Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Rheumatology

Neurogenic Heterotopic Ossification with Bilateral Intercostal Ossification: A Case Report

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DOI: https://doi.org/10.36347/sjmcr.2025.v13i01.041 | **Received:** 19.12.2024 | **Accepted:** 24.01.2025 | **Published:** 25.01.2025

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Abstract **Case Report**

Introduction: Neurogenic heterotopic ossification (NHO) is one of the most common forms of heterotopic ossifications, mainly affecting the large proximal joints of the limbs, it can involve one or more joints, the intercostal localization of heterotopic ossifications is rarely described in the literature. ***Case presentation*:** a 41-year-old female patient presented with mechanical arthralgia of the knees, left elbow and shoulders a disabling stiffness, following prolonged bed rest after spinal and pelvic impact trauma. On osteoarticular examination, the affected joints showed limited range of motion. Standard X-rays of the affected joints, a thoracic computed tomography and a bone scintigraphy confirmed the diagnosis of NHO with bilateral heterotopic ossification between the first two ribs. The patient was put on medical treatment with non-steroidal anti-inflammatory drugs while awaiting stabilization of the scintigraphic lesions for possible surgical management. ***Conclusion*:** In conclusion, we report a case of NHO with multiple localizations in a young patient, which is responsible major functional handicaps, implying the need for prevention of this very disabling condition, whose management is very delicate.

Keywords: Neurogenic heterotopic ossification, arthralgia, stiffness.

INTRODUCTION

Heterotopic ossification is the ectopic formation of histologically normal bone in an extraskeletal site, and can involve various types of soft tissue (skin, muscle, tendons, ligaments …) [1]. Neurogenic Heterotopic ossification (NHO) is one of the most common forms of heterotopic ossification, occurring following central neurological damage secondary to spinal cord injury, cerebral anoxia, traumatic brain injury or stroke [2]. Clinically, NHO manifests itself as a joint stiffness, with or without inflammatory signs, and may progress to ankylosis of the affected joint, with muscular atrophy and nerve damage due to nerve entrapment, impairing the patient's quality of life and usually requiring surgical management. NHO mainly affects the large proximal joints of the limbs [3], and preferentially the hips, elbows, knees and shoulders [2], just as it can affect several joints in the same subject, with or without bilateral topography [4, 5]. The intercostal location of heterotopic ossifications is rarely described in the literature [6]. We report a case of NHO with multiple locations and intercostal involvement in a 41-year-old woman.

CASE PRESENTATION

A 41-year-old woman, having a history of spinal and pelvic impact trauma following a 2-stage fall. The patient was subsequently hospitalized in a traumatology department for staged vertebral fractures with a pelvic fracture. During her hospitalization, the patient experienced respiratory distress with consciousness disorders, for she was transferred to an intensive care unit where she remained hospitalized for 1 month, followed by prolonged bed rest for around a year at home.

Two years later, the patient presented to our clinic with mechanical polyarthralgia of the knees, left elbow and shoulders, with a stiffness. On osteoarticular examination, the patient walked with a walker, and presented with 45° flessum of the elbows with a pain on active and passive mobilization, stiffness of both shoulders with 90° abduction, and limited flexion of both knees (heel- to-buttock distance 35 cm on the right, 30 cm on the left). Neurological examination was normal.

A standard x-rays was requested, it showed ectopic ossifications juxta-articulatory of shoulders,

knees and left elbow (Figures 1, 2 and 3). Biological tests were normal (Table 1).

Table I: Biological tests performed on the patient

Biological parameter	Patient value	Reference values
Hemoglobin (g/dl)	13	12-16
White blood cell count(/uL)	8900	4000-10000
Neutrophils (/uL)	3300	1000-4000
Platelets (/uL)	228000	150000-400000
Sedimentation rate (MM)	30	0-10
C- reactive protein (mg/L)	6.6	0-3
Serum alkaline phosphatase (U/L)	96	40-129

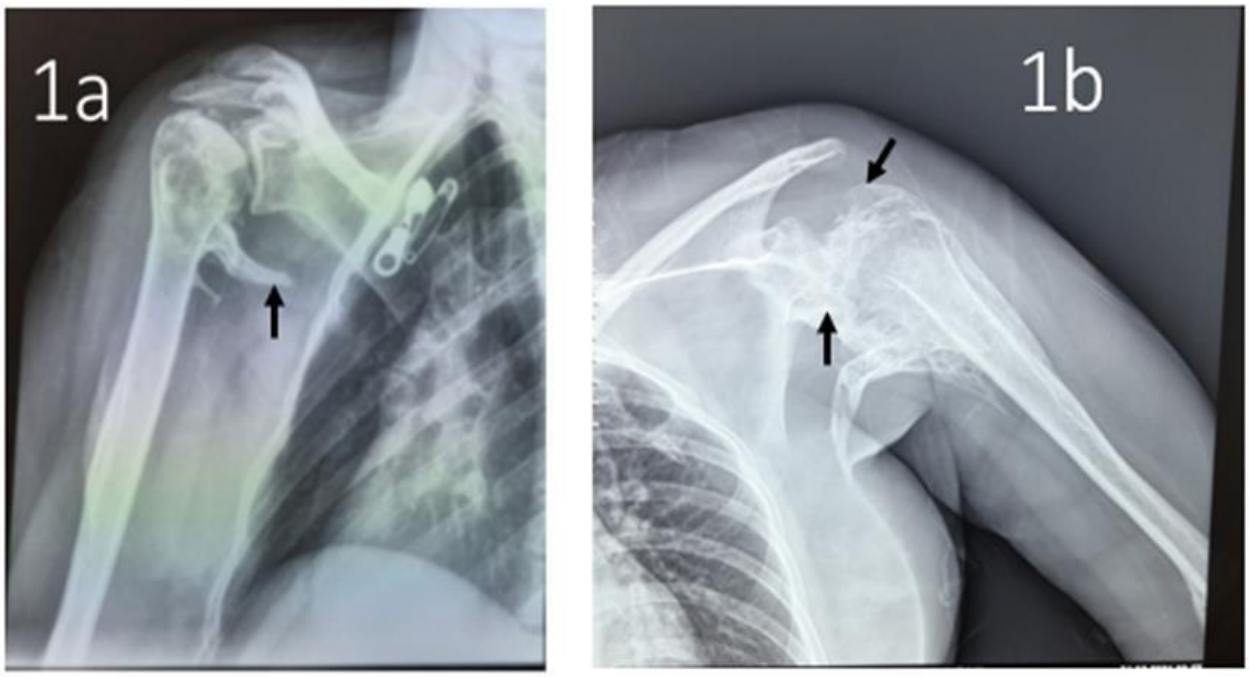

Figure 1: a) Standard front X-ray of the right shoulder showing heterotopic ossification at the humeral neck; b) Standard front X-ray of the left shoulder showing heterotopic ossification encompassing the entire glenohumeral joint

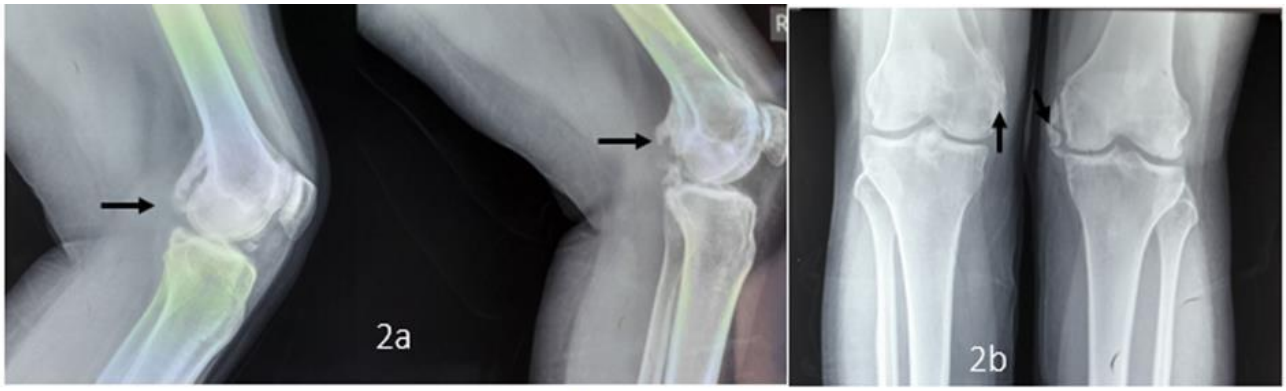

Figure 2: a) Standard profile radiograph of both knees showing retroarticular ossification; b) standard frontal radiograph of both knees showing medial heterotopic ossification of the femorotibial joint

Figure 3: a- Standard profil X-ray of the left elbow showing heterotopic ossification encompassing the entire humero- ulnar and superior ulno-radial joint

A thoracic computed tomography (CT) revealed severe calcifying and ossifying damage to the tissues and periarticular areas involving the bilateral glenohumeral and costal joints (Fig 4).

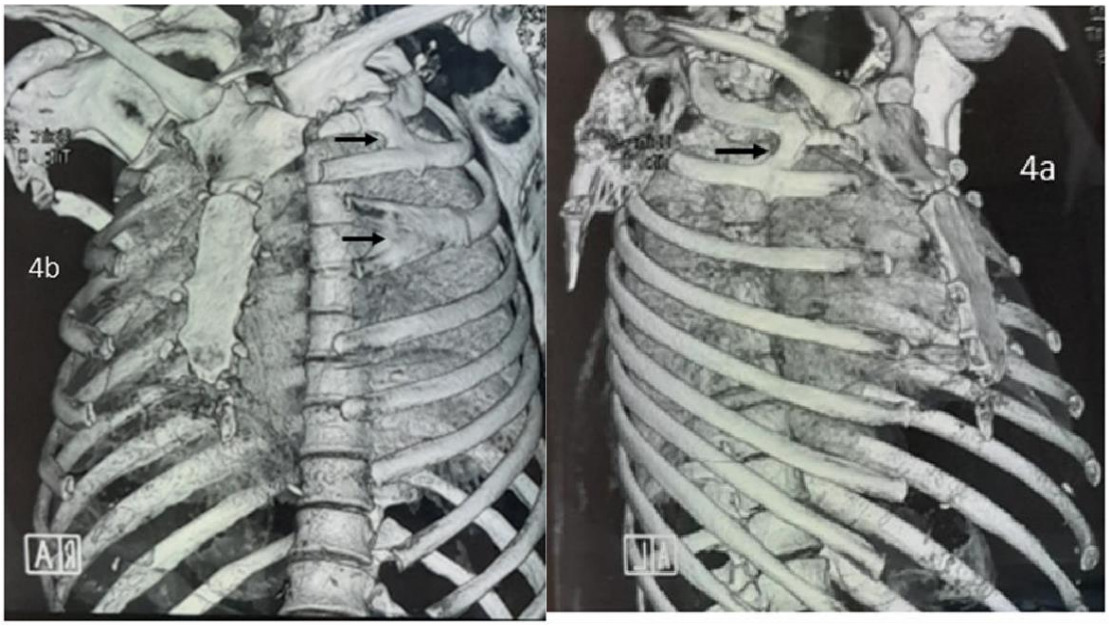

Figure 4: Thoracic CT scan showing intercostal bony bridges connecting the anterior of the first two ribs bilateral with bifurcation of the anterior arch of the third right rib

A bone scan revealed areas of weak to moderate hyperfixation of the para-articular ossifications of the shoulders and left elbow in the late period, with moderate hyperfixation of the osteoma of the first right chondro-sternal junction. Another bone scan was carried out a year later, which showed moderate to very intense posterior para-articular ossifications of both knees in late time, reflecting the progressive nature of the disease, with foci of hyperfixation of the right 1st rib and the left 3rd and 4th ribs in relation to probable fractures (Figure 5). The patient was put on medical treatment with non-steroidal anti-inflammatory drugs, while waiting for a stabilization of the scintigraphic lesions for possible surgical management.

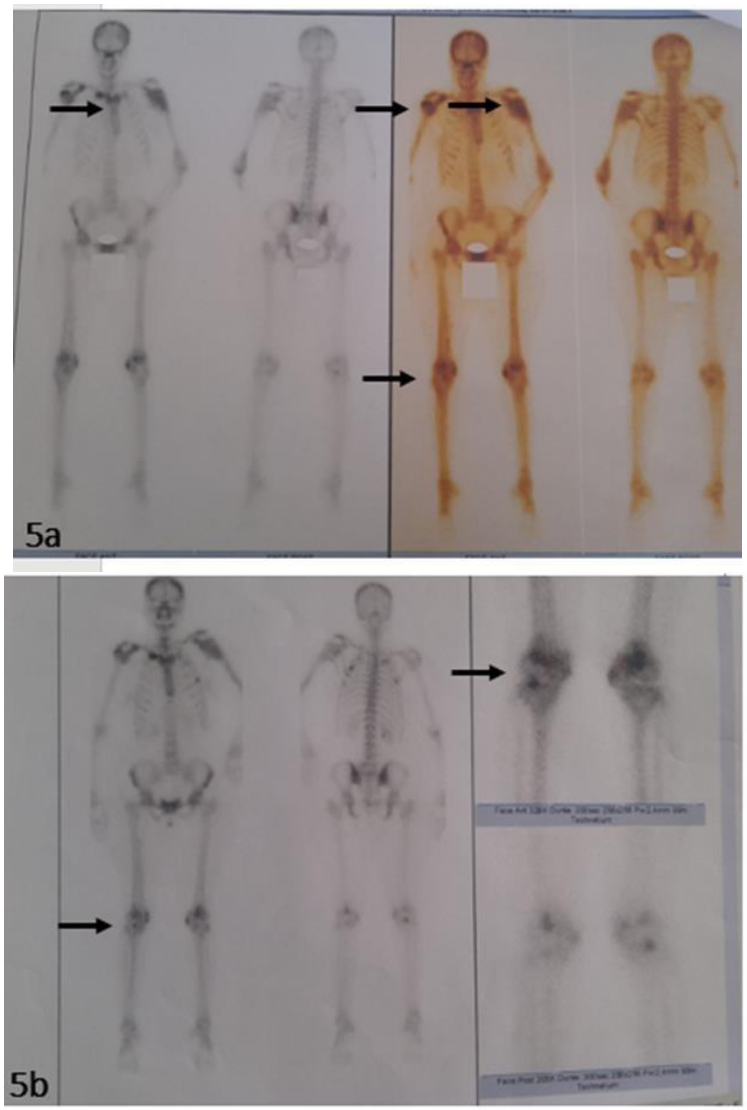

Figure 5: Bone scan after HMDP-Tc 99m injection performed at one-year intervals: 5a) Foci of weak to moderate hyperfixation of para-articular ossifications of the shoulders and left elbow at time late with moderate hyperfixation of the osteoma of the first right chondrosternal junction; 5b) Scintigraphic appearance in favour of several posterior para-articular ossifications of both knees with moderate to very intense fixation in late time, testifying to their progressive nature

DISCUSSION

NHO is a heterotopic ossification that can occur after musculoskeletal trauma, joint surgery or injury to the central or peripheral nervous system. It affects 44% of patients who have undergone a hip surgery, and 10-20% of patients with nervous system lesions [7]. In our case, NHO of multiple localizations occurred in a patient who had suffered of a traumatic injury and has been confined to bed for a long time. The pathophysiological mechanism of NHO is not well elucidated, however, two theories have been put forward to explain this pathology: the first being prolonged immobilization associated with forced manipulation of the joints by nursing staff to avoid joint stiffness, and the second being the damage to the central nervous system leads to pathological activation of certain factors that allow stimulation of mesenchymal cells and their transformation into osteoblasts responsible for this ectopic ossification [1, 8, 9]. The probable risk factors for NHO include young age, male gender, prolonged coma, delayed rehabilitation, pressure sores, bone fractures and urinary tract infections [10-12]. NHO mainly affects the large proximal joints of the limbs, especially the hip joint, which is the most severely affected joint. It affects a single joint in 40% of patients, and two joints in a third of cases [13], and manifests itself as arthralgia with or without joint stiffness, depending on the stage of NHO; these symptoms usually occur within 3 to 12 weeks of the trigger [7]. Our patient had multiple localizations (two knees, two shoulders and the left elbow) of NHO and presented late, after 2 years, in our training with stiffness in all involved joints. Early diagnosis (2 to 5 weeks after the triggering event) of this condition can be done by a biological assessment of urinary prostaglandin E2 (PGE2) and serum alkaline phosphatase levels, the latter

of which increases in the first two weeks and can multiply by 2 to 5 times the normal value in the 10thth week and normalize around the 18thth week. Early detection can also be done using imaging, including triphasic bone scintigraphy which is sensitive since it can detect lesions from the 2 -week and it also allows to follow the course of the lesions and to place the surgical indication, while the x-rays standard and CT scan detect lesions only from the 6th week. Magnetic resonance imaging (MRI) during the acute phase of NOH it allows to show an increase in vascularization and density of the tissues affected by the iso or hypersignal zones compared with muscle in T1 and hypersignal T2, during this phase, the MRI is non-specific and poses a problem of differential diagnosis with several other pathologies. However, the biopsy of lesions is not recommended as well as the imaging is sufficient to make the diagnosis [7].

In our case, given that the consultation was late, the radiographic lesions were very suggestive, with an obvious clinical context, and were sufficient to retain the diagnosis of NHO. Our case report highlights a rather unusual location of heterotopic ossification involving the first right chondro-costal joint, with intercostal bone bridges and a fracture of 3 ribs. Greiffenstein P *et al*., reported two cases of patients with intercostal heterotopic ossification (HO) after surgically managed chest wall trauma [15]. And A. M. Vergara *et al*., also described an intercostal localization of HO in a patient after Coronavirus disease 2019 (COVID 19) pneumonia, following which the patient stayed in the intensive care unit for respiratory distress [15]. In the literature, the incidence and impact of the thoracic localization of NHO are imprecise, and this localization is difficult to identify, given that the symptoms are non-specific (pain, limitation thoracic range of motion) and intercostal bone bridges are thin and difficult to visualize on standard radiographs [14].

In terms of therapeutic management, the first thing to mention is the prevention of this pathology, which, once formed, will be very tricky to treat. So, in patients with risk factors for developing NHO, prophylactic treatment must be put in place, even if their side effects may limit their use, but non- steroidal anti-inflammatory drugs are indicated, notably indomethacin, which is a non-selective cox inhibitor. Other preventive treatments are also possible, such as radiotherapy, range-of-motion exercises and bisphosphonates, and others are under investigation, including BMP (bone morphogenetic proetein) antagonists and the administration of retinoic acid. For curative treatment, surgery remains the only useful option when the ectopic bone is already mature in a patient with diminished joint amplitudes hampering daily activity, even if surgery itself can encourage HO formation. Hence the need to establish the surgical indication and choose the right time, which should be 12 to 18 months after HO formation [7]. This case report illustrates the difficulty of managing this pathology and the major functional impact on our patient's quality of life [7-14].

CONCLUSION

In conclusion, we report a case of NHO with multiple localizations in a young patient, which is responsible major functional handicaps, implying the need for prevention of this very disabling condition, whose management is very delicate.

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