Case 1304

Melorheostosis
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Section: Musculoskeletal system
Case Type: Clinical Cases
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Patient: 21 years, male

Clinical History:
The patient presented with swelling and chronic pain of the forearm without any medical history.

Imaging Findings:
The patient presented with swelling and chronic pain of the forearm. A radiogram demonstrated cortical lesions characteristic of melorheostosis. No pathologic proof has been obtained, the diagnosis was based on typical plain film findings.

Discussion:
Melorheostosis is a rare bone disease. The cause and pathogenesis are not known. The disease does not appear to be hereditary. It manifests after early childhood; occasionally, however, the initial signs appear in adult patients. Initial clinical manifestations include swelling and pain of joints and limitation of motion, muscle contracture, and tendon and ligament contracture. Radiological manifestations are commonly limited to a single limb in which one or more bones may be affected. The abnormalities are rarely seen in the skull and facial bones, vertebrae or ribs.

This disorder presents as an irregular thickening of the cortex along one side of a bone or the bones of one extremity. The thickening may be external, internal or both. The appearance has been likened to molten wax flowing down the side of a candle. This sclerotic bone appearance may involve one side of the tubular bones of the upper or lower extremity affecting the carpus and tarsus as well as the metacarpals, metatarsals or phalanxes. Soft tissue calcifications and ossifications are not infrequent and may lead to ankylosis of the joint.

If the disease begins early in life the epiphyses may fuse prematurely causing shortening of the involved extremity. The lesions usually cease progressing when skeletal growth is complete. Regression has not been noted. Growth disturbance can be severe leading to joint contracture, foot deformity or scoliosis. The disease can result in considerable deformity and disability.

Scintigraphy may reveal areas of increased uptake of radionuclide. With MR imaging, bone and soft tissue lesions are of low signal intensity on both T1 and T2 sequences.

The disease can accompany tuberous sclerosis, Gardner's syndrome, fibrous dysplasia and infantile cortical hyperostosis.

During the course of melorheostosis, life expectancy does not seem to be shortened.
The main differential diagnosis of hyperostotic lesions include osteoma, osteopoikilosis and osteopathia striata; however, the location of the disease and the cortical thickening are specific to melorheostosis. The cortical changes must be distinguished from periostitis that occurs as a response to an adjacent osseous process (such as neoplasm, infection or trauma) and from hyperostosis that accompanies congenital disorders.

**Differential Diagnosis List:** Melorheostosis

**Final Diagnosis:** Melorheostosis

**References:**

Freyschmidt J.
Melorheostosis: a review of 23 cases.

Greenspan A, Azouz EM.

Boraschi P, Ortori S, Falaschi F, Trippi D.
[The computed tomographic and magnetic resonance aspects of melorheostosis. A report of 2 cases].
Fig. 1

Description: Anteroposterior view; radiographic abnormalities are evident throughout with irregular patchy cortical thickening of the lower extremity of the ulna. The appearance is characteristic; the resemblance to wax dripping is responsible for the term "melorheostosis". Note the usual confinement to one side of the bone. Origin:
Description: Same patient; lateral view demonstrating characteristic cortical involvement. Origin: