CASO CLÍNICO/CASE REPORT

An Itch beyond the Skin: Syringomyelia Revealed by Localized Neuropathic Pruritus

Prurido para além da Pele: Siringomielia Revelada por Prurido Neuropático Localizado

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Abstract

A 67-year-old man presented with an eight-month history of persistent pruritus accompanied by a burning sensation in the left upper limb. Neurological examination revealed a pattern of hypoesthesia in the left upper limb that was inconsistent with a peripheral nerve or spinal root lesion; subsequent involvement of the right lower limb was also noted. The pruritus proved refractory to multiple therapeutic options. Neurophysiological studies showed no abnormalities. Magnetic resonance imaging of the cervicothoracic spinal cord revealed an intramedullary cystic lesion consistent with syringomyelia.

Neuropathic pruritus may result from peripheral or central sensory dysfunction, particularly within the spinothalamic tract, thalamus, or somatosensory cortex. As illustrated in this case, it may represent the predominant manifestation of syringomyelia, underscoring the importance of considering structural spinal cord causes in patients presenting with persistent and unexplained pruritus.

Resumo

Relatamos o caso de um homem de 67 anos com um quadro de oito meses de evolução de prurido persistente, acompanhado por uma sensação de ardor no membro superior esquerdo. O exame neurológico revelou um padrão de hipostesia no membro superior esquerdo que não era compatível com lesão de nervo periférico ou raiz espinhal; posteriormente, verificou-se envolvimento do membro inferior direito. O prurido mostrou-se refratário a múltiplas abordagens terapêuticas. Os estudos neurofisiológicos não evidenciaram alterações. A ressonância magnética da coluna cervicotorácica revelou uma lesão cística intramedular compatível com siringomielia.

O prurido neuropático pode resultar de uma disfunção sensitiva periférica ou central, nomeadamente ao longo do feixe espinotalâmico, do tálamo ou do córtex somatossensitivo. Como ilustrado neste caso, pode também constituir a principal manifestação da siringomielia, salientando-se a importância de considerar etiologias estruturais de mielopatia em doentes com prurido persistente e de causa não esclarecida.

Introduction

Pruritus is a common and unpleasant sensation that elicits an urge to scratch and may arise from dermatological, systemic (such as renal failure and cholestasis), psychosomatic or neurological conditions.[1] Neuropathic pruritus (NP) results from injury or dysfunction of the somatosensory nervous system, accounts for an estimated 8% of chronic itch cases,[2] and may impose a substantial burden on quality of life.[1] It may arise from lesions affecting the peripheral or central nervous system and is often accompanied by neuropathic sensory phenomena, such as pain, dysaesthesia or hypoesthesia.[3]

Case Report

We describe the case of a 67-year-old male patient with no relevant past medical history, referred from primary care to the Neurology outpatient clinic for an eight-month history of persistent pruritus associated with a burning sensation in the left upper limb. The symptoms had an insidious onset and progressively worsened over the following months, becoming more intense and persistent over time. There was no trigger, nor alleviating factors. No other symptoms were reported, including headache, cough, motor deficits or urinary dysfunction. Prior to referral, a basic laboratory investigation (complete blood count, creatinine, liver enzymes, serum electrolytes, B12 vitamin and folate levels) was unremarkable and cervical computed tomography (CT) only revealed incipient degenerative changes. Treatment with antihistamines, topical analgesics, and multivitamin supplements had not provided symptomatic relief.

At the Neurology consultation, no visible skin lesions were found on general inspection. Neurological examination revealed a pattern of hypoesthesia in the left upper limb that was not consistent with a peripheral nervous system distribution, with sparing of the fifth digit and medial forearm. No other relevant abnormalities were identified on neurological examination, including motor deficits, pathological hyperreflexia, or additional sensory deficits (namely suspended sensory level). Treatment with pregabalin was initiated but subsequently discontinued by the patient due to severe constipation. Nerve conduction studies and electromyography were unremarkable, making a peripheral nervous system lesion unlikely.

At four-month follow-up, symptoms persisted despite a therapeutic trial of duloxetine. Six months later, the patient developed pruritus in the right lower limb. Given the suspicion of a central nervous system etiology, brain and cervicothoracic spinal cord magnetic resonance imaging (MRI) was performed, revealing an intramedullary cystic lesion extending from T1 to T3, with cerebrospinal fluid-like signal intensity, and a thin left paramedian cavity extending cranially to C5 (**Fig. 1**). These findings were consistent with syringomyelia, and the patient was referred for neurosurgery evaluation to assess potential surgical management. After repeating the MRI of the spinal cord, the neurosurgeon decided to maintain a radiological surveillance strategy. The patient was also referred to a pain management consultation.

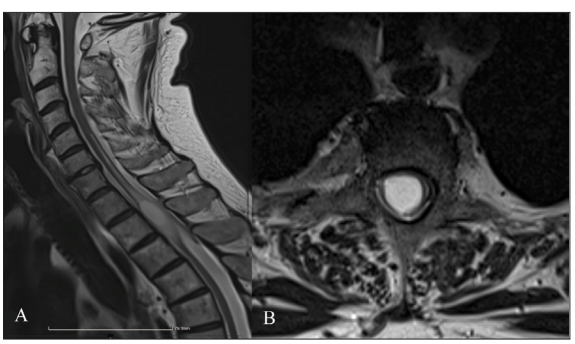

Figure 1. (A) Sagittal T2-weighted MRI image demonstrates an intramedullary fluid-filled cavity compatible with syringomyelia, with expansion from T1 to T3 and a thin, tapering syrinx extending superiorly to the C5 level. (B) Axial T2-weighted image at the T2 level illustrates a central cavity with peripheral displacement and compression of the spinal cord.

Discussion

Neuropathic pruritus, unlike conventional pruritus, occurs in the absence of primary skin lesions and reflects dysfunction of the somatosensory pathway of the nervous system. Its pathophysiology is thought to involve peripheral neuronal hyperexcitability, neuroinflammation, channel dysfunction and/or impaired central inhibitory control, leading to abnormal activation or amplification of itch-processing pathways.[4] Peripherally, itch is mainly transmitted by small-diameter primary afferents, particularly unmyelinated C-fibres and, to a lesser extent, Aδ-fibres, which also participate in nociceptive signaling. Centrally, pruriceptive input is relayed through the superficial dorsal horn and ascends via the spinothalamic tract to the thalamus and cortical areas involved in itch perception, including the somatosensory cortex, insula and anterior cingulate cortex.[3-5] Increased excitability and reduced inhibitory control may amplify itch transmission, contributing to spontaneous itch, al-

loknesis- itch evoked by normally non-pruritic stimuli- and hyperknesis- enhanced itch perception in response to pruritic stimuli.[4]

Spinal cord lesions are a recognized cause of central neuropathic pruritus, as they may disrupt the neural networks involved in itch modulation.[3] Reported causes include intramedullary tumours, inflammatory myelopathies such as neuromyelitis optica spectrum disorder, spinal cord injury, stroke, and syringomyelia.[3] In syringomyelia, progressive expansion of the syrinx within the spinal cord parenchyma, most commonly at cervical or cervicothoracic levels, may disrupt the dorsal horn circuits and adjacent ascending somatosensory pathways, including the spinothalamic tract.[6] This may generate a pruritogenic excitation–inhibition imbalance, with abnormal activation or amplification of itch-processing neurons and impaired inhibitory modulation of pruriceptive transmission. In our patient, disruption of spinal somatosensory pathways by syringomyelia, particularly within the anterolateral/spinothalamic system, may explain the associated dysaesthesia and hypoesthesia.[1,3,4]

The diagnosis of neuropathic pruritus remains challenging, as no specific diagnostic test is currently available, and recognition relies largely on clinical suspicion, careful neurological examination, and the exclusion of dermatological and systemic causes.[3] In our patient, pruritus was the predominant and most disabling symptom and occurred in the absence of relevant skin changes, while the associated hypoesthesia and dysaesthesia supported a neuropathic origin. The absence of dermatological or systemic findings prompted further neurological investigation. Initial investigations focused on a peripheral nervous system disorder due to the absence of clear signs of myelopathy on neurological examination, delaying the diagnosis of syringomyelia.

One of the most remarkable aspects of this case is the clear dissociation between the semiology and the extension of the spinal cord lesion. We hypothesize that the selective involvement of spinothalamic and dorsal horn itch-processing pathways, with relative sparing of adjacent motor tracts and other long sensory tracts, may account for this clinicoradiological dissociation.

Management of neuropathic pruritus is often difficult, as no therapies are currently approved specifically for neuropathic pruritus and pharmacological treatment is largely extrapolated from evidence on neuropathic pain. Commonly used agents include gabapentinoids, sodium channel blockers, antidepressants and, in localized cases, topical anaesthetics.[3,7] Despite these approaches, symptoms are often refractory and may substantially impair quality of life. In our patient, the optimal therapeutic scheme was not yet achieved, partially due to the side effects of these drug classes.

In conclusion, this case reinforces the importance of considering structural spinal cord lesions, including syringomyelia, in patients presenting with persistent, localized, and otherwise unexplained pruritus of suspected neuropathic origin. ■

Awards and Previous Presentations

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Contributorship Statement / Declaração de Contribuição

CMS: Conception, clinical evaluation of the patient, writing and final approval.

GC: Retrieval and neuroradiological interpretation of the MRI findings and final approval.

MJP: Conception, clinical evaluation of the patient, critical review and final approval.

CMS: Conceção, avaliação clínica do doente, redação e aprovação final.

GC: Obtenção e interpretação neurorradiológica dos achados de ressonância magnética e aprovação final.

MJP: Conceção, avaliação clínica do doente, revisão crítica e aprovação final.

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