

Erythromelalgia: a Red Neuralgia

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Acta Neurol Taiwan 2016;25:49-50



Figure 1. Marked erythema of left sole (A) compared with baseline skin color (B).

A 35-year-old woman presented with acute onset of left foot numbness on awakening in the morning two days ago, followed by burning pain at night before sleep. The pain could be relieved by taking analgesics, but the numbness persisted. She had similar attack over the distribution of left ulnar nerve half a year ago. The presentation was acute onset numbness of the ulnar two fingers and weakness of left hand. The patient reported mild local heat but no erythema was noted. Compressive neuropathy was impressed after nerve conduction study (NCS), but the recovery was poor. She denied exposure to chemicals or toxins. Family history of systemic lupus erythematosus (SLE) was noted. Physical examination showed erythema and elevated skin temperature of left sole (Figure 1A) and mild swelling of left foot. The patient was not aware of the skin changes and denied feelings such as local heat or swelling. Neurological examination was unremarkable except left claw hand deformity and weakness. NCS was done several hours later, and the skin changes disappeared before the study. The study confirmed the diagnosis of left ulnar neuropathy with low compound motor action potential (CMAP). It also revealed a borderline low CMAP of left tibial nerve while the sensory studies were normal. Under the impression of erythromelalgia with family history of SLE, laboratory tests including the hemogram, serum vitamin B12 and folic acid levels, thyroid function, erythrocyte sedimentation rate (ESR) and autoimmune markers were checked, and the results were

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Received July 20, 2015. Revised November 16, 2015

Accepted December 28, 2015.

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within normal limits. Sexually transmitted diseases such as human immunodeficiency virus (HIV) infection and syphilis were also excluded. We repeated NCS 2 weeks later which showed low CMAP amplitude of left tibial nerve (2.20 mV, in contrast to 5.23 mV in the first study). After 3 months, we contacted the patient via telephone and she reported residual numbness and weakness of left foot.

Erythromelalgia, also known as Mitchell's disease, is a rare neurovascular peripheral pain disorder. The patients often suffer from intermittent heat, redness, swelling and pain more commonly affecting the lower extremities that can be exacerbated by warming, exercise and dependence on legs, and relieved by cooling and elevation⁽¹⁾. Primary erythromelalgia is caused by mutation of SCN9A gene, which encoding a voltage-gated sodium channel (NaV1.7) selectively expressed in dorsal root ganglion and sympathetic ganglion neurons⁽²⁾. The disorder could be either sporadic or familiar with autosomal dominant inheritance. Secondary causes include myeloproliferative disorder or paraneoplastic syndrome, medications such as cyclosporin and calcium channel blockers, infectious

diseases such as HIV, autoimmune vasculitis and mercury or mushroom poisoning⁽¹⁾. Because of the similar clinical presentations of the two episodes (acute onset of numbness and weakness, followed by incomplete recovery) and the recurrent nature of erythromelalgia, we speculated the first attack was also due to erythromelalgia. The skin changes might be ignored because of its transient presence, as in her second attack.

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