Brachioradial Pruritus: Orphan Patients in Search of Understanding and Care

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ABSTRACT

The authors present a case study around an entity named ‘brachioradial pruritus’ a neurogenic itch which typically occurs on the dorsolateral forearm but may also involve the upper arm and shoulders, and less commonly the back. The disease at this time does not have a known treatment and the authors not only discuss the isolation of these patients which is exacerbated by the ignorance or indifference of most medical personnel to this often debilitating condition but also from their shared learning experiences from a web based platform propose an algorithm that may be useful for others who need to deal with this condition.

Keywords: Algorithm, Brachioradial Pruritus, Orphan Patients, Virtual Grand Rounds in Dermatology, Web Based Platform

In the early 1980s one of us (DJE) encountered many patients in Hawaii with a singular and peculiar painful and pruritic dermatosis on the arms. The first article on the disorder that would be called Brachioradial Pruritus (BRP) came from Florida and dated back to 1968 (Waismann, 1968). Fortuitously, the next article (Heyl, 1983) which reported cases from South Africa, appeared in the Archives of Dermatology in 1983 when we were first seeing similar patients in Hawaii. Initial conclusions were that this was a solar dermopathy. Our 1986 article seemed to confirm that most patients were from sunny climes (Waleyk & Elpern, 1986). Sadly, however, we have not progressed much in the past twenty-five years and do not have a real understanding of what BRP is, and lacking that no accepted therapeutic protocol.

Patients with BRP inhabit the murky domain of the “orphan patient” (OP). Walter and Dorinda Shelley defined the OP in their 1988 letter as “one with a unique, inchoate, baffling, and often disabling disease not yet clearly discernible in the medical literature. Although technically this patient has an orphan disease — one defined as affecting fewer than 200,000 people in the United States — in reality he or she is an outlier, standing alone” (Shelley & Shelley, 1988).

Patients with BRP fall into the category of OPs. The prevalence of BRP in the United States is unknown, but the condition has been documented in six states (Waismann, 1968; Waleyk & Elpern, 1986; Fisher, 1997; Bernhard, 2007; Massey & Massey, 1986; Kestenbaum, 1979). Because the disease is uncommon, patients af-
flicted with this condition often find themselves standing alone, without any solid medical or social support. In addition, many physicians are not familiar with the condition, and those that are do not have an effective therapeutic protocol. This means that patients are either dismissed as having trivial symptoms, or are repeatedly offered ineffective treatment. In truth, BRP does not, at this time, have a known treatment. The problem we wish to address here is not the lack of effective treatment but the isolation of these patients which is exacerbated by the ignorance or indifference of most medical personnel to this often debilitating condition. Many patients with the disease feel alone and hopeless, and become frustrated with the medical system.

BRP is a neurogenic itch which typically occurs on the dorsolateral forearm but may also involve the upper arm and shoulders, and less commonly the back. It may be unilateral or bilateral (Veien, 2001). It has been reported in patients of all skin types (Walcyk & Elpern, 1986), genders (Wallengren, 2005) and geographic climates. Patients generally describe BRP as an intense periodic itch that originates “under the skin.” Scratching is irresistible, and it usually makes the itch worse. The itch is only reliably relieved by ice-packs. Other topical and oral treatments appear to be less effective.

It is accepted that BRP is a primary neuropathy, but there is disagreement as to the anatomical location of the lesion causing the neuropathy. Two theories predominate; the solar hypothesis and the cervical nerve root injury hypothesis. The solar hypothesis places the cause of peripheral nerve damage on overexposure to solar radiation (Kestenbaum, 1979; Wallengren, 2005; Veien, 2001), whereas the nerve root hypothesis postulates that injury to any of the C5-C8 nerve roots is the culprit (Heyl, 1983; Crevitz, 2006; Barry, 2004; Goodkin, 2003). Many BRP patients appear to have both mechanisms at work.

A panoply of treatments have been reported to reduce the severity of symptoms in some patients, but have failed to show reproducible benefit in others. No universally accepted treatment for BRP exists at present. This means that physicians should approach each BRP patient as if he/she were unique, a method that requires considerable background knowledge, flexibility and complete honesty on the part of the physician. The idea here is not only to alleviate the patients’ symptoms, but also to give them a sense of comfort and reassurance – comfort that what they are experiencing is a known medical condition and reassurance that their physician is familiar with BRP and will give the patient his full attention and effort.

Below are two cases posted on Virtual Grand Rounds in Dermatology (VGRD) (vgrd.blogspot.com), an online forum where unique and challenging dermatology cases are posted by physicians and commented on by colleagues from around the globe. There have been 325 posts on the VGRD blog since 2004, and 859 comments to date, giving an average of 2.6 comments per post. Of note, there have been 55 comments made on the BRP post, over 90% of which were made by patients suffering from this condition.

**CASE 1. 32 YEAR OLD EQUESTRIAN**

I am an otherwise healthy woman desperately searching for a doctor who may be able to discuss some treatment options for my very itchy arms! I am 32 year old, 110 lb, active, female resident of New England and have been suffering with itchy arms (without rash) for at least four years now. When I finally sought treatment- I was referred by my primary MD to a dermatologist who informed me that I had dry, sensitive skin, and prescribed a topical cream and an oral steroid - I carefully followed his advice for 2 years - with absolutely no relief from the itch. The only thing that relieved the symptoms (which flare more intensely in the evening hours) was ice packs. I was often woken from sleep by the itch.

The itch occurs in cyclical pattern- symptomatic for months, followed by a month or two of none or very mild symptoms, then followed by another flare. It is present in sunny months as well as winter.
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