Melorheostosis of the Index Finger: A Case Report

Ellen K. Quarles, Michael L. Richardson

We describe a case of melorheostosis involving multiple bones of the left index finger. This patient presented with a many-year history of a finger mass, and demonstrated the typical radiographic findings of melorheostosis on radiographs and computed tomography (CT) of the hand. Following excisional biopsy and bone grafting, this patient is doing well and undergoing physical therapy.

Case Report

A 35 year old left-handed female presented with a many-year history of a mass in her left index finger. She experienced dull discomfort to palpation, and complained of stiffness, but denied any numbness, swelling or instability. She also exhibited full range of motion in her finger. Inspection of the hand revealed a small mass on the ulnar side of the middle phalanx of the left index finger that was firm and bony in nature.

Radiographs displayed dense bone formation involving much of the middle phalanx, with a flowing, slightly expansile margin along its ulnar aspect (Figure 1). Small foci of dense bone were also noted in the proximal end of the distal phalanx, and in the distal end of the proximal phalanx. These areas of osteosclerosis were felt to involve both the cortex and the medullary space in all three sites. In addition to melorheostosis, multiple bone islands and osteopoikilosis were also included in the differential diagnosis. Due to the polyostotic nature of this process, more worrisome entities such as an indolent case of parosteal osteosarcoma were felt to be quite unlikely.

Computed tomography (CT) of the hand (Figure 2) demonstrated findings identical to those shown on the radiographs. The soft tissues were unremarkable, and the visualized tendons followed a normal course. No soft tissue masses were noted. The radiographic and CT findings were felt to be consistent with melorheostosis. While the CT did not change the differential diagnosis, the hand surgeon found it helpful for preoperative planning.

The patient was admitted for surgery, where an excisional biopsy was performed. Dense bone fragments were removed from the interior of the middle phalanx in an effort to reestablish the medullary canal. Allograft bone graft material was then placed in the resection cavity of the bone. Histologic examination of the biopsy specimen showed fragments of dense bone with fibrotic marrow, consistent with melorheostosis. No evidence of active inflammation or neoplasm was noted. The patient has some residual pain and stiffness in her finger following surgery, but is otherwise doing well on physical therapy.

Citation: Quarles EK, Richardson ML. Melorheostosis of the index finger: a case report. Radiology Case Reports. [Online] 2008;3:140.
Copyright: © 2008 Michael L. Richardson. This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs 2.5 License, which permits reproduction and distribution, provided the original work is properly cited. Commercial use and derivative works are not permitted.

Abbreviations: CT, computed tomography; MR, magnetic resonance

Ellen C. Quarles (Email: ekayt@u.washington.edu) is in the Department of Pathology, University of Washington School of Medicine, 1956 NE Pacific Street, Seattle, WA, 98195, United States of America.

Michael L. Richardson (Email: mrich@u.washington.edu) is in the Department of Radiology, University of Washington School of Medicine, 4245 Roosevelt Way NE, Seattle, WA 98105, United States of America.

Published: January 24, 2008
DOI: 10.2484/rcr.v3i1.140
Melorheostosis of the Index Finger: A Case Report

Figure 1. AP (A) and lateral (B) radiographs of the left index finger, demonstrating dense bone formation in all three phalanges. The dominant lesion involves the middle phalanx with smaller non-contiguous lesions in the proximal and middle phalanges. Both the cortex and the medullary space are involved by this process in all three sites.

Discussion

Melorheostosis was first described by Léri and Joanny in 1922 (1). This name comes from the Greek melos, meaning limb and rhein, meaning to flow. The radiologic appearance has been likened to “melting candle wax” (2). Synonyms for melorheostosis include Léri’s Disease and flowing periosteal hyperostosis.

Melorheostosis is an idiopathic disorder that does not appear to be hereditary. Males and females are equally affected (2). Melorheostosis is a rare disorder, with a prevalence of about 0.9 per million (3). It is usually first diagnosed as an incidental finding on plain radiographs. Approximately half of the diagnosed cases are found in patients 20 years of age or younger (2, 4).

Melorheostosis typically affects long bones of the limbs and sometimes the small bones of the hands and feet. Extremely rarely, melorheostosis involves the vertebral column (5, 6). Melorheostosis is almost always unilateral, with only six cases of bilateral involvement reported (7). The characteristic hyperostosis of melorheostosis is distributed longitudinally along one or more sides of a single bone or set of contiguous bones (5). The hyperostosis seen in our patient followed this longitudinal pattern along the ulnar side of the phalanges of the left index finger.

Plain radiographs of melorheostosis usually demonstrate hyperostosis of either the osseous cortex, the medullary space or both (8). The endosteal space may be obliterated in some cases. Cortical hyperostosis may be accompanied by intervening soft-tissue calcification or ossification (6, 8). Contiguous bones of an extremity are often involved, although monostotic involvement has also been reported.

If radionuclide imaging is performed, it typically demonstrates multiple areas of increased uptake, corresponding to the areas of sclerosis noted on radiographs (9, 10). This may be helpful in distinguishing melorheostosis from other hyperostotic lesions of bone such as osteopoikilosis or osteopathia striata, which usually appear normal on radionuclide imaging (10).

CT and magnetic resonance (MR) imaging can also be used to demonstrate the osseous and extraosseous findings of melorheostosis (6, 11). CT reveal foci of hyperostosis and soft tissue calcification as areas of high attenuation, while MR demonstrates these same foci as areas of low signal intensity on all pulse sequences (6, 11). Less well-calcified soft tissues may appear higher in signal intensity and even enhance following intravenous gadolinium administration (11). Cross-sectional imaging may help to accurately localize these findings, particularly in areas of complex anatomy such as the spine. In our case, the CT did not change the differential diagnosis, but assisted the hand surgeon in his preoperative planning.

The most common complaints include dull pain, stiffness, as with our patient, decreased joint motion, and paresthesias over the involved bones (12). While some patients with melorheostosis are asymptomatic, oth-
ers demonstrate severe contraction deformities and pain (7). The symptoms and physical impairment caused by melorheostosis vary considerably in severity, and may also include limb swelling, soft tissue masses and limb deformation (4). Soft tissue involvement, especially periosseous fibrosis, is common (5, 13, 14).

Other associated abnormalities have been reported, including neurofibromatosis, soft tissue calcification, skin pigmentation, cutaneous hemangiomas, lymphatic vessels, linear scleroderma, arteriovenous malformations, and arterial aneurysms (4, 13). However, an associations with vascular malformations is considered rare (15).

Melorheostosis remains an idiopathic disorder -- no certain etiology or pathogenesis has yet been found. However, one possible clue its pathogenesis is that the margins between affected and unaffected bone in melorheostosis are usually somewhat abrupt. The hyperostotic linear areas seen in the affected bones tend to have a segmental distribution that does not correspond with the arterial blood supply or innervation of the limbs. One possible explanation for this phenomenon and prominent theory of the origin of the disease was advanced by Inman and Saunders in 1944 (16) and furthered by the work of Murray and McCredie in 1979 (17). These authors have suggested that the lesions of melorheostosis may be due to damage to spinal sensory nerves in early embryonic life. These authors presented evidence that affected areas of the skeleton corresponded to sclerotomes, i.e. regions of bone and periosteum that are hypothetically innervated by a single spinal segment (17). In our patient, the lesion in the index finger would correspond to the proposed sclerotome of C7. However, the very existence of sclerotomes remains somewhat controversial (4, 18).

Treatment of melorheostosis depends upon the exact symptoms and findings exhibited by a given patient. Surgical excision or resection is sometimes used to increase range of motion in joints of patients with bone enlargement or soft tissue calcification. Other reported treatments include: osteotomies and bone lengthening for patients with limb shortening or angulation deformities, surgery for ankylosis, tendon lengthening, limb lengthening, excision of fibrous tissue, fasciotomies, capsulotomies, osteotomies, excision of hyperostoses, arthrodesis, contralateral epiphysiodesis, sympathectomies and amputation (7, 19). Non-surgical treatments have included diphosphonates, non-steroidal anti-inflammatory medications, analgesics, nifedipine, physical therapy, manipulations, braces, serial casting, and nerve blocks (7, 19).

Unfortunately, neither conservative nor surgical methods of treatment appear to be very effective. Following surgery, many patients typically demonstrate recurrence of their disease (20). Although the affected areas progressively worsen throughout the lifetime of the patients, the rate of progression tends to decline as the individuals age. Surgery
was chosen as the best course of action in our 35 year-old patient, with the hope that recurrence and subsequent progression of the disease will be slow in her case.

References

1. Léri A, Joanny L. Une affectation non decrite des os: hyperostose “en coulee” sur toute la hauteur d’un membre ou melorrheostose. Bull Mem Soc Med Hop Paris 1922; 46:1141-1145.

2. Kalbermatten NT, Vock P, Rufenacht D, Anderson SE. Progressive melorheostosis in the peripheral and axial skeleton with associated vascular malformations: imaging findings over three decades. Skeletal Radiol 2001; 30:48-52. [PubMed]

3. Wynne-Davies R, Gormley J. The prevalence of skeletal dysplasias. An estimate of their minimum frequency and the number of patients requiring orthopaedic care. J Bone Joint Surg Br 1985; 67:133-137. [PubMed]

4. Freyschmidt J. Melorheostosis: a review of 23 cases. Eur Radiol 2001; 11:474-479. [PubMed]

5. Garver P, Resnick D, Haghighi P, Guerra J. Melorheostosis of the axial skeleton with associated fibrolipomatous lesions. Skeletal Radiol 1982; 9:41-44. [PubMed]

6. Motimaya AM, Meyers SP. Melorheostosis involving the cervical and upper thoracic spine: radiographic, CT, and MR imaging findings. AJNR Am J Neuroradiol 2006; 27:1198-1200. [PubMed]

7. Rozencwaig R, Wilson MR, McFarland GB, Jr. Melorheostosis. Am J Orthop 1997; 26:83-89. [PubMed]

8. Levine SM, Lambiase RE, Petchprapa CN. Cortical lesions of the tibia: characteristic appearances at conventional radiography. Radiographics 2003; 23:157-177. [PubMed]

9. McCay T, Mulvihill J, Leonard JC. Multifocal melorheostosis. Clin Nucl Med 2006; 31:504-505. [PubMed]

10. Whyte MP, Murphy WA, Siegel BA. 99mTc-pyrophosphate bone imaging in osteopoikilosis, osteopathia striata, and melorheostosis. Radiology 1978; 127:439-443. [PubMed]

11. Judkiewicz AM, Murphey MD, Resnik CS, Newberg AH, Temple HT, Smith WS. Advanced imaging of melorheostosis with emphasis on MRI. Skeletal Radiol 2001; 30:447-453. [PubMed]

12. Hove E, Sury B. Melorheostosis. Report on 5 cases with follow-up. Acta Orthop Scand 1971; 42:315-319. [PubMed]

13. Campbell CJ, Papademetriou T, Bonfiglio M. Melorheostosis. A report of the clinical, roentgenographic, and pathological findings in fourteen cases. J Bone Joint Surg Am 1968; 50:1281-1304. [PubMed]

14. Morris JM, Samilson RL, Corley CL. Melorheostosis. Review of the literature and report of an interesting case with a nineteen-year follow-up. J Bone Joint Surg Am 1963; 45:1191-1206. [PubMed]

15. Roger D, Bonnetblanc JM, Leroux-Robert C. Melorheostosis with associated minimal change nephrotic syndrome, mesenteric fibromatosis and capillary haemangiomas. Dermatology 1994; 188:166-168. [PubMed]

16. Inman V, Saunders J. Referred pain from skeletal structures. J Nerv Ment Dis 1944; 99:660-667.

17. Murray RO, McCredie J. Melorheostosis and the sclerotomes: a radiological correlation. Skeletal Radiol 1979; 4:57-71. [PubMed]

18. Ivanusic JJ. The evidence for the spinal segmental innervation of bone. Clin Anat 2007; 20:956-960. [PubMed]

19. Perlman MD. Melorheostosis: a case report and literature review. J Foot Surg 1990; 29:353-356. [PubMed]

20. Pruitt DL, Manske PR. Soft tissue contractures from melorheostosis involving the upper extremity. J Hand Surg [Am] 1992; 17:90-93. [PubMed]