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Scratching The Surface: Notalgia Paresthetica

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A 64-year-old woman presented to her primary care physician for a localized pruritic lesion on her upper mid back.

History. The patient described the lesion as a pruritic and slightly darker area on her upper mid back that had been present for the past 5 years. She endorsed increased size of the lesion per her husband's observation, and worsening pruritus. The patient noted the pruritus to be quite bothersome and would persistently scratch the area with a ruler. She found limited to no benefit with topical corticosteroid creams.

The patient had no significant medical history related to skin conditions, endocrine related conditions such as diabetes, allergies, or neurological disorders. The initial physical examination demonstrated a 6 cm x 8 cm slightly hyperpigmented patch with diffusely irregular borders accompanied by no secondary excoriation or satellite lesions (Figures 1 and 2).

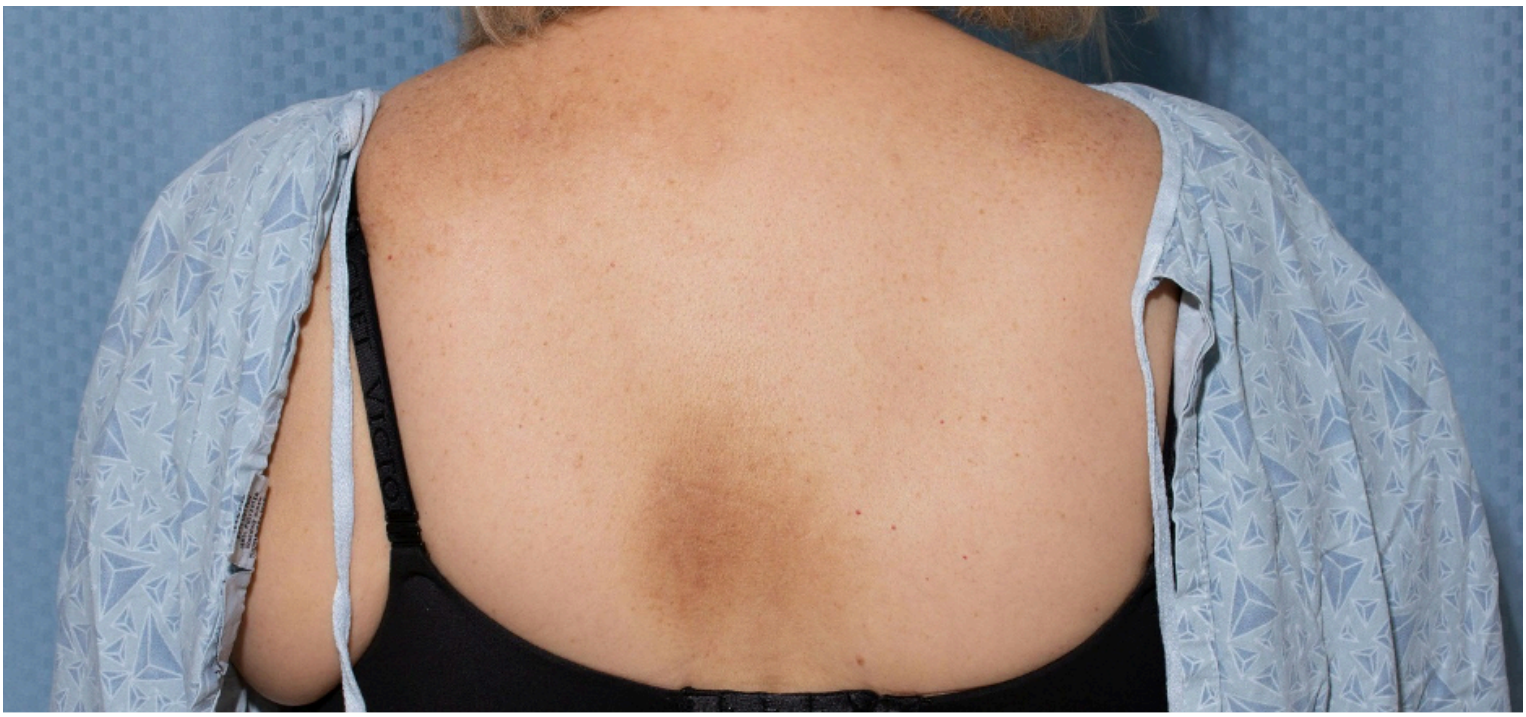

Figure 1. *The hyperpigmented patch inferomedially to the scapular blades.*

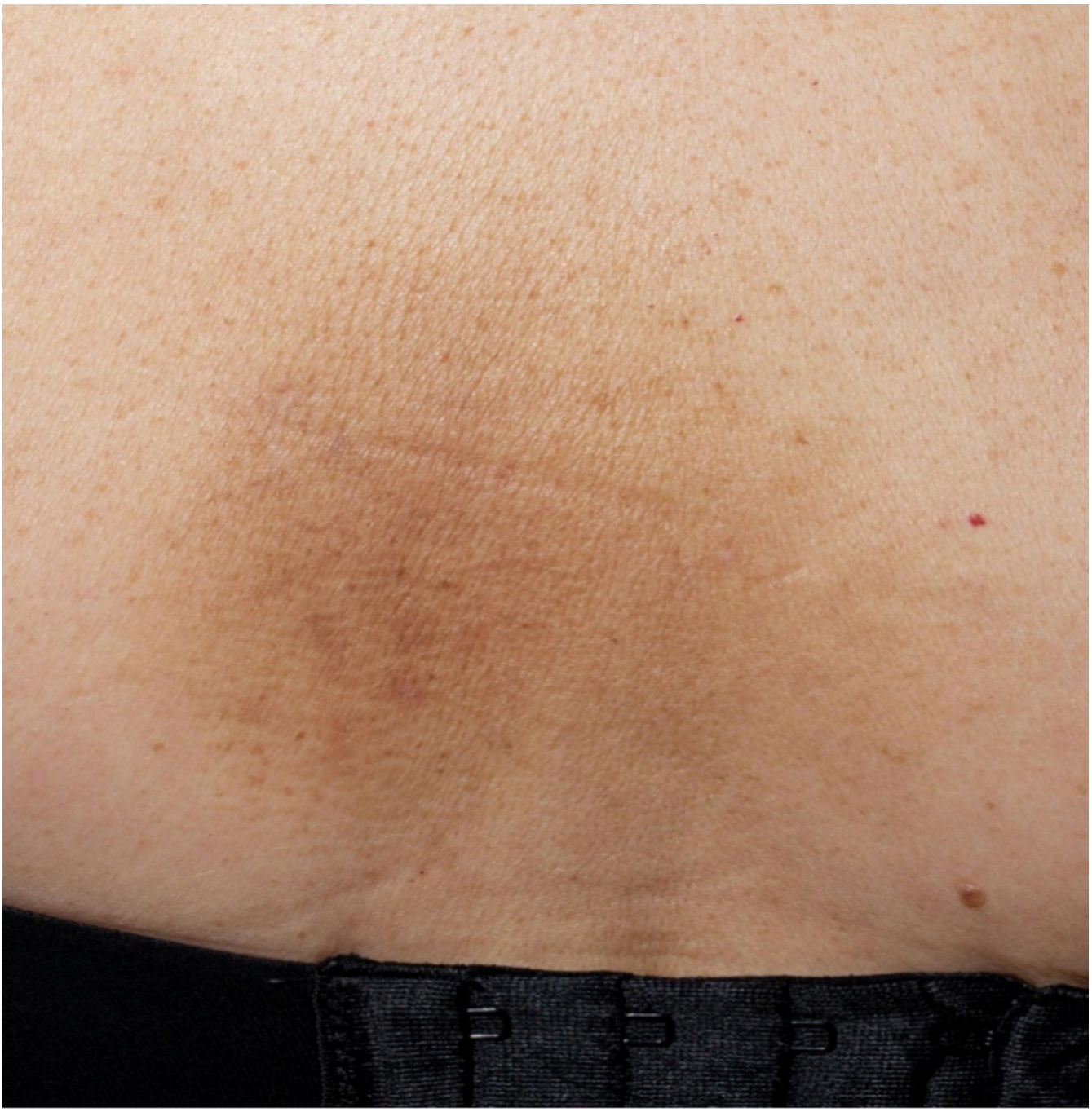

Figure 2. *Skin examination findings observed at a close range.*

Diagnostic testing. A skin biopsy was done at the time of initial symptom onset 5 years prior to the patient's current presentation, which revealed non-specific hyperkeratosis. She had continued to use topical corticosteroids without relief and was lost to dermatological follow-up. Given the chronicity and severity of symptoms, the patient re-presented to her primary care physician and was referred to the department of dermatology at our academic medical center for further re-evaluation, where a diagnosis of notalgia paresthetica was made based on clinical presentation. No further diagnostic testing or biopsy was deemed necessary.

Differential diagnoses. The differential diagnoses to be considered for such pigmented lesions include notalgia paresthetica (NP), contact dermatitis, patchy parapsoriasis, lichen simplex chronicus, macular amyloidosis, and tinea versicolor.[1]

Contact dermatitis is a skin reaction caused by external irritants or allergens and can occur anywhere on the body. Patchy parapsoriasis is a skin disorder with scaly patches on the skin and has potential association with lymphoproliferative disorders. Lichen simplex chronicus is a skin disorder with localized thickening and lichenification (thickened, leathery skin) due to chronic scratching of the affected area and it is often associated with eczema, psoriasis, or contact dermatitis. Macular amyloidosis is a skin disorder characterized by the deposition of amyloid protein in the skin, leading to the development of scaly brownish or greyish macules or patches on the upper back. Tinea versicolor is a fungal skin infection caused by the yeast *Malassezia*, which presents with small, scaly, discolored patches on skin.

In this case, the differential diagnoses were assessed and were ruled out based on the clinical presentation. Although not required to establish the diagnosis of NP, a biopsy can be considered to rule out additional pathology.

Treatment and management. Many treatment modalities exist, however, there is no strong evidence to suggest that any one treatment is superior to another. There are several treatments available, including topical agents, systemic agents, procedural modalities, and physical therapy.

Our patient's symptoms improved with a prescription of topical cream consisting of compounded amitriptyline 2%, gabapentin 5%, and ketamine, which she applied to the affected area four times daily. She was reassured that the spot is benign, and the pruritic sensation was secondary to a cutaneous sensory neuropathy.

Outcome and follow-up. There was significant improvement over the course of 7 months with the prescribed topical cream. No adverse effects were reported. Follow-up appointments were scheduled to provide further support and the patient provided satisfaction with the course of treatment a year later.

Discussion. Originally described in 1934 by Astwazaturow,[2] NP was described as an itchiness in the posterior region of the body. There have been several names given to the syndrome, including puzzling posterior pigmented purpuric patches, hereditary localized pruritus, puncta pruritica (itchiness), localized shoulder pruritus, and cutaneous lichen amyloidosis, but NP is the most accepted term.[3] It does not exhibit racial preference and is encountered across the globe.[4] Women are more likely to develop NP than men. In the majority of cases, the onset of symptoms occurs between the ages of 54 and 62 years.[6] However, the age of onset can manifest at a significantly younger age (with cases reported in children aged 6 years old) when it is associated with multiple endocrine neoplasia type 2A.[5] Isolated cases have also been reported in older adults.

It is widely accepted as a sensory neuropathy, arising from changes in the cutaneous branches of the posterior rami, notably the upper branches of T2-T6 spinal nerves.[6] However, the exact cause of NP is not fully understood. Contributing factors include dermal innervation, hereditary factors, viscerocutaneous reflex mechanisms, exposure to certain neurotoxic chemicals, and injuries to the spinal nerves resulting from trauma or compression.[7] In our case, there was no discerning factors that may have redisposed her to NP.

Our patient had the typical symptoms of pruritis and paresthesia, characterized by burning, tingling, surface numbness, tenderness, and foreign body sensations. There is often a patch of hyperpigmentation or hypopigmentation secondary to chronic scratching of the skin in patients with NP, as with our case. In most cases, these sensations are felt medial to scapula and lateral to the thoracic spine.[7] While symptoms are typically unilateral, interscapular and bilateral distributions have also been reported.

NP follows a chronic course, marked by periods of remissions and flare-ups. While not life-threatening, the cutaneous symptoms significantly impact the patients' quality of life.

In this case, the diagnosis was made using clinical examination. A biopsy may prove useful in excluding alternative diagnoses, but is not necessary.[1] Furthermore, spinal imaging is generally not recommended unless concurrent neurological or musculoskeletal symptoms are present.[8] Additional treatment options include topical agents (including capsaicin, amitriptyline/ketamine, lidocaine, anesthetic cream, or tacrolimus) and oral agents (such as gabapentin, amitriptyline, or oxcarbazepine).[6] Nonpharmacologic interventions such as physical therapy, intralesional therapy including botulinum toxin type A, narrowband ultraviolet b therapy,[9] nerve blocks,[10] and nerve ablation therapy have shown to be useful.[11] It has been proposed to begin treatment with topical agents with or without physical therapy and limit repeated botulinum toxin injections or nerve ablation therapy, given subsequent risk of iatrogenic motor neuropathy.[12]

Conclusion. This case report illustrates the clinical presentation and management of notalgia paresthetica, an often-undiagnosed condition which can cause significant adverse symptoms. Early recognition and appropriate treatment can greatly improve the quality of life.

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