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INCIDENTAL AXIAL MELORHEOSTOSIS OF THE THORACIC VERTEBRA AND RIB IN AN ASYMPTOMATIC ELDERLY PATIENT: A MULTIMODALITY IMAGING CASE REPORT

Dr. L.N. Moni Sri, MD[1], Dr. G. Murugan, MD[2], Dr. Ashwin Kumar A[3*], Dr. A. Baskar[4], Dr. Vettri Vignesh[5], Dr. L.N. Bhavya Sri[6]

[1]Postgraduate Resident (2nd Year), Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India. Email: ln.monisri@yahoo.com
[2]Professor and Head of the Department, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India.
Email: Dr.gmurugan@gmail.com
[3]Associate Professor, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India, Email: ashgilli@gmail.com
[4]Associate Professor, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India, Email: Baskarradio@sbmch.ac.in
[5]Assistant Professor, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India, Email: doctorvettri@gmail.com
[6]Fellowship in Onco-Imaging, Tata Medical Centre, Kolkata. Email: Lnbhavyasri@gmail.com

Abstract

Melorheostosis is a rare benign sclerosing bone dysplasia typically affecting the appendicular skeleton in a sclerotomal distribution. Axial involvement is extremely unusual, with only a few dozen reported cases involving the spine or ribs. We present an 80-year-old asymptomatic male who underwent computed tomography (CT) of the chest for evaluation of unrelated respiratory symptoms. Incidentally noted was dense unilateral flowing cortical hyperostosis involving the right posterolateral aspect of the D3 vertebral body, extending into the posterior elements and adjacent third rib. Subsequent plain radiography confirmed the characteristic "dripping candle-wax" appearance, while magnetic resonance imaging demonstrated uniformly hypointense signal on all sequences without marrow edema or soft tissue component, consistent with melorheostosis. Given the absence of symptoms and lack of aggressive imaging features, conservative management with clinical observation was recommended. This case highlights the characteristic multimodality imaging features of axial melorheostosis, emphasizing the importance of recognizing this benign entity to prevent misdiagnosis as malignancy and avoid unnecessary invasive procedures. To our knowledge, combined vertebral and rib involvement detected incidentally in an asymptomatic elderly patient is rarely reported.

KEYWORDS: Melorheostosis; Sclerosing bone dysplasia; Axial skeleton; Candle-wax appearance; Incidental finding; Spine.

1. INTRODUCTION

Melorheostosis (Leri disease) is a rare benign sclerosing bone dysplasia that typically affects cortical bone in a segmental (sclerotomal) distribution. The classic imaging appearance resembling "dripping candle-wax" along the cortical margin was first described by Leri and Joanny in 1922 [1]. The prevalence is estimated at roughly 1 per 1,000,000 population [2]. Melorheostosis usually presents in childhood or early adulthood and most commonly involves the long bones of the limbs, particularly the lower extremities [3]. Symptoms often relate to pain, stiffness, deformity, or joint contractures in the affected limb.

In contrast, axial melorheostosis—affecting the spine, ribs, pelvis, or shoulder girdle is extremely unusual. Only a few dozen spinal or rib cases have been reported in the literature [4-7]. The imaging manifestations in the spine can range from isolated focal enostoses to bulky hyperostotic masses spanning several segments [8]. Because axial lesions are uncommon, they may be confused with other sclerosing disorders, including osteoblastic metastases, osteosarcoma, or Paget's disease. Recognition of the characteristic imaging features is therefore important to avoid unnecessary biopsy or intervention.

We describe a case of incidentally discovered melorheostosis involving the thoracic vertebra and adjacent rib in an asymptomatic elderly patient, and we review the pathophysiology, multimodality imaging features, and differential diagnosis of this rare axial presentation.

Case Presentation

An 80-year-old male underwent computed tomography (CT) of the chest for evaluation of respiratory symptoms unrelated to the musculoskeletal system. The patient denied back pain, radicular symptoms, limb weakness, prior trauma, constitutional symptoms, or history of malignancy. There was no known history of metabolic bone disease or inflammatory arthropathy. Physical examination demonstrated normal spinal alignment, preserved range of motion, and absence of focal tenderness or neurological deficit. Subsequent plain radiography of the thoracic spine and chest (Figure 1) confirmed dense

linear cortical hyperostosis with a characteristic "dripping candle-wax" appearance along the involved vertebra and adjacent rib. The lesion appeared confined to the cortex without aggressive features.

Incidental review of the CT images (Figure 2) revealed a focal area of dense cortical thickening involving the posterolateral aspect of the D3 vertebral body on the right side, extending into the posterior elements and adjacent third rib. The lesion demonstrated unilateral eccentric flowing cortical hyperostosis along the outer cortical margin, producing an undulating configuration. There was no evidence of cortical destruction, medullary lysis, periosteal reaction, vertebral body expansion, or associated paraspinal soft tissue mass. The intervertebral disc spaces were preserved, and there was no spinal canal compromise.

Magnetic resonance imaging (MRI) was performed for further characterization (Figure 3). The involved vertebral body and rib demonstrated uniformly low signal intensity on both T1-weighted and T2-weighted sequences, consistent with dense sclerotic bone [5,7]. There was no surrounding marrow edema, no abnormal enhancement, and no epidural or paraspinal soft tissue component. The spinal cord signal was normal.

Based on the characteristic multimodality imaging findings, particularly the unilateral flowing cortical hyperostosis without aggressive features, a confident radiologic diagnosis of axial melorheostosis was established [9]. Given the absence of symptoms and lack of complications, conservative management with clinical observation was recommended. At one-year follow-up, the patient remained symptom-free with no interval change.

DISCUSSION

Pathophysiology

Melorheostosis is a nonheritable mesenchymal dysplasia of unknown etiology. It is thought to arise from a somatic mosaic mutation during development, possibly involving the MAP2K1 signaling pathway [4]. Kang et al. [4] identified somatic activating mutations in MAP2K1 in affected tissues from patients with melorheostosis, providing insight into the genetic basis of this condition. Early hypotheses also implicated LEMD3 mutations, but these appear linked more with osteopoikilosis and Buschke-Ollendorff syndrome than sporadic melorheostosis [3].

The classic distribution of lesions often following a sclerotome (segmental innervation) pattern supports a developmental insult theory. Melorheostotic hyperostosis typically begins in childhood and progresses slowly, often stabilizing in adulthood. The process involves excessive intramembranous and endochondral ossification, leading to progressive thickening and hardening of cortical bone [2].

Imaging Characteristics

The hallmark of melorheostosis is cortical hyperostosis with a flowing, undulating margin, classically described as "dripping candle-wax" [1]. This is most apparent on plain radiographs (Figure 1), which typically show linear sclerotic streaks along one side of long bones or ribs. However, axial lesions may appear as focal dense enostoses or irregular thickening of vertebral or costal cortices [6,8].

CT confirms the morphology with superior bone detail: expansile cortical thickening that follows the surface of bone without medullary involvement (Figure 2). In our case, CT demonstrated high-attenuation undulating cortical hyperostosis of the vertebral body and adjacent rib, exactly resembling prior reports in axial cases [7,8]. The absence of cortical destruction, periosteal reaction, or soft tissue mass helps exclude malignant processes.

MRI reflects the low cellularity and dense mineralization of melorheostotic bone. Lesions are characteristically uniformly hypointense on all pulse sequences with no marrow edema or soft tissue component [5]. In our patient, all affected regions were very dark on T1 and T2 (Figure 3), with no gadolinium enhancement—findings consistently reported in the literature [5,7,9]. These MRI features are particularly valuable for excluding malignant tumors, which typically demonstrate intermediate signal, edema, or enhancement. Functional imaging (bone scan or PET) often shows moderate increased uptake due to active osteoblastic activity, though this was not performed in our asymptomatic patient.

Differential Diagnosis

The imaging findings of axial melorheostosis must be differentiated from other sclerosing bone lesions. Important considerations include osteoblastic osteosarcoma, parosteal osteosarcoma, osteoid osteoma, osteopetrosis, osteopathia striata, osteopoikilosis, chronic osteomyelitis, and sclerotic metastases (e.g., from prostate or breast primary) [2,10].

Melorheostosis differs from malignant lesions by its linear, flowing cortex-bound sclerosis and lack of aggressive features (no lysis, periosteal reaction, or soft tissue mass) [3,6]. Osteosarcoma typically demonstrates aggressive periosteal reaction, cortical destruction, and an associated soft tissue mass. Osteoid osteoma presents with a characteristic nidus and surrounding reactive sclerosis, often with nocturnal pain relieved by NSAIDs.

Myositis ossificans in the paraspinal region may be considered, but melorheostosis is confined to bone, whereas myositis ossificans arises in muscle and demonstrates characteristic zoning ossification. Osteopathia striata and osteopoikilosis produce multiple small sclerotic foci (striations or spots), not the bulky hyperostosis seen here [10]. Paget's disease typically

involves bone expansion, cortical thickening, and a characteristic "cotton wool" appearance on radiography, often with elevated alkaline phosphatase.
In our patient, the well-circumscribed, non-destructive nature of the lesions, their classic wax-like morphology, and the absence of any clinical symptoms were the keys to establishing the correct diagnosis [9].

Review of Axial Cases
Axial melorheostosis remains exceptionally rare. Motimaya et al. [8] reported cervical and upper thoracic spine involvement with characteristic imaging findings similar to our case. Bayya et al. [6] described a case of axial melorheostosis involving multiple vertebral levels. Leong et al. [7] presented a case of spinal melorheostosis with correlative imaging findings, emphasizing the uniformly low signal on MRI. Hoang et al. [5] illustrated the classic "dripping candle wax" sign in appendicular melorheostosis, which serves as the diagnostic hallmark.
Combined vertebral and rib involvement, as seen in our patient, is particularly unusual. Elsheikh et al. [11] described two radiologically distinct lesions in the shoulder region, highlighting the variable presentation of this dysplasia. Iordache et al. [12] recently reviewed the literature and emphasized the importance of imaging in diagnosis. To our knowledge, incidental detection in an asymptomatic elderly patient with combined vertebral and rib involvement has rarely been reported.

Management
There is no cure for melorheostosis; treatment is symptomatic. Most patients (especially those with incidental or asymptomatic findings) require no intervention [3]. Pain, if present, is managed with analgesics and physical therapy. Some reports have suggested benefit from bisphosphonates to reduce osteoblastic activity, although evidence remains anecdotal [2]. Surgical options (e.g., excision, osteotomy, spinal decompression) are reserved for severe deformity or nerve compression. Saxena et al. [9] recommended nerve root decompression for lumbar radiculopathy caused by melorheostosis compressing neural elements.
In our patient, no symptoms were attributable to the lesions, and there were no imaging features suggesting impending complications. Therefore, we elected clinical and imaging surveillance. At one-year follow-up, the patient remained symptom-free with no change in the appearance of the lesions. This conservative approach aligns with other reports of incidental melorheostosis [6,7].

CONCLUSION
Melorheostosis is a rare benign sclerosing bone dysplasia usually confined to the appendicular skeleton. Axial involvement is exceptionally rare, particularly in the spine and ribs. We report an incidental case of asymptomatic melorheostosis involving the thoracic vertebra and adjacent rib in an 80-year-old male, identified on CT performed for unrelated respiratory symptoms. The diagnosis was established by recognition of the characteristic "dripping candle-wax" cortical hyperostosis on plain radiography and CT, with confirmatory uniformly hypointense signal on MRI and absence of aggressive features. This case highlights the importance of multimodality imaging in diagnosing unusual presentations of melorheostosis. Awareness of its benign imaging hallmarks, cortex-bound linear sclerosis, lack of destruction or soft tissue mass, and characteristic low signal on all MRI sequences can prevent misdiagnosis as malignancy and avoid unnecessary biopsy or treatment. Conservative management with clinical observation is appropriate for asymptomatic patients.
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Author Contributions
L.N.M.S. contributed to data collection, image interpretation, and drafting of the manuscript. G.M. was involved in study supervision, critical review, and final approval of the manuscript. A.K.A. conceptualized the study, contributed to manuscript drafting, and served as the corresponding author. A.B. participated in image analysis and manuscript revision. V.V. contributed to literature review and manuscript editing. L.N.B.S. assisted in data interpretation and preparation of figures. All authors reviewed and approved the final version of the manuscript.

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Figure Legends

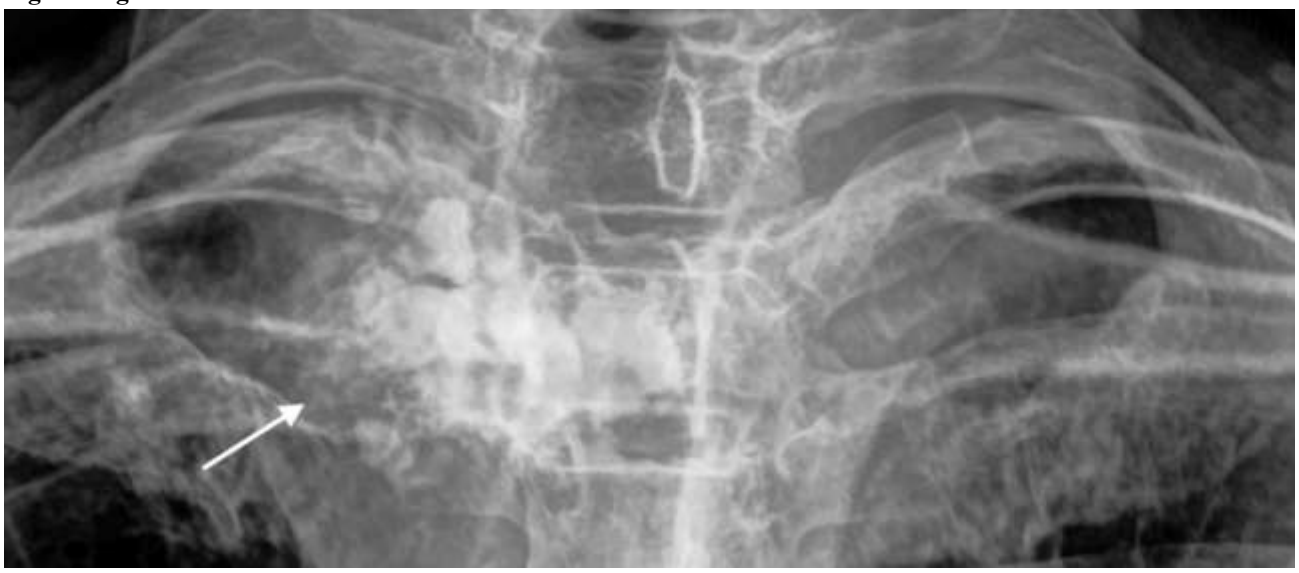

Figure 1. Plain radiograph of the thoracic region demonstrating unilateral flowing cortical hyperostosis involving the right posterolateral aspect of the upper thoracic vertebra and adjacent rib (arrow), producing the characteristic "dripping candle-wax" appearance. No cortical destruction, bone expansion, or aggressive periosteal reaction is identified.

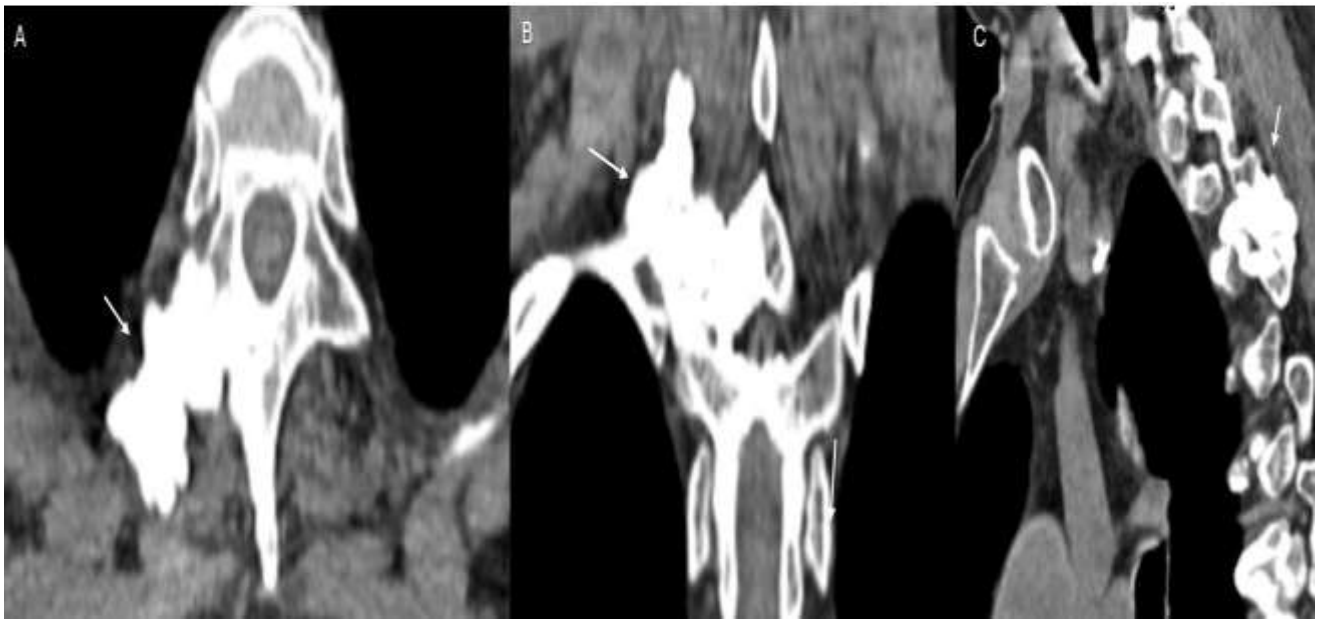

Figure 2. Computed tomography (bone window) images of the thoracic spine and adjacent rib demonstrating eccentric unilateral cortical hyperostosis. (A) Axial image shows dense flowing cortical thickening along the posterolateral aspect of the D3 vertebral body extending into the posterior elements (arrow). (B) Coronal reconstruction demonstrates linear undulating cortical hyperostosis along the vertebral body and adjacent rib (arrow). (C) Sagittal reconstruction confirms flowing cortical thickening without cortical breach, medullary destruction, or associated soft tissue mass (arrow).

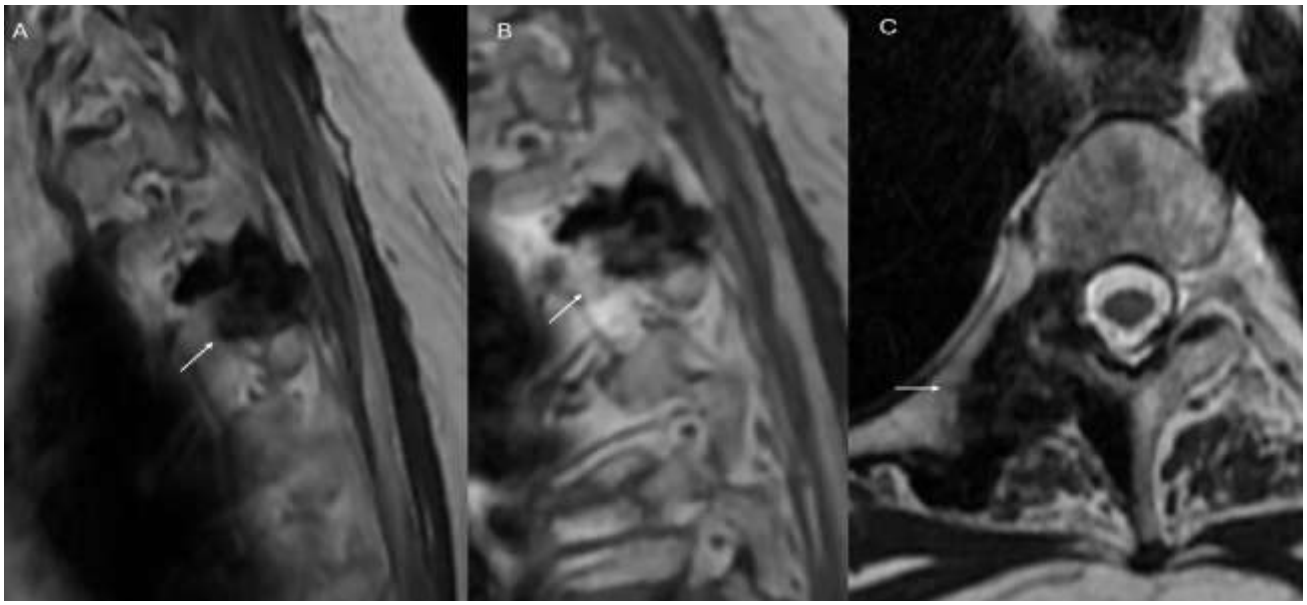

Figure 3. Magnetic resonance imaging of the thoracic spine demonstrating imaging features consistent with melorheostosis. (A) Sagittal T2-weighted image shows focal low signal intensity cortical thickening involving the posterolateral aspect of the D3 vertebral body (arrow). (B) Sagittal T1-weighted image demonstrates corresponding uniformly hypointense cortical hyperostosis without marrow replacement or expansion (arrow). (C) Axial T2-weighted image confirms eccentric cortical-based low signal thickening extending into the posterior elements and adjacent rib (arrow), with no surrounding marrow edema, epidural extension, or paraspinal soft tissue component.

Patient Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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