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¹⁸F-NaF PET/CT in extensive melorheostosis of the axial and appendicular skeleton with soft-tissue involvement

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Abstract

Melorheostosis is a rare, non-hereditary, benign, sclerotic bone dysplasia with no sex predilection, typically occurring in late childhood or early adulthood which can lead to substantial functional morbidity depending on the sites of involvement. We report on a patient with extensive melorheostosis in the axial and appendicular skeleton as well as in the soft-tissues, who was evaluated with whole-body ¹⁸F-NaF PET/CT scan. All melorheostotic lesions of the skeleton and of the ossified soft-tissue masses, demonstrated intensely increased ¹⁸F-NaF activity suggesting the application of this modality in assessing and monitoring the disease activity.

Melorheostosis is a rare, non-heridatary, benign, sclerotic bone-dysplasia of unknown etiology which typically occurs in late childhood or early adulthood [1, 2]. The characteristic radiographic appearance of melorheostosis consists of cortical and medullary hyperostosis of the affected bones, giving the impression of "dripping candle wax" [3, 4]. Melorheostosic lesions in the axial skeleton and soft-tissue ossified masses like in the case presented herein, are rare features of this uncommon disorder [4–6]. The presented data indicates ¹⁸F-NaF-avidity of all melorheostotic lesions both in the skeleton and in sites of extra-osseous

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ossifications, making ¹⁸F-NaF the optimal bone-seeking PET-radiopharmaceutical [7–9] to assess and monitor the disease activity.

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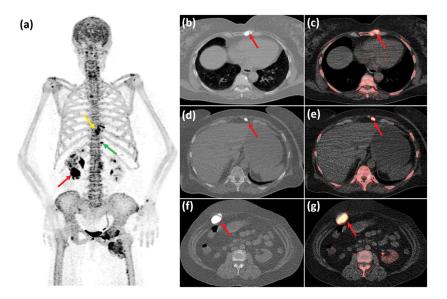


Figure 1.

A 67-year-old woman with established diagnosis of melorheostosis since the age of 22 was referred to our institute. Although independent in self-care activities, the patient has been suffering from chronic pain secondary to the melorheostosis with three to five episodes of pain flare-ups per year. The patient developed lymphedema of the left lower extremity and leg length discrepancy with the left leg being longer. In order to assess the disease activity the patient underwent whole body ¹⁸F-NaF PET/CT scan, which showed multiple ¹⁸F-NaF avid lesions corresponding to the hyperdense bone abnormalities and the soft-tissue ossifications. A prominent lesion with intensely elevated ¹⁸F-NaF activity (SUVmax: 45,4) resided in the left lower sternum (Fig. 1a: Maximum Intensity Projection (MIP) PET image of the torso, yellow arrow; Fig 1b & 1c: axial CT and axial fused PET/CT images of the chest, red arrows). Also, two sites of extra-osseous bone formation were observed in the anterior abdominal wall (Fig. 1a: green arrow; Fig 1d & 1e: axial CT and axial fused PET/CT images of the upper abdomen, red arrows) and in the right anterolateral abdominal wall (Fig. 1a: red arrow; Fig 1f & 1g: axial CT and axial fused PET/CT images of the abdomen, red arrows) which also showed markedly elevated ¹⁸F-NaFuptake (SUVmax: 30,3 & 42,6 respectively).

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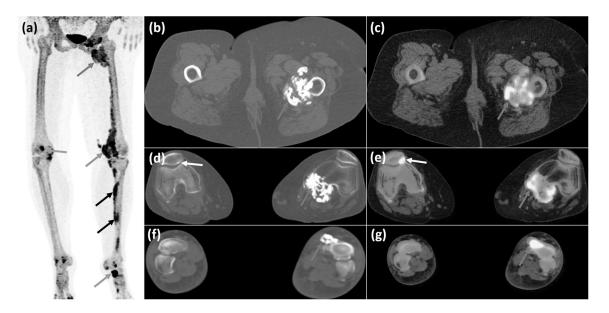


Figure 2.

In the lower extremities, abnormal left femoral osteosclerosis with associated bulky softtissue ossifications was seen between the left proximal femur and the pelvis (Fig 2a: Maximum Intensity Projection (MIP) PET image of the lower extremities, red arrow; Fig 2b & 2c: axial CT and fused PET/CT images of the femurs, red arrows), with intensely increased ¹⁸F-NaF activity (SUVmax: 52,2). Linear elevated ¹⁸F-NaF activity was seen along the left femoral shaft as well as in an extra-osseous ossified mass adjacent to the medial femoral condyle (Fig 2a: green arrow; Fig 2d & 2e: axial CT and fused PET/CT images of the knees, red arrows,) with high uptake as well (SUVmax: 51,7). Focal intense uptake (SUVmax: 55,3) in the right patella corresponded to an osteosclerotic lesion (Fig 2a: blue arrow; Fig 2d & 2e: white arrows). Sites of intense ¹⁸F-NaF avidity (SUVmax: 52,7) were seen in the medial left tibia (Fig 2a: black arrows) and in the left talor-navicular joint (SUVmax: 56,2) corresponding to melorheostotic lesion observed on CT (Fig 2a: orange

arrow; Fig 2f & 2g: axial CT and fused PET/CT images of the feet, red arrows).