Incidental Presentation Of A Rare Case: Melorheostosis

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INTRODUCTION:

Melorheostosis is a rare bone disease,non genetic, non developmental, sclerosing dysplasia of the bone and its etiology is still unclear. On xray, characteristic findings are periosteal hyerostosis along the cortex of longbones, resembling flowing or dripping of candle wax. Melorheostosis has no known cure and treatment aims to maintain quality of life.

CASE PRESENTATION

Patient is a 20 year old gentleman, known case of hypertension on treatment, presented with worsening swelling involving whole of left upper limb from arm to fingers, and heaviness of them affected limb. Stiffness noted over shoulder,elbow , wrist and MCP joints.Blood investigation serum PTH , ESR, TFT, corrected calcium was normal. Xray demonstrated typical melting wax appearance over humerus, radius and ulna. Patient subsequently was referred to tertiary centre for management.

DISCUSSIONS:

the pathology for melorheostosis is still unclear, there are some reports about the role of genetic predisposition, and in some reports gene mutation as well.

The condition can present in multiple locations, upper limb, lower limb, axial skeleton as well. Xray can show characteristic changes, besides that it has 9 typical signs. Thickening of outer layer of bone, skin affection, intermittent joint swelling, joint pain, limb deformity, nerve oppression, pain, paraesthesia and reduced range of motion. CT shows bone lesions with the classical aspects, and quantifies it in the spatial planes, MRI can describe extension into soft tissues. Bone scintigraphy can detect subclinical lesions and can be used to monitor progress.

There is no clear management guidelines. Protocol based on pamidronate improve symptoms, but not the progression.

CONCLUSION:

For diagnostic purposes, xrays are sufficient as characteristic findings can be seen as mentioned. Lab results are usually normal. Treatment is symptomatic for pain, usually with NSAID and alendronates.

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