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Neurogenic Myositis Ossificans of the Hip in a Bedridden Patient with Neurofibromatosis

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Abstract **Case Report**

Neurogenic myositis ossificans is an uncommon condition characterized by the formation of heterotopic bone in the periarticular soft tissues of individuals who have paraplegia or tetraplegia due to neurological disorders, frequently occurring after head or spinal cord injuries. Diagnosis relies on imaging, particularly CT scans, which commonly show the appearance of bony formations within the muscles around joints. Management involves the use of pain relievers, physical therapy, and surgery as a final option.

Keywords: Neurogenic Myositis ossificans, bedridden patient, Benign tumor, Imaging.

INTRODUCTION

Neurogenic myositis ossificans is an uncommon and benign pseudotumor condition characterized by the development of abnormal bone growth within muscles. This occurs in individuals with severe neurological disorders. The clinical symptoms are vague and typically involve the presence of a painful mass. This underscores the significance of imaging, particularly CT scans, which play a pivotal role in confirming the diagnosis. CT scans allow for a thorough examination of the affected muscles, enabling a comprehensive assessment to guide potential surgical interventions and rule out other possible conditions. We present an exceptional case of neurogenic myositis ossificans of the left hip observed in a patient with neurofibromatosis who was being bedridden for amputation of the right limb following a malignant transformation of neurofibroma.

OBSERVATION

A 26-year-old women patient, followed for neurofibromatosis type 1 who underwent amputation of the right leg due to degeneration of a neurofibroma, with a history of being bedridden. She presented with a one-year history of chronic left inguinal pain associated with swelling and limitation of motion. On physical examination, the letf hip joint was absolutely stiff with flexion deformity and fixed external rotation, and power was 4/5 all over the left lower extremity with intact sensation. Pelvic CT scan showed multiple intramuscular peri-coxofemoral calcifications, with more extensive region on the right, the coxofemoral, sacroiliac joints, flaky and confluent, showing a pseudotumor appearance; however, a periosteal reaction or associated bone lesion was absent (Fig 1).

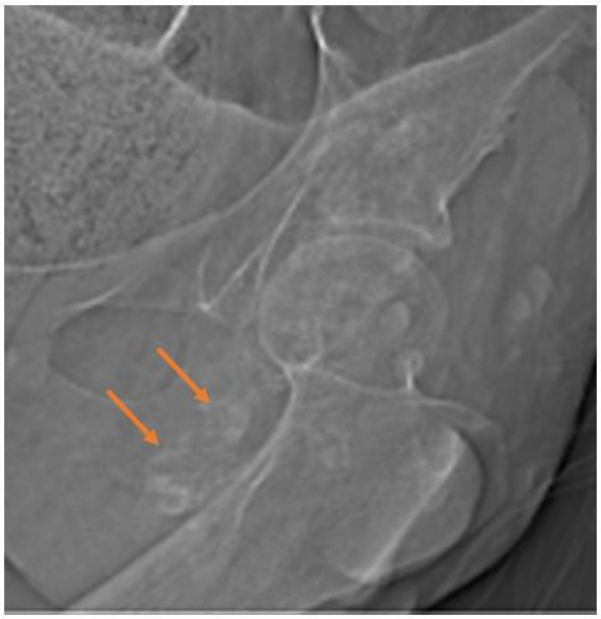

Figure 1: Conventional radiograph of the left hip

Figure 1 Conventional radiograph of hip showing circumferential calcification with a lucent center and a radiolucent cleft that separates the lesion from the cortex of the femur (Orange arrow).

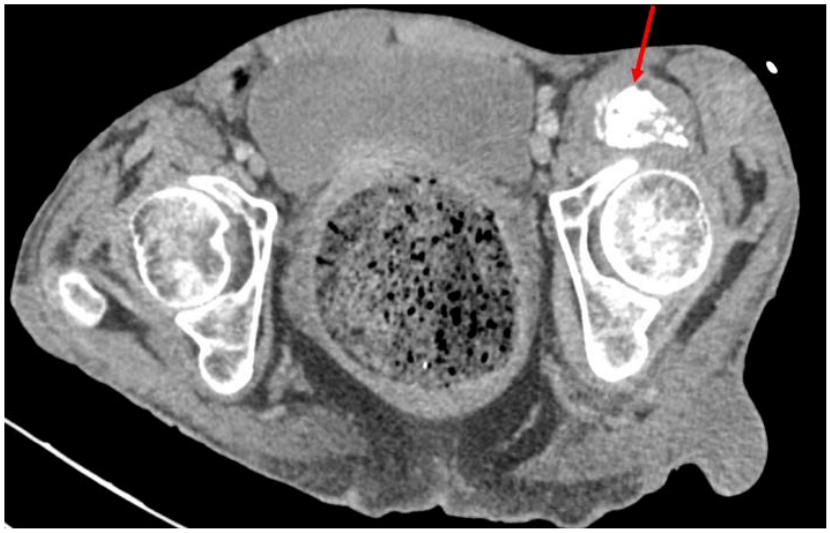

Figure 2: AxialCT enhanced scan, Soft tissue window

Figure 2 axial enhanced pelvic CT scan, soft tissue window show a fairly well-defined and lobulated mass in the anterior compartment of the hip abutting the adjacent muscles containing multiple calcifications, non-enhancing with contrast (Red arrow).

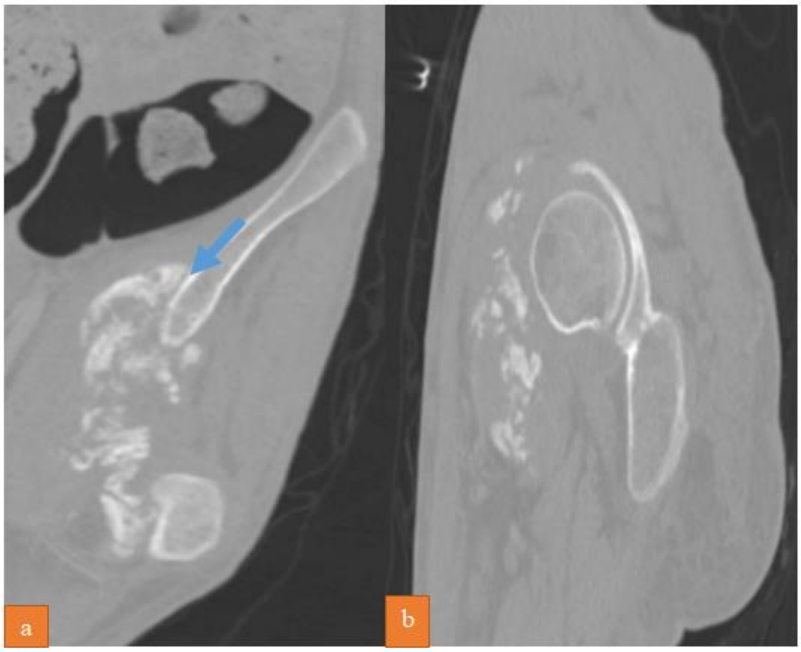

Figure 3a: Pelvic CT scan, bone Window, a, Coronal view, b. Sagittal view

Figure 2 a, b: Coronal (a), Sagittal (b) enhanced pelvic CT scan, Bone window, showing irregular ossifications within the soft tissues anterior to the joint with a radiolucent cleft separates it from the cortex (Blue arrow)

DISCUSSION

Heterotopic ossification (HO) can manifest as a result of various triggers, including local bone or soft tissue injuries, total joint replacement surgeries, burns, and the development of spastic or paralyzed joints following central nervous system injuries or disorders. Neurogenic heterotopic ossification typically presents in patients who have experienced traumatic brain or spinal cord injuries [1].

It is interesting to note that this patient did not have any evidence of brain or spinal cord injury. However, she was in an iatrogenically induced state of

neuromuscular paralysis as part of her treatment after the amputation.

In a clinical context, individuals with heterotopic ossification (HO) may exhibit a wide range of symptoms, spanning from pain and restricted range of motion (ROM) to the complete fusion of the affected joint, known as ankylosis. Interestingly, these symptoms may not always align with what is observed on radiological scans. Consequently, diagnosing HO based solely on clinical presentation can be a complex task. Moreover, HO can resemble other conditions at different stages of its development.

At the outset of evaluating patients presenting clinical symptoms indicative of heterotopic ossification (HO), conventional radiography is often the go-to imaging method. Initially, there is an absence of calcification, but soft tissue swelling may be observed. Typically, calcification starts becoming evident within a span of 2 to 6 weeks, and the lesion gradually develops the classic, well-defined peripherally calcified appearance by the end of 2 months. Over the subsequent 4 months or so, these lesions usually tend to decrease in size and increase in density [2].

CT scans often mirror the appearance of lesions observed on conventional radiographs, showing a pattern of bone formation that initiates at the periphery and progresses toward the center. As the lesion matures, a distinctive zonal pattern of mineralization emerges, marked by the presence of a fully formed bony cortex. The presence of this classic zonal appearance on CT scans is highly specific for heterotopic ossification (HO). Cross-sectional imaging is valuable for pinpointing the exact location of the lesion. However, if a CT scan is conducted during the early stages of maturation, it may reveal a soft tissue mass with irregular or absent mineralization, potentially resembling a soft tissue sarcoma or, particularly in the case of the juxtacortical subtype, osteosarcoma. In such cases, performing a follow-up CT scan after several weeks is beneficial, as it provides an opportunity for better characterization as the lesion continues to mature and evolve [3].

Magnetic resonance (MR) imaging findings can vary based on the age of the heterotopic ossification (HO) lesion. In its early stages, the features are non-specific, often displaying a heterogeneous high T2-weighted signal within the lesion, with an occasionally indistinct hypointense rim indicating calcification. When gadolinium contrast is administered intravenously, it can result in early and intense, yet heterogeneous, enhancement of the lesion, potentially leading to confusion with osteomyelitis. As the lesion continues to mature over several weeks to months, MR imaging may reveal a more clearly defined hypointense rim, which corresponds to the mature cortical bone, along with the development of high T1- and T2-weighted signals in the central region. In late-stage lesions, enhancement is typically absent, although a few may exhibit minimal enhancement [4].

The cause of HO in our patient is likely to be microtrauma as a result of repeated cycles of prolonged immobilisation, followed by forcible mobilisation of the joints. Management of HO is often targeted at preventing the condition from occurring in unaffected patients. Single-dose radiotherapy or oral nonsteroidal anti-inflammatory drugs have been described to be helpful in the prevention of HO.[1] Medical therapy and radiotherapy are available for the treatment and prophylaxis of HO, but excisional surgery remains the treatment of choice to restore joint mobility. [1]. Due to the extensive mature HO, our patient was managed with only oral analgesics. Surgical management was not indicated for her, as it would not have improved her mobility due to her prior condition.

CONCLUSION

Neurogenic ossificans HO is commonly seen after a spinal cord or traumatic brain injury, but less commonly in patients with neurofibromatosis. Due to the nonspecific nature of the radiological findings of early HO, this clinical condition is easily overlooked and may hence be underdiagnosed. The present case is rare, as the patient's HO occurred in her non-paretic limb as she undergone an amputation of her right limb. Our patient had bony ankylosis of her left hip joint with radiological findings of extensive mature ossification of the periarticular soft tissue. The corresponding CT images, along with plain radiographs, showed extensive mature HO.

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