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CASE REPORT

Melorheostosis Involving the Cervical and Upper Thoracic Spine: Radiographic, CT, and MR Imaging Findings

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SUMMARY: Melorheostosis, an uncommon mesenchymal dysplasia, rarely affects the axial skeleton. We describe the imaging findings of melorheostosis involving the cervical and upper thoracic spine. Radiographs and CT showed unilateral well-marginated undulating zones of cortical hyperostosis involving multiple vertebrae that were contiguous with a coalescent ossified right paravertebral mass. MR imaging showed zones of signal intensity void on all pulse sequences without contrast enhancement. Conservative management was elected because of lack of interval clinical and imaging changes for 8 years.

Melorheostosis (Leri disease) is a rare nonhereditary sclerosing mesenchymal dysplasia of bone originally described more than 80 years ago.[1] This dysplasia occurs in early childhood[2] and in 40%–50% of cases is evident by 20 years of age.[2-5] The bony lesions commonly present as wavy longitudinal bars of hyperostosis resembling molten wax flowing down one side of a candle[6]; hence, the common reference to this abnormality as having a "flowing candle wax" appearance.[7-10] One or multiple adjacent bones are often affected, often in a sclerotomal distribution.[7,11-13] Melorheostosis commonly involves limb bones and may extend across synovial joints.[14-16] Lesions may be associated with ossification in adjacent soft tissues.[14-16] Melorheostosis limited to the spine is rare.[2,17] Melorheostosis affecting thoracic vertebrae with involvement of facet joints associated with back pain has been described recently.[14] The current report describes the radiographic, CT, and MR imaging features of melorheostosis involving multiple lower cervical and T1 vertebrae.

Case Reports

Eight years ago, a 46-year-old man presented with neck pain after minor trauma. Cervical spine radiographs showed a slight segmental kyphosis from the C4 level to the C7 level and a densely sclerotic right-sided vertebral and paravertebral abnormality that extended from the C5 to the T1 level (Fig 1*A*, *-B*). CT scans showed a high-attenuation undulating cortical hyperostosis with a "dripping candle wax appearance" involving the right and/or dorsal aspects of the C5, C6, C7, and T1 vertebral bodies, right lamina, and transverse processes of the C4, C5, C6, and C7 vertebrae; right C5 and C6 pedicles; and C4, C5, and C6 spinous processes. The unilateral osseous abnormality narrowed and partially obliterated the right C5–C6 and C6–C7 facet joints (Fig 1*C, -D, -G*). The enlarged portions of the vertebrae from the hyperostosis caused right foraminal narrowing at the C5–C6 and C6–C7 levels and caused deviation of the cervical spinal cord to the left of midline. The right-sided hyperostosis at the involved vertebrae extended laterally to form a large, coalescent ossified mass in the right paravertebral region. The intervertebral disks were decreased in height but not infiltrated by the hyperostosis. Radionuclide bone scans showed intense radiotracer uptake within the right lateral aspect of the C5–T1 vertebrae without any other focal abnormality. At the time of imaging, the differential diagnosis for these findings included tumoral calcinosis, hetertopic ossification, ivory osteomas, and melorheostosis. Follow-up examination after 1 year revealed no change in the CT findings. The patient was lost to follow-up.

Seven years later, the patient returned after trauma related to a motor vehicle crash with complaints of stiffness and worsening neck pain, especially involving the right paraspinal soft tissues. Radiographs showed no interval change in the size or configuration of the vertebral and right paravertebral hyperostosis compared with the prior radiographic examinations. MR imaging was performed to assess for ligamentous injury. There was no evidence of increased signal intensity on fat-suppressed long-TR/long-TE images in the interspinous ligaments or paraspinal soft tissues excluding ligamentous injury. MR imaging showed zones of signal intensity void on all pulse sequences corresponding to sites of hyperostosis seen on radiographs and CT (Fig 1*E, -F, -H, -I*). The hyperostosis predominantly involved the outer cortical surface. Hyperostosis also involved the endosteum with marrow space encroachment; however, the marrow adjacent to sites of hyperostosis had normal signal intensity on all pulse sequences. After gadolinium contrast administration, no abnormal enhancement was seen within the hyperostotic lesions. The diagnosis of melorheostosis was made on the basis of the characteristic distribution, location, and combined radiographic, CT, and MR imaging features of the abnormalities.

Discussion

Melorheostosis is a rare benign sclerosing bone dysplasia of unknown etiology that localizes in regions of innervation of spinal sensory nerves and correlates with those sclerotomes and myotomes.[18,19] This disorder tends to be segmental and unilateral and may affect only one bone (monostotic), one limb (monomelic), or multiple bones (polyostotic).[1,2,20] The lower extremity is more frequently involved than the upper extremity.[1,2,20] Melorheostosis rarely involves the spine, skull, and facial bones.[1,2,20] Lesions may occasionally be associated with skin lesions, vascular anomalies, and joint contractures,[2] though they were not present in this reported case. Histologic findings include variable degrees of cortical thickening consisting of chondroid islands surrounded by mature lamellar and woven bone, as well as adjacent zones of fibrocartilage with irregular surface fibrillation.[14,17,19] Soft tissue abnormalities consisting of osseous, chondroid, vascular, and fibrocartilaginous tissue have been reported in ≤76% of cases of melorheostosis.[7,10,14]

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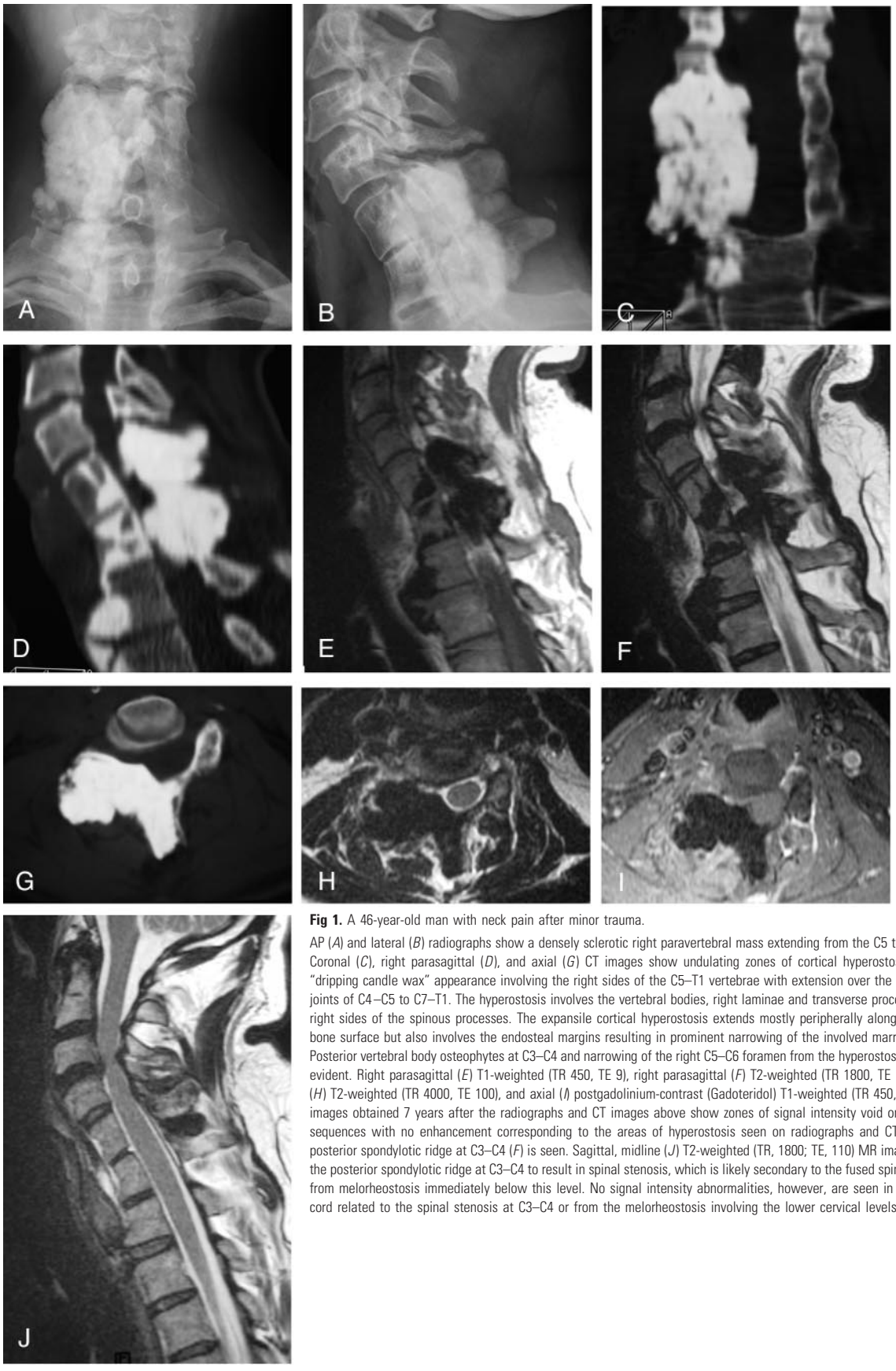

Fig 1. A 46-year-old man with neck pain after minor trauma.

AP (*A*) and lateral (*B*) radiographs show a densely sclerotic right paravertebral mass extending from the C5 to T1 level. Coronal (*C*), right parasagittal (*D*), and axial (*G*) CT images show undulating zones of cortical hyperostosis with a "dripping candle wax" appearance involving the right sides of the C5–T1 vertebrae with extension over the right facet joints of C4–C5 to C7–T1. The hyperostosis involves the vertebral bodies, right laminae and transverse processes, and right sides of the spinous processes. The expansile cortical hyperostosis extends mostly peripherally along the outer bone surface but also involves the endosteal margins resulting in prominent narrowing of the involved marrow space. Posterior vertebral body osteophytes at C3–C4 and narrowing of the right C5–C6 foramen from the hyperostosis are also evident. Right parasagittal (*E*) T1-weighted (TR 450, TE 9), right parasagittal (*F*) T2-weighted (TR 1800, TE 110), axial (*H*) T2-weighted (TR 4000, TE 100), and axial (*I*) postgadolinium-contrast (Gadoteridol) T1-weighted (TR 450, TE 9) MR images obtained 7 years after the radiographs and CT images above show zones of signal intensity void on all pulse sequences with no enhancement corresponding to the areas of hyperostosis seen on radiographs and CT scans. A posterior spondylotic ridge at C3–C4 (*F*) is seen. Sagittal, midline (*J*) T2-weighted (TR, 1800; TE, 110) MR image shows the posterior spondylotic ridge at C3–C4 to result in spinal stenosis, which is likely secondary to the fused spinal column from melorheostosis immediately below this level. No signal intensity abnormalities, however, are seen in the spinal cord related to the spinal stenosis at C3–C4 or from the melorheostosis involving the lower cervical levels.

The differential diagnosis for superficial hyperattenuated vertebral and adjacent paraspinal abnormalities includes melorheostosis, tumoral calcinosis, tumoral calcium pyrophosphate dihydrate deposition disease (CPPD), ivory osteomas, heterotopic ossification (myositis ossificans), and parosteal and periosteal osteosarcoma. The unilateral and multifocal cortical locations, distinct imaging features, and lack of interval change for the abnormalities in this report are, however, characteristic for melorheostosis and are identical to prior reported imaging findings for biopsy-confirmed melorheostosis involving lower thoracic and lumbar vertebrae.[14,17] The imaging features of this case are also sufficiently different from the other disorders in the differential diagnosis. For example, tumoral calcinosis of the spine, which is often associated with systemic disorders of calcium metabolism or renal dialysis, typically occurs as high attenuation paraosseous lesions that result from dystrophic calcifications in soft tissues composed of calcium hydroxyapatite crystals, collagenous fibrous septa, and collections of histiocytes and foreign body giant cells.[21-23] Unlike melorheostosis, tumoral calcinosis of the spine is often not unilateral and appears primarily as extradural lesions with heterogeneous mixed signal intensity on both T1-weighted and T2-weighted images, which are often associated with erosion of adjacent bone.[21-23] CPPD can occur as tumor-like lesions in the spine, typically located at the ligamentum flavum and synovial joints.[24] CPPD lesions contain needle- and rhomboid-shaped calcium pyrophosphate crystals with associated chondroid elements, fibrocollageneous tissue, and variable amounts of acute and chronic inflammatory reaction.[24] CPPD lesions may cause erosion of adjacent bone.[24] The MR imaging findings of spinal CPPD are also similar to those for tumoral calcinosis, thus differing from those for melorheostosis.[2,14,21-24] Although ivory osteomas can have histologic features similar to melorheostosis, large osteomas of the spine are very rare and have been reported to involve only single vertebra.[25] Heterotopic ossification, also referred to as myositis ossificans, typically occurs as lesions in soft tissue with predominant peripheral distributions of ossification and thus differs from the appearance of cortical-based hyperostosis seen with melorheostosis.[26] The imaging features of parosteal and periosteal osteosarcomas also differ markedly from melorheostosis because these tumors contain ossific mineralized matrix that is often irregular and not uniformly attenuated on radiographs and have focal or poorly defined soft-tissue masses with high signal intensity on T2-weighted images.[27,28] Tumor invasion into medullary bone with high signal intensity on T2-weighted images occur in 41% of low-grade and 50% of high-grade parosteal osteosarcomas.[27] Finally, the juxtacortical soft tissue masses of periosteal osteosarcomas typically cause extrinsic erosion of cortical bone, perpendicular periosteal reaction, and reactive marrow zones with high signal intensity on T2-weighted images, which are findings not seen with melorheostosis.[28]

Although radiographic and bone scintigraphic appearance of melorheostosis has been well described (ie, undulating cortical thickening and marked increased uptake of radionuclide[7]), CT and MR imaging helps confirm and accurately localize the zones of hyperostosis in the spine and provide assessment of the degrees of narrowing of the spinal canal and foramina. Further, even though the MR imaging appearance of soft tissue masses associated with melorheostosis is variable, mineralized and nonmineralized soft tissue abnormalities should be recognized as another manifestation of this disease.[7,14] MR imaging aids in confirmation of the diagnosis and in the accurate detection and determination of the extent of soft tissue involvement.

Although melorheostosis is a rare condition affecting the axial skeleton, it should be a definite consideration in the differential diagnosis of unilateral or segmental lesions of cortical hyperostosis in the spine because accurate detection can prevent an unwarranted biopsy.

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