Dermatological-Neurological Interactions
Timothy Berger, MD

The term neurocutaneous dermatoses is defined as a dermatitis that arises secondary to pathology located at any point along the circuitry of the nervous system. They can be divided into 3 subgroups- anatomical, neurovascular and primary sudomotor, based on the underlying mechanism presumed to be responsible primarily for producing their clinical features.

Neuropathic pruritis: persistent picking and scratching at the skin is often very evident upon evaluation. This often suggests an underlying neurological etiology. Lichenification and prurigo, papules/nodules are often the most common manifestations of the unrelenting cycle of itching and scratching. These lesions are often accompanied by lichenification (hyperplasia in response to chronic itching).

Intractable localized or segmental pruritis without evidence of a primary dermatological process is named dependent on the body part affected. Brachioradialus pruritis, nostalgia paresthetica and anogenital pruritis are three of the more common conditions. These are often manifestationsof mechanical injuries affecting the vertebral column or nerve roots. The anatomical distribution of them corresponds to the respective level of injury: brachioradial pruritis to the cervical, nostalgia paraesthetica the thoracic and anogenital pruritis to the lumbosacral region.

Brachioradialus pruritis is characterized by localized,severe, refractory, intractable itiching and secondary skin changes such as lichenification. The skin over the proximal heads of the brachioradialus muscles is affected, usually associated with clinical evidence of chronic sun damage. On occasion, the pruritis extends across the upper back. Heyt demonstrated radiographic evidence of osteoarthritis in four of five patients, and hypothesized that nerve injury, resulting from either lesions of the cervical spine or from mechanical compression by nearby structures. Goodkin et al., have provided further radiographic proof that cervical spine disease may be an important contributing factor. In their research, they fouind that 50% of their patients had radiologically verified degenerative changes in the cervical spine relating to the level of pruritis presented. Cervical root compression as a cause, was further supported by a report of spinal cord tumor causing pruritis at the C5-6 dermatomes. Using this information, it is now thought that brachioradialus pruritis may respresent a form of neuropathic pruritis. Sunlight, however, can also be an eliciting factor and compression of the cervical nerve root a predisposing factor in these patients. Wallengren and Sundler studied cutaneous biopsies from the affected skin of patients with brachioradialus pruritis to normal skin from age-matched controls. They visualized the cutaneous innervation by antibiodies to protein gene 9.5, a general neuronal marker, calcitonin gene-related peptide, a marker for thin sensory nerve fibers, and the VR-1 receptor, a marker for capsaicin-sensitive nerve fibers. They found that pruritis was seasonal in most of their patients and histologically the biopsied specimens displayed changes similar to those seen in skin affected by ultraviolet light. The number of nerve fibers in the diseased skin, as detected by nerve markers was significantly reduced as compared to the controls. Of note, some patients' fiber counts

were normalized as the symptoms abated. Wallengren and Sandler argue that spinal disease alone cannot explain symptoms of brachioradialus pruritis, based on their observations of symptom free periods interspersed with relapses. An affected family suggests the presence of hereditary brachioradialus pruritis, either with auto-somal dominant or x-linked inheritance.

Patients often apply ice packs to the affected arm. Sunscreen may also be of partial benefit. Topical analgesics are also helpful, as well as, some surgical and pharmaceutical interventions. Cervical physiotherapy, cervical manipulation and acupuncture have also been reported to be helpful in the treatment of this disorder. Tait and colleagues found that 71% of patients treated with conservative chiropractic care reported resolution of symptoms following manipulative treatment, including those who had documented cervical spine disease with or without cervical symptoms.

Notalgia paresthetica is usually a unilateral sensory neuropathy found mainly in the older patient characterized by infrascapular pruritis, burning pain, tenderness or hyperalgesia. Occassionally, other neurological symptoms are present as well. These may include numbness, tingling and formication. Usually, the dermatomal distribution is T2-6 without evidence of primary dermatologic lesions. Pigmented patches and amyloidosis may occur from the rubbing of the skin. It is hypothesized that the course that the nerve root when taking the 90 degree course as they exit the spine may contribute to the increased susceptibility to mechanical injury.

Pathogenesis is possibly linked to musculoskeletal compression and resultant pinal nerve impingement of the posterior primary rami of the aforementioned spinal nerves. Raison-Peyron et al., have radiologically demonstrated evidence of spinal arthrosis in patients with NP. Savk has also demonstrated a correlation between patients with NP and spinal degenerative changes. In both cases, the patients demonstrated the same dermatomal distribution of pruritis as structural abnormalities.

Some hereditary cases have occurred, mainly in young patients associated with multiple endocrine neoplasia type 2A. The associated cutaneous lesions are hypothesized to be secondary to pathology in the neural crest-derived dorsal sensory nerves.

Treatments for this disorder have responded favorably to topical analgesics, OTC and pharmaceautical grade medications, paravertebral anaesthetic blocks, epidural steroid injections and physiotherapy. Most of these treatment modalities affect directly or indirectly on the nervous system, supporting the neurologic etiology theory.

Anogenital pruritis is a general term for itching which is almost always limited to the genital or perianal skin. There is usually little or no symptoms at other anatomical sites. Cutaneous manifestations are identical to those of lichen simplex chronicus elsewhere on the body. Attacks are characterized with violent itching with nocturnal pruritis common. Generally, the pruritis is restricted to one anatomical area and does not cross over to another. These conditions are very common and poorly managed. Anogenital itching may be a manifestation of pre-existing pathology or an idiopathic, primary condition, in

which persistent itch may be present. A thorough history and evaluation will differentiate AP from hemorrhoids or anal/genital neoplasms. Recently, there have been studies which have found that there is a possible neuropathic origin for idiopathic anogenital pruritis. This mechanism is similar to that of NP and BP. Patients with AP have been to be observed to have lumbar radiculopathy, representing nerve or nerve root compression at the L4-S2 vertebral levels. These levels have demonstrated degenerative changes of lower spine on radiopgraphs.

Treatment of AP usually involves eliminating exacerbating factors such as irritants and potential sensitizers, with modifications of hygiene habits. Short courses of anti-histamines and high-potency topical steroids.