Melorheostosis Involvement in the Upper Extremity

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What is Melorheostosis?

The term Melorheostosis comes from Greek origins with "Melos" meaning limb, "Rheo" meaning flowing and "Ostosis" meaning bone formation. Melorheostosis is characterized by thickening of the cortical bone in a pattern that has the appearance of dripping candle wax. This was first discovered by Leri and Janovy in 1922. Melorheostosis typically affects long bones. The lower extremities are more prevalent than the upper extremities. Short bones of the hand and foot may also be affected. Melorheostosis may mimic other conditions such as myositis ossificans, osteoma and panniculitis osteosarcoma. Other conditions associated with Melorheostosis include osteopoikilosis, osteopathia striata, scleroderma and Buschke-Ollendorf syndrome.

Methods

The eligible study cohort was identified by using the Rochester Medical Index database. The Rochester Medical Index classified patients diagnoses by using an internal coding system based on the Hospital Adaption of the International Classification of Diseases, Eighth Revision, (ICDA). The specific ICDA code for Melorheostosis was searched for the time period 1972-2010. Thirty-six cases were identified from this search. The individual charts were reviewed by five of the researchers. The specific data collected from the chart included: gender, age when diagnosed, final diagnosis, areas affected and bones affected. The researchers also identified in chart reviews whether surgery was performed, if occupational therapy, physical therapy, appointment with medical doctor, surgeon, or other rehabilitation professional. The incidence of Melorheostosis is 0.9 in 1 million. This disease can present at any age and both sexes appear equally affected. Onset is usually noted with deformity of the extremity, contracture, pain, stiffness and limited range of motion. Skin can show thickening, shininess, fibrosis, erythema, and linear scleroderma hyperpigmentation. This disease generally appears in late childhood or adolescence. The etiology of Melorheostosis is unknown. Below are two theories for the etiology of Melorheostosis: 1. It is thought to be a congenital lesion of the somatoypes due to the relationship between the distribution of sclerotomes, skeletal regions innervated by a single spinal sensory nerve. Murray and McDermott found a relationship embryonal neuromyopathy that results in a sciometric subtration. Most often in upper extremity patient is the C7 sclerotome was represented in the disease manifestation. 2. Hellenen et al studied the loss of function mutations in LEMD3's involvement in beta bone morphogenic protein signaling. 3

Results

Methods

- A total of 36 charts were identified and reviewed.
- Mean age of the study was 39.3 years.
- Median age was 39 years.
- The youngest patient was 2 to the oldest at 83 years.
- There were 11 males and 25 females identified.
- 13 patient's were identified with upper extremity involvement (Scapula to phalanges).

Impairments and functional limitations were noted in the majority of patients with upper extremity involvement. Although a cause and effect relationship can not be determined, pain, numbness and limited range of motion were noted in patients with Melorheostosis and associated upper extremity soft tissue conditions. The soft tissue associated conditions in combination with Melorheostosis appear more recalcitrant to treatment.

Conclusions

References