Imaging of Melorheostosis: Emphasis on MR Imaging Findings

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Purpose: To evaluate the usefulness of various radiographic imaging modalities in the diagnosis and characterization of melorheostosis.

Materials and Methods: We retrospectively evaluated the plain film (n=8), computed tomographic (CT) imaging (n=5) and magnetic resonance (MR) imaging (n=5) findings of eight patients with melorheostosis diagnosed by bone biopsy (n=4) and characteristic radiographic findings (n=8). MR images were obtained with a 1.5-T scanner focused on the region of maximal radiographic abnormality. Pulse sequences include T1-weighted SE, T2-weighted fast SE (n=5) and postcontrast imaging (n=4). In order to define subtle enhancement of the lesions, subtraction MR images were obtained in one case. Imaging findings were analyzed with particular emphasis on the distribution of lesions along the sclerotome, differential radiographic findings between diaphyseal and metaepiphyseal lesions of the long bones, as seen on plain radiographs, and the density and signal characteristics of hyperostotic lesions, as seen on CT and MR images.

Results: Characteristic distribution along the sclerotome was identified in five of eight cases mainly along C6 and 7 (n=2) and L3, 4 and 5 (n=3) sclerotomes. In diaphyseal melorheostosis (8/8), a characteristic finding, i.e., a wax flowing down from the candle, was identified on plain radiographs. In all three patients with metaepiphysal melorheostosis (3/8), multiple round or oval hyperostotic lesions were seen in the epiphysis and metaphysis of the long bones. On CT, the marrow cavity was partly obliterated by hyperostotic lesions in all five patients with endosteal hyperostosis. Among these, central ground glass opacity with a sclerotic rim was seen in three patients. Periosteal hyperostosis was seen in two of five cases, being visualized as irregular excrescences in the periosteal region and surrounding soft tissue. Individual hyperostosis was visualized as hypointense on T1-weighted images and as a hyperintense center with a surrounding hypointense rim on T2-weighted images (5/5). On postcontrast images, central enhancement was noted in all four cases. In one of these, in which the degree of central enhancement was subtle, subtraction images (postcontrast SE - precontrast SE) also revealed a central signal increment. Central enhancement corresponded to the hyperintense center seen on T2-weighted images (4/4) and the ground-glass opacity seen on CT (2/2).

Conclusion: Radiographic imaging plays a crucial role in the diagnosis of melorheostosis. The future role of gadolinium-enhanced MR imaging in the characterization of the lesion may be important though further evaluation and pathologic correlation is required.

Index words: Bones, osteochondrodysplasia
Bones, radiography
Bones, CT
Bones, MR

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Melorheostosis is a rare non-inheritable disorder characterized by a flowing hyperostosis of the cortex affecting a single limb and first described by Leri and Joanny in 1922 (1). Diagnosis has been based on characteristic plain radiographic findings; to our knowledge, MR imaging findings have been reported in only two cases and contrast-enhanced MR imaging findings in only one (2, 3).

We describe the plain film, CT and MR imaging findings of melorheostosis, having evaluated the usefulness of radiographic imaging in the diagnosis and characterization of melorheostosis with particular emphasis on contrast-enhanced MR imaging findings.

Materials and Methods

Between 1987 and 1998, eight patients (six males and two females, aged 3-55 [mean, 38.3] years) were found to be suffering from melorheostosis. Diagnosis was on the basis of characteristic radiographic findings (n=8) and the histopathologic findings of excisional bone biopsy (n=4).

CT (n=5) was performed with a HiSpeed Advantage scanner (GE Medical Systems, Milwaukee, U.S.A.) using the following parameters: 5-mm slice thickness, 10-mm interslice gap, 120 kVp, 280 mA, 10-mm collimation, a helical pitch of 1:1, and 25-cm field of view.

For MR imaging (n=5), a 1.5-T unit (Signa, GE Medical Systems, Milwaukee, U.S.A.) was used. Depending on the location of the lesions, a phased-array extremity coil was sometimes employed. Imaging sequences included spin-echo (SE) and fast spin-echo (FSE) techniques, with the following acquisition parameters: 400-600/10-12 (repetition time msec/echo time msec) for T1-weighted SE images; 2,500-3,500/96-108 (repetition time msec/effective echo time msec) with an echo train length (ETL) of eight for T2-weighted FSE images, and 350-600/10-12 (repetition time msec/echo time msec) for gadolinium-enhanced T1-weighted images. The fat-suppression technique based on frequency-selective excitation was used in T2-weighted FSE and gadolinium-enhanced T1-weighted SE imaging. For both T1- and T2-weighted images, slice thickness of 3.0-7.0 mm with a 0-2.5 mm interslice gap, number of excitation (NEX) of two to four and matrix number of 256x192 were used. Field of view (FOV) was 180 to 340 mm, depending on body size and section planes. Axial and coronal or sagittal images were obtained for each pulse sequence; contrast-enhanced T1-weighted images were obtained 20 seconds after intravenous injection of gadopentetate dimeglumine at a dose of 0.1 mmol/kg of body weight. In order to define lesion enhancement, subtraction MR images (postcontrast SE - precontrast SE) on Advantage Windows workstation (GE Medical systems, Milwaukee, U.S.A.) was used. CT and MR imaging focused on the regions of maximal abnormality. Plain radiographs were available for all patients.

Imaging findings were analyzed with particular emphasis on the distribution of lesions along the sclerotive, as seen on plain films, and the density and signal characteristics of hyperostotic lesions, as seen on CT and MR images.

Results

Plain Radiographic Findings

In all eight patients with diaphyseal melorheostosis, a characteristic finding, i.e., wax flowing down the side of a candle, was identified on plain radiographs (Fig. 1). In all three cases of metaepiphysial melorheostosis, multiple round or oval hyperostotic lesions were seen in the epiphysis and metaphysis of the long bones. In one case, combined osteopathia striata was identified in the ilio-pubic bone.

There were six cases of endosteal hyperostosis, in which the lesion does not extend through the periosteum, and two of the periosteal type, in which bony excrescences are seen along the periosteum. A 50-year-old male patient with extensive periosteal hyperostosis underwent surgery for progressive limb deformity.

In five of five cases, scintigraphy revealed increased uptake in the involved bones. Clinical and imaging data are summarized in the accompanying table.

CT Findings

All lesions in patients who underwent CT showed cortical thickening. In one case, abnormal dense cortical thickening scalloping normal cortex was seen. The lesions partially obliterated the marrow cavity, especially in endosteal-type cases. Central ground-glass opacity with a surrounding sclerotic rim was identified on CT in three of five patients (Fig. 1F). Extensive cauliflower-like bony excrescences are noted in one periosteal type.

MR Findings

All five lesions for which MR images were obtained showed low signal intensity on T1-weighted SE images, and central high signal intensity with a low signal inten-
Fig. 1. Images in a three-year-old female with involvement of the right lower extremity.

A. Plain radiograph shows patchy linear hyperostosis in the right pubis (black arrow) and endosteal hyperostosis in the medial side of right femur (white arrow).
B. The diaphysis of the right tibia reveals characteristic wavy hyperostosis flowing down the side.
C. Axial T1-weighted SE image (550/15) of the right tibia shows endosteal hyperostosis of low signal intensity.
D. Axial fat-suppressed T2-weighted FSE image (3,700/100) demonstrates the central high signal intensity (arrow) with surrounding rim of low signal intensity.
E. Postcontrast axial T1-weighted SE image (350/15) with fat suppression reveals central enhancement (arrow) of the lesion.
F. CT shows the central ground-glass opacity with sclerotic rim (arrow) at the comparable site of the MR images.
G. Postcontrast sagittal T1-weighted SE image (550/14) with fat suppression of the right foot also demonstrates central enhancement (arrow) of the round or oval lesions in the talus and medial cuneiform.
H. Photomicrograph of the abnormal cortex reveals irregularly thickened tabeculae and some fibrovascular tissues (Hematoxylin-eosin stain; original magnification, × 100)
ity rim on T2-weighted FSE images (Fig. 1, 2). On postcontrast MR images, central enhancement with a low signal intensity peripheral rim was identified in two children and two adults. Signal increment was noted in the center of the lesions on the subtraction images obtained in the case of a 45-year-old male patient (Fig. 2E). In two children, central ground-glass opacity on CT images coincided with central enhancement on postcontrast MR images (Fig. 1E,F).

**Pathologic Findings**

The microphotographs obtained by excisional bone biopsy revealed irregularly thickened trabecular bones and some fibrovascular tissues (Fig. 1H). Osteoclastic or osteoblastic activity was not seen.

**Discussion**

Melorheostosis is a rare benign sclerosing bony dysplasia characterized by linear hyperostosis of cortical bone and joint abnormalities, and affects children and adults. Because diagnosis is based on the presence of characteristic radiographic features rather than the histopathology of the lesions, radiographic imaging plays an important role in the diagnosis of this condition (4).

The etiology of melorheostosis is not known. According to Zimmer (5), the pattern of involvement indicates that it arises from maldevelopment of the limb bud caused by metameric disturbance. Campbell (6) believes that the disorder affects not only bone but also many tissues of mesodermal origin and that the marked soft tissue changes found at birth tend to support Zimmer’s concept. There is, however, no evidence that melorheostosis is a genetic disorder (7). Although most investigators believe it is congenital, symptoms do not manifest until late childhood or early adolescence (4).

Melorheostosis usually involves a single limb particularly the lower extremity, though in rare instances, more than one limb is affected. The pelvis, skull, spine, and facial bone may also be involved (6, 8-10). Murray and McCredie (11) have suggested that the skeletal lesions may represent the late result of a segmental sensory nerve lesion. The typical distribution of hyperostosis corresponds to a single sclerotome (a zone supplied by an individual spinal sensory nerve), or part of one; the skeletal lesions seen in all our patients corresponded to sclerotomes or a part of one. Two children showed the same distribution of lesions.

Hyperostosis tends to progress through childhood and the disease exhibits a slow, chronic course with periods of exacerbation and arrest (6). Most evidence of progression of the condition is based on the increased hyperostosis seen on radiographs obtained at periodic intervals, and on the findings of several biopsies. It can also progress in adults, manifesting as an aggravation of symptoms, soft-tissue abnormalities, and bone involvement, although in adults the process is not as active or rapidly progressive as in children (6). We observed the progression of this disease in a 45-year-old male patient (Fig. 2E).

**Table: Clinical and Imaging Data in Patients with Melorheostosis**

<table>
<thead>
<tr>
<th>No</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>CT findings</th>
<th>MR findings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>T1WI</td>
</tr>
<tr>
<td>1</td>
<td>39</td>
<td>F</td>
<td>Varus deformity</td>
<td>Right pubis, femur, tibia, talus, cuneiform, phalanges</td>
<td>Low SI Central high SI</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>M</td>
<td>Bowing</td>
<td>Right pubis, femur, tibia, talus, cuneiform, phalanges</td>
<td>Low SI Central high SI</td>
</tr>
<tr>
<td>3</td>
<td>34</td>
<td>F</td>
<td>Incidental</td>
<td>Left ulnar</td>
<td>Central thickening</td>
</tr>
<tr>
<td>4</td>
<td>40</td>
<td>M</td>
<td>Incidental</td>
<td>Left scapula, rib, clavicle, humerus, radius, carpal bones, phalanges</td>
<td>Central thickening with ground-glass opacity</td>
</tr>
<tr>
<td>5</td>
<td>45</td>
<td>M</td>
<td>Joint pain</td>
<td>Right tibia</td>
<td>Low SI Central high SI</td>
</tr>
<tr>
<td>6</td>
<td>35</td>
<td>F</td>
<td>Deformity</td>
<td>Right scapula, calvicle, humerus, radius, ulnar, carpal bones, phalanges</td>
<td>Central thickening with bony excrescences</td>
</tr>
<tr>
<td>7</td>
<td>35</td>
<td>F</td>
<td>Joint pain</td>
<td>Right Phalanges</td>
<td>Central high SI</td>
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<tr>
<td>8</td>
<td>35</td>
<td>M</td>
<td>Incidental</td>
<td>Right femur, patella, tibia, phalanges</td>
<td>Central high SI</td>
</tr>
</tbody>
</table>

T1WI = T1-weighted images, T2WI = T2-weighted images, SI = signal intensity

*This case was combined with osteopathia striata
disorder in a 50-year-old male who underwent surgery for limb deformity.

The features of the disease in children are slightly different from those in adults. Clinically, pain occurs infrequently in children and is not intense; radiographically, the endosteal type is frequently reported in childhood though it can also be seen in adults (12).

Melorheostosis has been reported in association with other disorders including linear scleroderma, osteopoikilosis, osteopathia striata, neurofibromatosis, tuberous sclerosis and hemangioma or other vascular lesions (13-19). The entity was described by Walker in 1964 as mixed sclerosing bone dystrophy (20). In one of our cases, melorheostosis was combined with osteopathia striata in the pubic bone, but linear scleroderma or other skin change was not seen in our patient.

The histologic features of melorheostosis have been reported by a number of authors (4, 11, 17). Microscopic examination of cortical specimens reveals nonspecific, hyperostotic, periosteal bone exhibiting thickened trabeculae and some fibrovascular tissues in the marrow spaces. The immature bone consists largely of primary haversian systems, particularly on the periosteal surface, that are almost obliterated by the deposition of sclerotic, thickened, and somewhat irregular lamellae. Osteoclastic or osteoblastic activity can be noted but is never prominent.

Roentgenographic findings have on occasion been used for diagnosis. Irregular, dense, eccentric hyperostosis affecting both the cortex and adjacent medullary canal of a single bone, or one side of the limb, like melted wax dripping down the side of a candle, is highly characteristic. In the epiphysis of long bones, more dis-
crete round foci may resemble the findings of osteopoikilosis, whereas in flat bones such as those in the pelvis, or the scapula, radiating or localized sclerotic patches are seen (21).

CT findings are described as simple cortical thickening and osseous excrescence on the external and/or internal cortical surfaces (21). On occasion, these changes partly or completely obliterate the marrow cavity, particularly in the endosteal type. In two children and two adults, we found that density was less than that of normal cortex in the endosteal area occupying the marrow cavity just beneath the thickened cortex. This ground-glass opacity was centrally located and surrounded by a sclerotic rim. We speculated that the difference in density between the abnormal dense cortex and the central ground glass opacity in the diaphysis was either due to the varying amounts of intramembranous ossification found along the periosteum, or the process of change of the lesion into a mature dense cortex. Greenspan (4) also believes that radiographic findings appear to reflect developmental errors at the sites of intramembranous and endochondral bone formation, predominantly the former.

To our knowledge, MR imaging findings of melorheostosis have been published in only two cases (2, 3), only one of which included the findings of postcontrast MR imaging (3). They found that a metaphyseal lesion of the distal femur showed low signal intensity on both T1- and T2-weighted images, with peripheral rim enhancement, speculating that the lesion was one which was less mature and the enhancing rim was consistent with the vascular fibrous reaction about centrally located aberrant bony proliferation (3).

Empirically, we had expected that on MR images, cortical hyperostosis might show low signal intensity on all pulse sequences, but on T2-weighted images, all the lesions in fact showed central high signal intensity, with a low signal intensity rim. Unexpectedly, we experienced central enhancement on postcontrast images in four patients for whom contrast-enhanced MR images were available. In one case, in which subtle central enhancement was observed, subtraction imaging also revealed central signal increment.

In our study, the central portion of the lesions showing contrast enhancement was in two cases in which both CT and MR findings were available - comparable to the area of central high signal intensity seen on T2-weighted images and the ground-glass opacity seen on CT. We considered that this central enhancement was caused by fibrovascular reaction about the thickened rim of trabecular bones. Interestingly, all the central ground-glass opacity seen on CT, and the central enhancement seen on postcontrast MR images, was found only in children and symptomatic young adults. In view of these clinical findings, central enhancement seems to be related to disease activity.

In conclusion, radiographic imaging plays a crucial role in the diagnosis of melorheostosis. The central ground-glass opacity seen on CT and central enhancement seen on postcontrast MR images are probably related to lesion activity, though further study is required.

References

 Raptors 2009-2010 Season Ticket

- Home Games
  - March 3 - April 10
  - Game 1
  - Game 2
  - Game 3
  - Game 4
  - Game 5
  - Game 6
  - Game 7
  - Game 8

- Away Games
  - February 28
  - March 1
  - March 3
  - March 4
  - March 5
  - March 7
  - March 8
  - March 9

- Preseason Games
  - October 16
  - October 18
  - October 20

- Postseason Games
  - April 25
  - April 27
  - April 29
  - May 1

- Other Games
  - October 22
  - October 24
  - October 26
  - October 28
  - November 1
  - November 3
  - November 5
  - November 7
  - November 9
  - November 11
  - November 13

- Special Games
  - December 1
  - December 3
  - December 5
  - December 7
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  - December 11
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