Neurocutaneous disease

Neurocutaneous dysesthesias

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Learning objectives
After participating in this learning activity, participants should be able to properly detect patients with neurocutaneous disease, formulate a treatment regimen for cutaneous dysesthesias, and recognize patients who could benefit from referral to another specialist.

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Dysesthesia is a generic term for a cutaneous symptom—such as pruritus, burning, tingling, stinging, anesthesia, hypoesthesia, tickling, crawling, cold sensation, or even pain—without a primary cutaneous condition in a well-defined location that is often caused by nerve trauma, impingement, or irritation. There are multiple types of dysesthesias depending on the body location and the nerves involved. While location, exact symptoms, and etiologies might vary, the underlying theme is that these conditions are of neurologic origin and have dermatologic consequences. For many of these conditions, the symptoms are localized to the skin, and patients frequently present to the dermatologist; it is important for dermatologists to be knowledgeable about these symptoms and their underlying causes. In part II of this continuing medical education review, the primary diagnoses associated with underlying cutaneous dysesthesias will be explored, including scalp dysesthesia, trigeminal trophic syndrome, meralgia paresthetica, notalgia paresthetica, and brachioradial pruritus. The typical demographics in terms of symptoms, location, and patient populations will be discussed in addition to the specific etiologies, workups, and possible treatment options. (J Am Acad Dermatol 2016;74:215-28.)

Key words: brachioradial pruritus; burning scalp syndrome; dysesthesia; macular amyloidosis; meralgia paresthetica; neurocutaneous; notalgia paresthetica; scalp dysesthesia; trigeminal trophic syndrome.

A summary of neurocutaneous diseases is available in Table I.

SCALP DYSESTHESIA

Key point

- **Thought to be caused by either psychiatric conditions or nerve trauma either indirectly through muscle tension or directly through surgical trauma**

Demographics

Scalp dysesthesia, also known as burning scalp syndrome, is indicated by burning, pruritic, or stinging sensations felt on the scalp of patients in the absence of a primary cutaneous disorder.[1,2] These

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Abbreviations used:

ASIS:	anterior superior iliac spine
BRP:	brachioradial pruritus
LFCN:	lateral femoral cutaneous nerve
MP:	meralgia paresthetica
NP:	notalgia paresthetica
OMT:	osteopathic manipulative treatment
TENS:	transcutaneous electrical nerve stimulators
TTS:	trigeminal trophic syndrome

sensations can be either diffuse or localized.[2] Although previous case reports were comprised entirely of women,[1,2] there has been no thorough epidemiologic evaluation, and the true sexual predilection is unknown. The incidence is also unclear, because most studies have used patient response questionnaires to look for "sensitive skin" localized to the scalp and therefore can not ensure that primary cutaneous diseases were not included.[3-5]

Etiology

Scalp dysesthesias have been seen in three patient populations; patients with a history of a psychiatric condition,[1] cervical spine disease,[2] and/or a history of a facial or brow lift.[6] The psychiatric conditions described in association with scalp dysesthesias were dysthymic disorder, generalized anxiety, and somatization.[1] However, another retrospective review found no association between psychiatric disease, stress levels, and cutaneous symptoms.[2] Therefore, the relationship between psychiatric conditions and scalp dysesthesias is not well understood, as is whether there is causation or rather an unknown confounder. It is unclear if this association drives the primary disease or enhances patient propensity to be disturbed by these sensations.

The other two etiologic theories derive from neurologic trauma causing symptoms either more centrally with cervical spine disease or iatrogenically induced to peripheral nerves through surgical procedures. In the case of cervical disease, patients with scalp dysesthesias had a higher rate of C5-C6 radiographically localized cervical spine disease.[2] The nerves that originate from this C5-C6 location innervate the posterior neck and do not directly supply the scalp; the posterior scalp originates from C2.[7] The hypothesis is that the nerve impingement does not cause a dermatome-related symptom, but rather that the symptoms are related to chronic tension placed on the occipitofrontalis muscle and scalp aponeurosis as a result of primary cervical spine disease.[2] When scalp dysesthesia is caused by a previous facial or brow lift, it is often because of surgical trauma to the superficial nerves of the face and scalp.[6]

Workup

Before making a diagnosis of scalp dysesthesia, the practitioner must ensure no primary cutaneous disorders are causing these symptoms, such as irritant contact dermatitis, seborrheic dermatitis, atopic dermatitis, or psoriasis.[1] If patients report a history of headaches or temporal pain caused by palpation, tension headaches or temporal arteritis need to be considered.[1] Once other diagnoses have been eliminated based on history or physical examination, the diagnosis of scalp dysesthesia can be entertained. As mentioned above, there are two main theories as to the etiology of the symptoms patients experience with scalp dysesthesia: psychologic and anatomic. Therefore, the two separate etiologic theories have two separate workup algorithms. While there might be a connection with underlying psychiatric conditions, there are not enough data to support psychiatric screening in these patients.[1] Because of the relationship with previous facial or brow lift surgical procedures, this is an important component of the history to ascertain. Because there has been a reported connection with cervical spine disease, one could argue for routine screening—however, this has only limited evidence.[2]

Treatment

Data regarding appropriate treatment strategies are lacking; many treatment options are based on case reports and have not been compared against each other, including oral gabapentin, topical gabapentin, topical corticosteroids (both high potency and low), antidepressants (eg, venlafaxine and amitriptyline),[2] doxepin,[1] and pregabalin.[8] One case series with patients who had cervical spine–related symptoms had no improvement in their symptoms with physical therapy.[2]

TRIGEMINAL TROPHIC SYNDROME

Key points

- **Trigeminal trophic syndrome most commonly affects the V2 branch of the trigeminal nerve, resulting in ulceration of the nasal ala**
- **The most common cause of trigeminal trophic syndrome is trigeminal nerve ablation for trigeminal neuralgia and cerebral vascular accidents**
- **Treatment includes physical barriers, pharmacologic interventions, and surgical repairs**

Demographics

Trigeminal trophic syndrome (TTS) is a condition in which abnormal sensations, as a result of trigeminal nerve injury, leads to self-inflicted ulceration of