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PATHOPHYSIOLOGY AND TREATMENT OF NOTALGIA PARESTHETICA- A SENSORY NEUROPATHIC SYNDROME OF THE BACK SKIN: AN OVERVIEW

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ABSTRACT

Notalgia paresthetica (NP) is a sensory neuropathic syndrome of the back skin, classically of the unilateral infrascapula. It is primarily a localized pruritus syndrome. Notalgia paresthetica was first named in 1934 and described as episodic itching or pain on a small patch of the mid back, usually an area of skin just past easy reach. Additional features of Notalgia paresthetica may include localized burning, pain, tenderness, hyperalgesia, or dysesthesias. Notalgia paresthetica may be associated with a poorly circumscribed tan or hyperpigmented patch in the symptomatic area. Notalgia paresthetica tends to be a chronic condition with periodic remissions and exacerbations. While not life threatening and not generally associated with other co-morbidities, it does frequently decrease quality of life causing much discomfort and nuisance to the affected patients. In the present article, we have concentrated on pathophysiology, mortality, morbidity, clinical features, symptoms, causes, treatment as well as prognosis of Notalgia paresthetica. The aim of present article is to provide in depth knowledge about clinical aspects related to Notalgia paresthetica.

KEYWORDS

Notalgia paresthetica, NP, Notalgia, sensory disturbance of the back, unilateral itching of back, interscapular itching, skin dysesthesia of back, localized pruritus syndrome, sensory neuropathic syndrome, hyperalgesia, unilateral back itching, subscapular pruritus, posterior pigmented pruritic patch and nostalgia.

INTRODUCTON

Notalgia paresthetica (NP) is a common, refractory sensory neuropathic syndrome with the hallmark symptom of localized pruritus of the unilateral infrascapula. It is generally a chronic, non-curable condition with periodic remissions and exacerbations. While the dermatologic syndrome may be multi-factorial in etiology, its possible association with underlying cervical spine disease needs to be evaluated for proper treatment. Radiographic studies of the spine may be more considered than they are currently. Collaborative multi-specialty evaluation by dermatology, radiology, neurology, and orthopaedics may be indicated

in primary management of this condition. First line therapy for notalgia paresthetica with associated cervical disease may include non-dermatologic spinal treatments such as spinal manipulation, physical therapy, massage, cervical traction, cervical muscle strengthening, and oral non-steroidal anti-inflammatory medications and muscle relaxants. Notalgia paresthetica may in fact be a dermatologic sign of an underlying systemic disease.

Notalgia paresthetica also known as Hereditary localized pruritus or Posterior pigmented pruritic patch, and Subscapular pruritus[1] is a chronic sensory neuropathy. Notalgia paresthetica is a common localized itch, affecting mainly the interscapular area especially the T2-T6 dermatomes, but occasionally with a more widespread distribution, involving the shoulders, back, and upper chest[2]. The characteristic symptom is pruritus (itch or sensation that makes a person want to scratch) on the back, usually on the left hand side below the shoulder blade (mid to upper back). It is occasionally accompanied by pain, paresthesia (pins and needles), and hyperesthesia (unusual orpathologically increased sensitivity of the skin to sensory stimuli, such as pain, heat, cold, or touch), which results in a well circumscribed hyperpigmentation of a skin patch in the affected area.

The causes or origin of this condition (etiology) have not yet been completely defined. Patients are usually older persons, and commonly find themselves scratching their back on doorposts etc, as the location can be hard to reach. The correlation of notalgia paresthetica localization with corresponding degenerative changes in the spine suggests that spinal nerve impingement may be a contributing cause.

PATHOPHYSIOLOGY

The exact pathophysiology of the cutaneous findings of notalgia paresthetica remains unknown. Although the etiology of notalgia paresthetica is unclear, two of the multiple proposed possible mechanisms include (1) localized increased sensory innervation of the affected skin areas and (2) neuropathy from degenerative cervico-thoracic disc disease or direct nerve impingement[3, 4, 5].

Savk et al in 2000 showed more than half of their patients had significant radiographic changes in the vertebrae corresponding to the dermatome of the cutaneous lesion. Further, all study patient's demonstrated normal neurological examination and standard electrodiagnostic results. All had skin histopathology compatible with post inflammatory hyperpigmentation. There were no amyloid deposits or other described pathology on pathologic exam of the skin. Springer et al in 1990 concluded that the symptoms of notalgia paresthetica may in part be related to an increase in the sensory epidermal innervation in the affected skin areas. Histological studies have shown cutaneous changes in a few cases including lichen amyloid which may be secondary to the localized chronic scratching and rubbing[6].

MORTALITY/MORBIDITY

While not life threatening and not generally associated with other non-spine co-morbidities, the cutaneous symptoms of notalgia paresthetica frequently decrease quality of life causing much discomfort and nuisance to the affected patients. There may be some increased morbidity because of the possible underlying cervical and thoracic spine and disc disease. Notalgia paresthetica tends to be a chronic condition with periodic remissions and exacerbations. There is no described increased mortality with this disorder. Notalgia paresthetica may be seen in all races without any described racial predilection. Notalgia paresthetica may be seen in both males and females, although there seems to be an increase in females. Notalgia paresthetica is more common in adulthood, typically in ages 40-80[7].

SYMPTOMS AND CLINICAL FEATURES

Notalgia paresthetica (NP) patients often present with the hallmark symptom of localized pruritus of the unilateral infrascapula. Notalgia paresthetica classically presents with skin findings of a unilateral, ill defined, tan, pink, or

hyperpigmented non-indurated patch of the infrascapular back (mid back). The affected skin area usually ranges in size from 3-10cm. Secondary skin changes such as lichenification, lichen amyloid, excoriations, eczema, xerosis, and secondary infection may be noted. There may be associated mild sensory alternations to light touch, vibration, and pin prick. Examination of the spine may be normal or reveal tenderness, decreased range of motion in the neck, and possible associated cervical muscle spasm.

CAUSES

The exact cause of the cutaneous findings of notalgia paresthetica remains unknown. Notalgia paresthetica may in fact be a dermatologic sign of an underlying systemic disease. Notalgia paresthetica may not be solely a skin disease per se but a cutaneous sign of an underlying degenerative cervical spine disease. The striking association of notalgia paresthetica with degenerative or traumatic cervico-thoracic spine disease suggests that early spinal nerve impingement may contribute to the pathogenesis of the skin symptoms of the disease. Additional studies are needed to further assess the relationship of notalgia paresthetica with cervical spine disease. Whether this is a causal or coincidental finding remains to be determined in larger studies. While topical therapies may in some cases seemingly help decrease the localized symptoms in notalgia paresthetica, systemic or broader scope spinal evaluation may be warranted to fully evaluate refractory cases. Cervical spinal imaging and treatment may be appropriate as primary or first line therapy in many cases of notalgia paresthetica.

LAB STUDIES

Although laboratory tests are generally not required in the workup of notalgia paresthetica, a basic pruritus workup may be helpful in select cases based on history and contributory symptoms.

IMAGING STUDIES

Although imaging tests have traditionally not been a part of the workup of notalgia paresthetica, basic cervical and possibly thoracic x-rays or MRI may be warranted in the initial management of the disorder. Imaging studies may be particularly helpful in patients with contributory spine symptoms of pain, tenderness, spasm or decreased range of motion and any history of spinal trauma or injury.

HISTOLOGIC FINDINGS

Skin biopsy and tissue histology are usually not indicated for the diagnosis of notalgia paresthetica. Biopsies may be done to exclude other diagnosis and neoplasms. There are no described criteria for tissue diagnosis of notalgia paresthetica. Prior studies have shown various histologic findings including post inflammatory hyperpigmentation and lichen amyloid.

TREATMENT

Treatments of notalgia paresthetica with topical modalities have generally failed and are refractory because of the difficult to reach location. To date, there has been no clearly described etiology and no uniformly effective treatment for notalgia paresthetica. Topical therapies aimed at the back skin may be in fact being ineffectual or partially effective as basic emollients. Since notalgia paresthetica does have periodic spontaneous remissions and exacerbations, it may be difficult to accurately measure response to various therapies. A placebo response may be considered with some therapies. During the initial assessment of patients with notalgia paresthetica, it is important to obtain a thorough past history of osteoarthritis, prior neck trauma, motor vehicle accident, vertebral fracture, cervical neoplasm or malignancy, or cervical disc disease. Even in the absence of positive medical history, radiographs or MRI of the cervical spine may aid in early diagnosis and treatment of degenerative spine disease. The striking association of notalgia paresthetica with degenerative or traumatic cervico-thoracic spine disease suggests that early spinal nerve

impingement may contribute to the pathogenesis of the skin symptoms of the disease. Additional studies are needed to further assess the relationship of notalgia paresthetica with cervical spine disease.

Whether this is a causal or coincidental finding remains to be determined in larger studies. While topical therapies may in some cases seemingly help decrease the localized symptoms in notalgia paresthetica, systemic or broader scope spinal evaluation may be warranted to fully evaluate refractory cases. Cervical spinal imaging and treatment may be appropriate as primary or first line therapy in many cases of notalgia paresthetica. In the future, first line therapy for notalgia paresthetica with associated cervical disease may include non-dermatologic, non-invasive treatments such as spinal manipulation, physical therapy, cervical soft collars, massage, cervical traction, cervical muscle strengthening and increased range on motion, transcutaneous electrical nerve stimulation, cervical discectomy with fusion, oral non-steroidal anti-inflammatory medications (ibuprofen, celecoxib, ketoralac) and oral muscle relaxants (carisoprodal, cyclobenzapril, methocarbamol, metaxalone). Other medical and surgical measures for degenerative disc cervical disease and nerve impingement as introduced may also be considered. For more generalized and chronic pruritus, full laboratory workup including complete blood count, chemistry panel including renal and liver functions, chest x-ray, and other studies may be warranted to exclude underlying physiologic causes of pruritus. Alternatively, proper management of NP may involve a multi-specialty cooperative effort of dermatology with radiology, orthopaedic surgery, neurology, and possibly adjunctive fields including acupuncture, chiropractic, and physical therapy.

SURGICAL CARE

Surgical therapy for notalgia paresthetica with associated cervical disease may include discectomy with fusion, disc replacement surgery, minimally invasive injectable disc repair techniques, and other surgical measures for degenerative cervical disease and nerve impingement.

CONSULTATIONS

Proper evaluation and management of notalgia paresthetica may involve a multi-specialty cooperative effort of dermatology with radiology, orthopaedic surgery, neurology, pain management, and possibly adjunctive fields including acupuncture, massage, chiropractic, and physical therapy. Consultations with other specialists may be warranted based on radiologic findings and individual patient history and physical exam.

MEDICATION

While to date there has been no uniformly effective treatment for the cutaneous symptoms of notalgia paresthetica, common first line medications include potent topical steroid creams. Currently available therapeutic options for the localized itch syndromes include capsaicin cream[8], eutectic mixture of local anesthetic (EMLA) cream, topical steroids, pramoxine cream, topical cooling or ice pack applications, oral steroids, Tiger balm, menthol creams, flurandrenolide tape (Cordran Tape), intralesional corticosteroid injections, botulinum toxin injections[9], oral antihistamines, hydroxyzine, doxepin, topamax, anticonvulsant medications, carbamazepine (Tegretol) antidepressant medications, gabapentin (Neurontin), oxcarbazepine[10], topiramate, thalidomide[11], and many others. It is possible that some of the current systemic therapies may in fact exert their effect through the spinal nerves and central nervous system thereby supporting the neuropathic etiology of notalgia paresthetica[12].

First line therapy for notalgia paresthetica with associated cervical or cervicothoracic disease may include non-dermatologic medications such as oral non-steroidal anti-inflammatory medications (ibuprofen, celecoxib, ketoralac) and oral muscle relaxants (carisoprodal, cyclobenzapril, methocarbamol, metaxalone). Other medical and surgical measures for degenerative cervical disc disease and nerve

impingement as introduced may also be considered.

Drug Category: Corticosteroid, Topical (Very High Potency)

Drug Name	Clobetasol Propionate
Description	Class I super potent topical steroid; suppresses mitosis and increases synthesis of proteins that decrease inflammation and cause vasoconstriction. Decreases inflammation by stabilizing lysosomal membranes, inhibiting PMN and mast cell degranulation.
Adult Dose	Apply bid for up to 2 wk; not to exceed 50 g/wk
Paediatric Dose	<12 years: Not recommended >12 years: Administer as in adults
Contraindications	Documented hypersensitivity; viral or fungal skin infections
Interactions	None reported
Precautions	May suppress adrenal function in prolonged therapy
Pregnancy	C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Drug Category: Corticosteroid, Topical (High Potency)

Drug Name	Fluocinonide
Description	High-potency steroid, inhibits cell proliferation, is immunosuppressive, antiproliferative, and anti-inflammatory. Also have antipruritic and vasoconstrictive properties.
Adult Dose	Apply sparingly bid/qid as severity warrants
Paediatric Dose	Administer as in adults
Contraindications	Documented hypersensitivity; herpes simplex infection; fungal, viral, or tubercular skin lesions
Interactions	None reported
Precautions	May cause adverse systemic effects if used over large areas, denuded areas, on occlusive dressings, or during prolonged treatment periods
Pregnancy	C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Drug Category: Anti-Pruritus

Drug Name	Hydroxyzine hydrochloride
Description	Antagonizes H1 receptors in periphery. May suppress histamine activity in subcortical region of CNS.
Adult Dose	25-100 mg PO qd/qid
Paediatric Dose	0.6 mg/kg/dose PO q6h
Contraindications	Documented hypersensitivity
Interactions	CNS depression may increase with alcohol or other CNS depressants
Precautions	Associated with clinical exacerbations of porphyria (may not be safe for porphyric patients); ECG abnormalities (alterations in T-waves) may occur; may cause drowsiness
Pregnancy	C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Drug Category: Analgesic, Topical

Drug Name	Capsaicin
Description	Natural chemical derived from plants of Solanaceae family. Penetrates deep for temporary relief of minor aches and pains of muscles and joints associated inflammatory reactions. Derived from plants of Solanaceae family. May render skin and joints insensitive to pain by depleting substance P in peripheral sensory neurons. Has demonstrated effectiveness in several studies of diabetic neuropathic pain and in other types of neuropathic pain.
Adult Dose	Apply to affected area tid/qid for 3-4 consecutive wk and evaluate efficacy; not to exceed 4 applications/d; wash hands with soap and water after applying
Paediatric Dose	Not established
Contraindications	Documented hypersensitivity; broken or irritated skin
Interactions	None reported
Precautions	For external use only; avoid contact with eyes; do not use tight bandage; discontinue use if condition worsens or symptoms persist for 14-28 d
Pregnancy	C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

Drug Category: Corticosteroid, Topical (Medium Potency)

Drug Name	Triamcinolone
Description	For inflammatory dermatosis responsive to steroids; decreases inflammation by suppressing migration of polymorphonuclear leukocytes and reversing capillary permeability. Available in ointment (0.1%) and cream (0.025%, 0.1%, 0.5%).
Adult Dose	Apply thin film bid/tid to response
Paediatric Dose	Apply as in adults
Contraindications	Documented hypersensitivity; fungal, viral, and bacterial skin-infections
Interactions	None reported
Precautions	Do not use in decreased skin circulation; prolonged use, applications over large areas, and use of potent steroids and occlusive dressings may result in systemic absorption; systemic absorption may cause Cushing's syndrome, reversible HPA axis suppression, hyperglycemia and glycosuria
Pregnancy	C - Fetal risk revealed in studies in animals but not established or not studied in humans; may use if benefits outweigh risk to fetus

PROGNOSIS

Notalgia paresthetica tends to be a chronic disease with periodic remissions and exacerbations. The prognosis for control of the symptoms is good. It is not curable[13, 14].

CONCLUSION

Notalgia paresthetica may not be solely a skin disease per se but a cutaneous sign of an underlying degenerative cervical spine disease. The striking association of notalgia paresthetica with degenerative or traumatic cervico-thoracic spine disease suggests that early spinal nerve impingement may contribute to the pathogenesis of these skin symptoms of the disease. Additional studies are needed to further assess the relationship of notalgia paresthetica with cervical spine disease. Whether this is a causal or coincidental finding remains to be determined in larger studies. While topical therapies may in some cases seemingly help decrease the localized symptoms in notalgia paresthetica, systemic or broader scope spinal evaluation may be warranted to fully evaluate refractory cases. Cervical spinal imaging and treatment may be appropriate as primary or first line therapy in many cases of notalgia paresthetica. Patient education involves discussion of possible underlying causes and associations with cervico-thoracic spinal disease. Patients need to be advised of potential disease flare with exacerbations of their spinal disease.

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