findings is expected.

MELORHEOSTOSIS

Case Report | Journal of Orthopaedic Case Reports 2019 September-October: 9(5):Page 7-10

Melorheostosis in an Adolescent with Limb Length Discrepancy and Management with Epiphysiodesis with Eight Plates

Nikolaos A Laliotis[1], Panagiotis Konstandinidis[1], Chrysanthos K Chrysanthou[1], Lizeta Papadopoulou[1]