Brachioradial pruritus in a young man presenting with transverse myelitis

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ABSTRACT

Pruritus with unknown origin is a common complaint and secondary causes should be investigated. We report a 22-year-old man with intractable localized pruritus and painful burning, stinging sensations in the bilateral arms, shoulders, neck and upper back for 3 weeks. A dermatologist was consulted in the emergency department. On physical examination, there were multiple excoriated erythematous macules and depigmented scars in a linear distribution over the dermatome from C4 to C6. Decreased muscle power in bilateral upper limbs was also noted. A consultant dermatologist strongly suggested that an image survey on the cervical spine was necessary. Surprisingly, a magnetic resonance image (MRI) of the cervical spine revealed a lesion with T1 hypointensity and T2 hyperintensity occupying two-thirds of the intramedullary region at the C2–C5 levels, consistent with transverse myelitis. Brachioradial pruritus with transverse myelitis was diagnosed. It is important for clinicians to be aware that brachioradial pruritus should be a differential diagnosis of pruritus of unknown origin and that the disease entity is not responsive to systemic corticosteroids and antihistamines; however, it is dependent on treatment of the underlying process. Once the diagnosis of brachioradial pruritus is made, plain film or MRI of the cervical spine should be arranged to determine the presence of possible nerve compression lesions.

KEYWORDS

Brachioradial pruritus
Notalgia paresthetica
Pruritus of unknown origin
Transverse myelitis

Introduction

Brachioradial pruritus is a rare form of neuropathic itch.1 It was first described by Waisman2 as solar pruritus in 1968 and was thereafter regarded as photodermatosis for many years.3,4 Brachioradial pruritus is a localized pruritus in the skin on the dorsolateral aspect of the arm or the skin over the brachioradialis muscle.5 We report here a case of brachioradial pruritus presenting with intractable pruritus in a young man.

Case report

A 22-year-old man presented with severe pruritus on the bilateral arms, shoulders and upper back for 3 weeks. He reported insidious onset of progressive numbness from the distal to proximal part in four limbs 1 month ago, which was followed by itching and painful sensations 1 week later. There were no skin lesions such as vesicles on the bilateral shoulders, upper chest, nape and upper back. He also had progressive muscle weakness in both hands 2 weeks prior to admission. The patient had difficulty grasping and using chopsticks. He then visited our emergency department and consulted a dermatologist.
Physical examinations revealed multiple excoriated erythematous macules with depigmented scars in a linear distribution over the dermatome from C4 to C6 (Figures 1 and 2). Neurological examination showed a decreased pin prick test in these areas. Decreased muscle power was found in the hypothenar muscles (4/5), deltoid muscles (3/5), biceps muscles (4/5), brachial muscles (4/5), brachioradial muscles (4/5), and triceps muscles (4/5). The laboratory data revealed that the white blood cell count was $11.6 \times 10^9/L$ (normal range: $3.9 \times 10^9–10.6 \times 10^9/L$), segment was $0.844\%$ (normal range: $0.42–0.74$), alanine aminotransferase levels were $43 \text{ U/L} \ (0.72 \mu \text{kat/L}; \text{normal range: } 0–36 \text{ U/L} \ [0–0.60 \mu \text{kat/L}]),$ alkaline-phosphatase levels were $81 \text{ U/L} \ (\text{normal range: } 28–94 \text{ U/L}),$ total bilirubin was $1.2 \text{ mg/dL} \ (\text{normal range: } 0–1.3 \text{ mg/dL}),$ direct bilirubin was $0.4 \text{ mg/dL} \ (\text{normal range: } 0–0.4 \text{ mg/dL}),$ blood urea nitrogen levels were $9 \text{ mg/dL} \ (3.21 \mu \text{mol/L}; \text{normal range: } 6–21 \text{ mg/dL} \ [2.1–7.5 \mu \text{mol/L}]),$ creatinine was $0.9 \text{ mg/dL} \ (80 \mu \text{mol/L}; \text{normal range: } 0.64–1.27 \text{ mg/dL} \ [57–112 \mu \text{mol/L}]),$ thyroid-stimulating hormone levels were $0.435 \text{ mIU/L} \ (\text{normal range: } 0.35–5.5 \text{ mIU/L}),$ free T4 levels were $1.59 \text{ ng/dL} \ (20.5 \text{ pmol/L}; \text{normal range: } 0.89–1.76 \text{ ng/dL} \ [11.5–22.7 \text{ pmol/L}]),$ T3 levels were $164.41 \text{ ng/dL} \ (2.53 \text{ nmol/L}; \text{normal range: } 60–181 \text{ ng/dL} \ [0.92–2.79 \text{ nmol/L}]),$ varicella zoster virus-IgG was positive, varicella zoster virus-IgM was negative, herpes simplex virus-IgM was negative, and rapid plasma regain was negative.

Neuropathic itch was highly suspected since the common secondary causes of intractable pruritus were ruled out and there were neurological signs on the specific dermatome (C4–C6 dermatomes). An image survey of the cervical spine was strongly suggested by a dermatologist. Surprisingly, a magnetic resonance image (MRI) disclosed enlargement of the cervical spinal cord from levels C2 to C5 with an intramedullary lesion, occupying more than two-thirds of the spinal cord, with T1 hypointensity and T2 hyperintensity and enhancement of T2 peripheral portion (Figure 3). A diagnosis of transverse myelitis with the clinical presentations of brachioradial pruritus was made. The patient received desloratadine and dexchlorpheniramine maleate but there was no significant improvement of his intractable itch.

In our case, a cerebrospinal fluid study showed pleocytosis: the white blood cell count was $5/\mu\text{L} \ (\text{normal range: } 0),$ red blood cell count was $1\mu\text{L} \ (\text{normal range: } 0)$ and sugar, protein and lactate levels were normal. Transverse myelitis caused by viral infection such as herpes simplex virus, varicella-zoster virus, Epstein-Barr virus or cytomegalovirus infections was considered the most likely etiology by the neurologist. Empirical systemic acyclovir ($500 \text{ mg every 8 hours}$) was given for 7 days. The muscle power of both upper limbs recovered after 2 weeks. Meanwhile, the itchy, stinging and burning sensations on the arms, nape and upper back also subsided. The patient was followed up for 3 months and there was no recurrence of similar symptoms.
Brachioradial pruritus is characterized by a persistent painful, burning or stinging itch on the skin over the dorsolateral aspect of the arms and occasionally on the shoulder, neck and upper back. It is also known as a recurrent solar dermatomopathy or solar pruritus. The itchy sensation is delivered by unmyelinated C or Aδ fibers that convey messages centrally via the spinothalamic tract. Brachioradial pruritus is a type of neuropathic itch caused by a disturbance in the circuit along the sensory afferent pathway.

The most commonly involved location in brachioradial pruritus is the skin over the dorsolateral aspect of the arms and occasionally on the shoulder, neck and upper back. It mainly affects those aged 39–70 years and presents in predominantly females. In addition, unilateral involvement of brachioradial pruritus is more often observed than bilateral.

Table 1: Comparison of brachioradial pruritus and notalgia paresthetica.

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Brachioradial pruritus</th>
<th>Notalgia paresthetica</th>
</tr>
</thead>
<tbody>
<tr>
<td>Locations</td>
<td>Cervical spondyloarthropathy (C3–C7), ± solar exposure, UV radiation</td>
<td>Nerve root compression of posterior rami of T2–T6 nerve root</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Painful, burning or stinging itch</td>
<td>Burning pain, paresthesia, hyperesthesia and pruritus</td>
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<tr>
<td>Dermatomes</td>
<td>C3–C7</td>
<td>T2–T6</td>
</tr>
<tr>
<td>Skin manifestation</td>
<td>Unremarkable</td>
<td>Well-demarcated hyperpigmented patch</td>
</tr>
<tr>
<td>Type of itch</td>
<td>Neuropathic</td>
<td>Neuropathic</td>
</tr>
<tr>
<td>Treatment</td>
<td>Topical capsaicin, oral gabapentin, carbamazepine, lamotrigine and surgical release for tumor compression</td>
<td>Topical capsaicin, EMLA cream, oral gabapentin and paravertebral local anesthetic block</td>
</tr>
</tbody>
</table>

EMLA = eutectic mixture of local anesthetics.

Discussion

Pruritus of unknown origin is a common complaint and systemic causes should be considered, such as kidney disease, hepatobiliary disease, thyroid disease, diabetes mellitus, chronic infection (bacterial, fungal or parasitic), human immunodeficiency virus infection, medication use and underlining malignancy. In a retrospective study of pruritus of unknown origin in 50 patients, 7 patients had a systemic disease. However, when there is localized pruritus without skin manifestations, brachioradial pruritus, notalgia paresthetica, anogenital pruritus and burning mouth syndrome should be taken into consideration.

Brachioradial pruritus is characterized by a persistent painful, burning or stinging itch on the skin over the brachioradials muscles. It is also known as a recurrent solar dermatomopathy or solar pruritus. The itchy sensation is delivered by unmyelinated C or Aδ fibers that convey messages centrally via the spinothalamic tract. Brachioradial pruritus is a type of neuropathic itch caused by a disturbance in the circuit along the sensory afferent pathway.

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Brachioradial pruritus is believed to be associated with previous UV light radiation and solar exposure. Photodamaged nociceptors in the skin can fire toward the central nervous system and the signal of pruritus may be augmented by nerve compression induced by cervical spine diseases. Most patients (86%) have outdoor jobs or activities and this disease typically occurs in the spring and disappears by the end of summer. In 1983, Heyl first described the theory that brachioradial pruritus might be due to nerve injury or nerve compression from cervical spondyloarthropathy. Recently, familial cases with the pattern of X-linked recessive inheritance were also observed.

However, the exact cause of brachioradial pruritus is controversial. Goodkin et al showed that the location of abnormalities in cervical radiographic examinations was the most common in the C3–C7 positions. Thus, it is important for clinicians to carry out roentgenography or MRI of the cervical spine.

In addition, because of persistent scratching, secondary skin lesions such as prurigo nodularis or lichenification can be found. The “ice-pack sign” is a hallmark of brachioradial pruritus. Application of ice on pruritic skin can dramatically relieve the symptoms.

Another similar disease of localized pruritus on the interscapular area over the back is notalgia paresthetica (Table 1). This condition is characterized by burning pain, paresthesia, hyperesthesia and severe itching accompanied by a well-demarcated hyperpigmented patch. The itchy sensation is over the middle to upper back in T2 through T6 distribution of the dermatome.

There was no satisfactory outcome of treatment before the pathogenesis of brachioradial pruritus was fully elucidated. Topical capsaicin, a substance P antagonist, was thought to be an effective therapy (10/13 patients) compared with placebo arms after 3 weeks of application in a randomized control study. Gabapentin, a structural analogue of γ-amino butyric acid and an anti-epileptic drug is also effective within a few days with a dosage of 600–1800 mg daily. Other treatment options including carbamazepine, anti-inflammatory drugs, cervical spine manipulation, neck traction, physiological therapy, surgical decompression of radiculopathy and avoidance of sun exposure have also been reported. However, oral antihistamines and corticosteroids are ineffective.
Transverse myelitis is a neurologic syndrome caused by inflammation across both sides of the spinal cord. It is characterized by limb weakness and sensory disturbance below the involved level. Occasionally, bowel dysfunction and urinary retention are also noted. Common etiologies include infection (viral, bacterial or fungal), connective tissue disorders, vasculopathy, malignancy. Also, the condition is sometimes idiopathic. In our case, muscle weakness of both upper extremities suggested the diagnosis of transverse myelitis. Localized itching was not considered a feature in transverse myelitis until Bond and Keough described the first case of transverse myelitis causing localized pruritus on both proximal upper arms in 2003. The pruritus was still not alleviated by oral antihistamines and corticosteroids but was responsive to oxycodone. The exact mechanism is still unclear.

In conclusion, brachioradial pruritus can present with minimal or an absence of cutaneous manifestations, which makes its diagnosis very difficult. Moreover, it is important to note the differences between brachioradial pruritus and notalgia paresthetica. For clinicians, it is important to be aware that the disease is refractory to oral antihistamines and corticosteroids. Once the diagnosis of brachioradial pruritus is suspected, cervical roentgenography or MRI should be carried out.

References