

Symmetric Presentation of Melorheostosis Involving Multiple Flat Bones in the Thorax: A Case Report¹

Jung Suk Oh, M.D., Hyun Jin Park, M.D., Jee Young Kim, M.D.,
Ki Jun Kim, M.D.², Won Sang Jung, M.D.

Melorheostosis is a rare bone dysplasia of unknown etiology that usually affects a single limb and is characterized by cortical thickening, with a flowing candle wax appearance, extending vertically along the surface of the long bone. We report a case of polyostotic melorheostosis symmetrically involving multiple flat bones, ribs, and the scapulas. This is the first case of melorheostosis symmetrically involving multiple flat bones.

Index words : Melorheostosis

Thorax

Tomography, X-Ray Computed

Melorheostosis is a rare nonhereditary sclerosing mesenchymal dysplasia of bone that is diagnosed by a characteristic linear cortical hyperostosis with a flowing candle wax appearance (1-3). It commonly affects long bones, usually in the lower limb and on one side of the body. The involvement of flat bones is rare (1, 2). In addition, bilateral involvement is extremely rare, with only one case of long tubular bones with a symmetric distribution reported to date. We present the first case of melorheostosis with multiple flat bone involvement and symmetric distribution of the lesions.

Case Report

A 55-year-old man was diagnosed with sclerotic ribs

and scapulas on a chest radiograph (Fig. 1A) as part of a routine check-up. Neither the patient nor his family members had a history of significant disease. The patient's blood chemistry and physical examination revealed no significant abnormalities. A chest CT showed uneven cortical thickening in multiple ribs, both scapulas, and thoracic vertebral bodies. The cortical thickening of the ribs and scapulas was bilateral and symmetrical in distribution (Fig. 1B). Our differential diagnosis included the polyostotic form of melorheostosis, bilateral osteoma, and osteoblastic metastasis. For further evaluation, using the acquired volume data, we obtained multiplanar reconstruction (MPR) images, which revealed that cortical thickening was seen along the lower aspects of the ribs, infraspinatus fossas, the spines of both scapulas, and the anterior column of the thoracic vertebral bodies (Figs. 1C, D).

Discussion

Melorheostosis is a rare, osteosclerotic dysplasia with an unknown etiology and first described in 1922 by Leri and Joanny (1, 2). The diagnosis is based on the clinical

¹Department of Diagnostic Radiology, St. Vincent's Hospital, The Catholic University of Korea

²Department of Diagnostic Radiology, Incheon St. Mary's Hospital, The Catholic University of Korea

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Address reprint requests to : Won Sang Jung, M.D., Department of Diagnostic Radiology, St. Vincent's Hospital, The Catholic University of Korea, 93-6 Ji-dong Paldal-gu, Suwon-si, Gyeonggi-do 442-723, Korea.

Tel. 82-31-249-7492 Fax. 82-31-247-5713

E-mail: wonrad@gmail.com

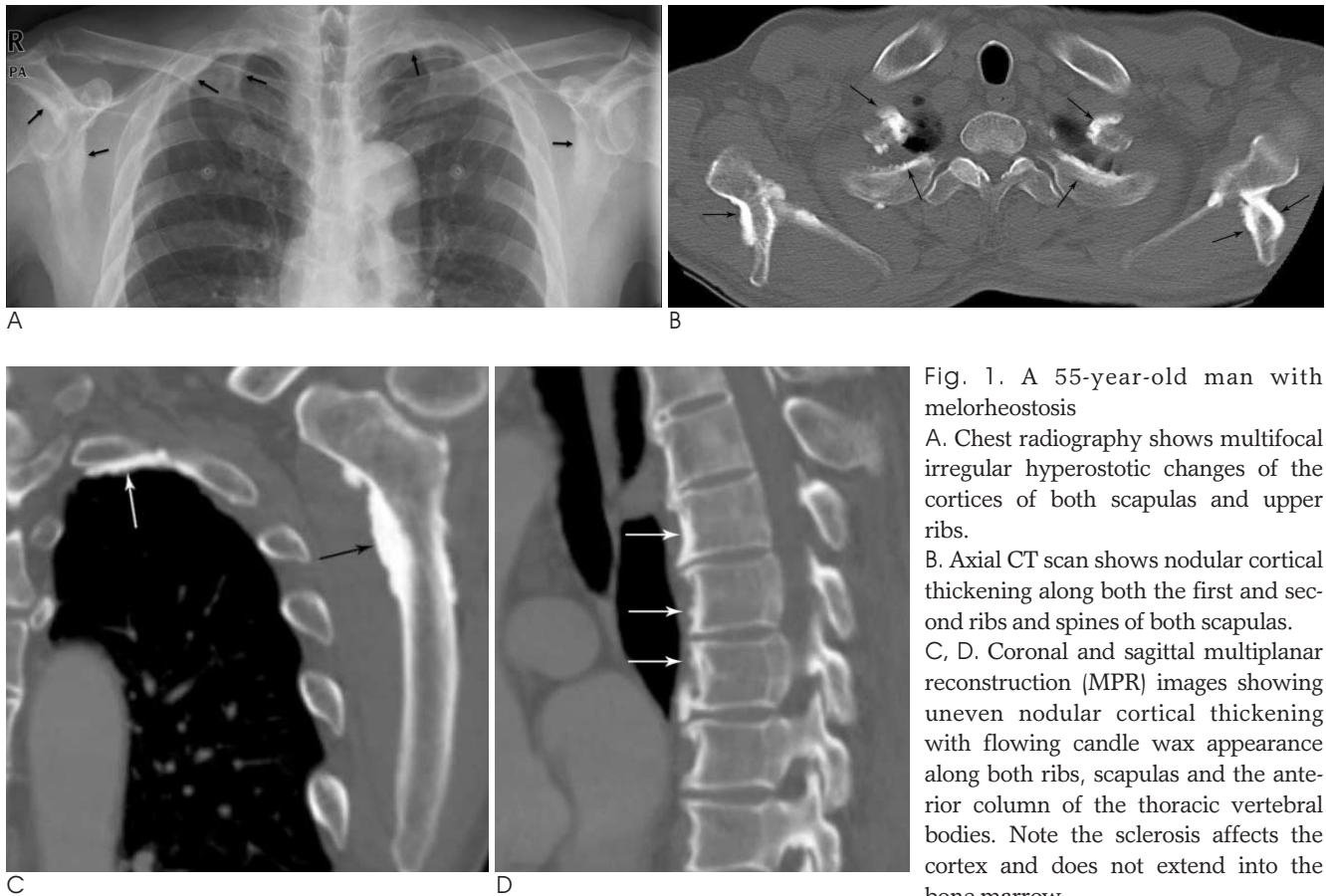


Fig. 1. A 55-year-old man with melorheostosis

A. Chest radiography shows multifocal irregular hyperostotic changes of the cortices of both scapulas and upper ribs.

B. Axial CT scan shows nodular cortical thickening along both the first and second ribs and spines of both scapulas.

C, D. Coronal and sagittal multiplanar reconstruction (MPR) images showing uneven nodular cortical thickening with flowing candle wax appearance along both ribs, scapulas and the anterior column of the thoracic vertebral bodies. Note the sclerosis affects the cortex and does not extend into the bone marrow.

and radiological findings. The radiological appearance of melorheostosis is a characteristic wavy cortical hyperostosis along the surfaces of the long bone with vertical extension, and resembles melted wax dripping down one side of a candle; the so called flowing candle wax appearance (2, 4). Melorheostosis frequently involves the long tubular bones. By far, the most common sites affected include the long bones of the lower limbs (4). This disorder tends to be segmental and unilateral, and based on the extension of the bone involvement of the lesion, it may be monostotic, monomelic, or polyostotic (4). A bone scintigraphy is invariably positive in patients with melorheostosis, revealing a moderate increase in uptake of tracer, predominantly localized to the cortex (3, 4). Computed tomography (CT) scanning more effectively reveals a clear demarcation from normal bone than standard radiographs (4). Abnormalities of the soft tissues overlying the bone and joint lesions are common, and in many cases, associated scleroderma have been reported (3, 4).

Most of the reported cases showed distribution of one long bone or one limb with a scleroderma distribution

(5). This characteristic distribution pattern and radiological features differentiates melorheostosis from other osteosclerotic dysplasias. Isolated flat bone involvement is uncommon (3), and multiple flat bone involvement without long bone involvement, as seen in our patient, is exceedingly rare. Moreover, symmetric involvement of multiple flat bones has not been previously reported.

Melorheostosis is a rare dysplasia with characteristic cortical thickening, commonly involving the long bones of one limb. Here we report the first case of melorheostosis with the characteristic CT findings and an unusual distribution pattern with multiple flat bones that were symmetrically involved without long bone involvement.

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흉곽에서 대칭적으로 편평골을 침범한 다발성 유선상과골증: 1예 보고¹

¹가톨릭의대 성빈센트병원 영상의학과

²가톨릭의대 인천성모병원 영상의학과

오정석 · 박현진 · 김지영 · 김기준² · 정원상

유선상과골증은 원인 미상의 드문 골 이형성증이다. 대개 단일 사지를 침범하며, 장골의 표면을 따라 수직적으로 흐르는 촛농 모양의 피질비대가 특징적이다. 저자들은 다골성 유선상과골증이 늑골과 견갑골과 같은 편평골을 대칭적이고 다발성으로 침범한 예를 보고하고자 한다.