CASE REPORT

Successful Thoracoscopic Sympathectomy for Primary Erythromelalgia in the Upper Extremities

Erythromelalgia is known as a rare syndrome of unknown etiology, characterized by redness with burning pain, edema associated with increased skin temperature in the upper and/or lower extremities. Various treatments such as drug therapies and sympathetic blockade were reported. We report two cases including a 57-year-old woman and a 64-year-old woman, showing the successful clinical outcome by bilateral thoracoscopic sympathectomy. (Jpn J Thorac Cardiovasc Surg 2004, 52: 524–526)

Key words: primary erythromelalgia, sympathectomy, thoracoscopic surgery

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Cases

Case 1. A 57-year-old woman was referred to the Nippon Medical School Hospital because the patient had been suffering from redness in the periphery of fingers on both hands since 1989. She had developed a burning pain and a swelling particularly in the night time associated with increased skin temperature since 1996. Those symptoms improved by dipping her hand in cool water. The patient visited the Department of Dermatology and was diagnosed with primary erythromelalgia. Attacks of burning pain tended to occur at night and lasted until next morning. She had neither family history nor factors for erythromelalgia in her job or everyday environment. The laboratory examination at the first visit to the Nippon Medical School, exhibited no abnormal findings of hypertension, diabetes mellitus, rheumatoid arthritis, gout nor systemic lupus erythematosis, but hepatitis with slight elevation of serum glutamic oxaloacetic transaminase (GOT) and glutamic pyruvic transaminase (GPT). Thermographs revealed marked hyperthermia of the palmer region comparing with other body surface (Fig. 1). Subsequently a punch skin biopsy from an affected area exhibited thickening of the capillary blood vessel walls in the superficial epidermis with blood congestion, histopathologically. She was administered aspirin from a dermatologist but symptoms were failed to improve. Then, she was referred to anesthesiologist in order to undergo thoracic epidural blockade and satellite ganglion blockade at the Th2 and Th3 levels. Even though duration of pain relief was short term (a few days), thoracic epidural blockade was effective to reduce her burning pain. The patient was referred to us to gets permanent pain-relief by sympathetic blockade using thoracoscopic sympathectomy.

Under general anesthesia using one lung ventilation, the patient was placed in the supine position with head-side up. Thoracoscopic surgery was carried out using two puncture techniques, bilaterally. The first port was made at the fifth intercostal space and the second port was made at the third intercostal space of the anterior axillary line. Sympathetic nerve was cut 5 mm in length at the level of the second and third ribs, bilaterally. A catheter, 1.0 mm in diameter, was placed in the pleural cavity for the purpose of releasing free air, if evident on chest X-ray film after surgery. The patient was relieved from every symptom immediately after surgery and showed uneventful postoperative course. The patient is enjoying her life without suffering for six years after surgery.

Case 2. A 64-year-old woman had been suffering from pain in the periphery of lower extremities, particu-

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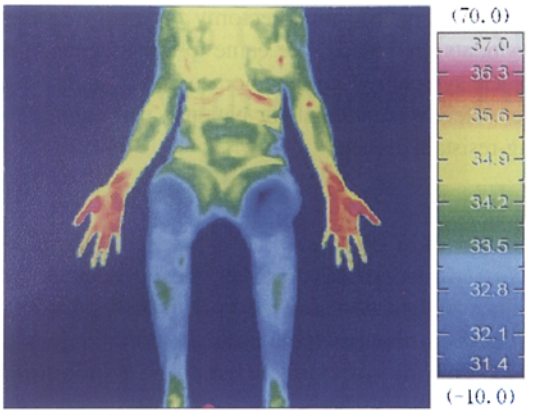

Fig. 1. Thermographs revealed marked hyperthermia of the palmer region comparing of other body surface in Case 1.

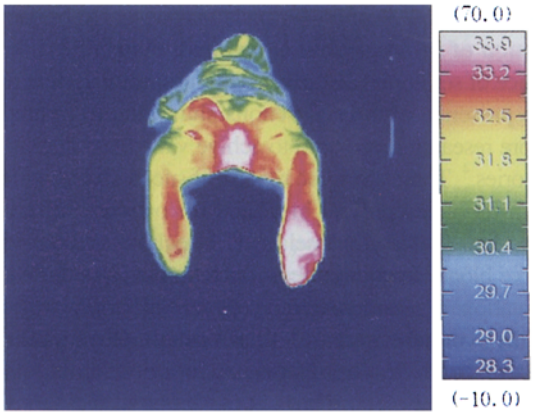

Fig. 2. Thermographs of the Case 2 revealed marked hyperthermia in the periphery of lower extremities, particularly on foot.

larly on foot since 1996, which resolved spontaneously. In 1999, a recurrence of the symptoms impaired her to walk because of severe pain. In addition, she suffered from left palmer pain and was diagnosed with primary erythromelalgia at other hospital, resulting in medication therapy with aspirin, anti-depressive agent and some tranquilizer. The fact that her symptoms failed to improve, plus bed rest was required prompted her to seek thoracoscopic sympathectomy at Nippon Medical School in 2001. The patient had no family history of relevant conditions. Palmer pain was reduced by dipping her hand in cool water. Thermographs of the patient 2 revealed marked hyperthermia in the periphery of lower extremities, particularly on foot (Fig. 2). After reconfirmation of effect by thoracic epidural blockade and satellites ganglion blockade, a thoracoscopic sympathetic blockade of the left sympathetic nerve was carried out at the level of the 2nd and 3rd ribs in February, 2001. Sympathetic blockade was performed with coagulation. Plus, lumbar sympathetic nerve blockade using alcohol was performed as a permanent nerve blockade. After surgery symptoms disappeared, the patient could engage in rehabilitation without pain. However, six months later, she started to complain of cold feeling on her foot. Furthermore her increased sweating around the left axillary region suggested either loss of efficacy by thoracic sympathectomy or compensatory sweating by sympathetic blockade. In December 2001, because of recurrent pain, thoracoscopic sympathectomy and lumbar sympathetic nerve blockade using alcohol were carried out again. Even though there was no regeneration of sympathetic nerve macroscpically under thoracoscopic observation, additional coagulation using electrocautery was performed at the level of the 2nd and 3rd ribs and an additional sympathectomy was done at level of the 4th rib. After surgery, pain remarkably reduced and she showed uneventful postoperative course.

Discussion

Erythromelalgia reported by Mitchell et al.[1] in 1878 is a rare syndrome consisting of burning pain, redness and elevation of skin temperature, and to date its etiology remains unknown.[2] Erysthromelalgia clinically tends to be confused with other disorders, such as complex regional pain syndrome (CRPS; formerly defined as reflex sympathetic dystrophy or causalgia). Symptoms of CRPS failed to improve even after cooling down of affected areas. In contrast, the patients presented were relieved of severe burning pain by dipping affected areas into ice water, suggesting the presence of erythromelalgia but other disorder above mentioned. Erysthromelalgia has been classified into primary and secondary according to the presence of basic pathological conditions.[3] As for patients showing the secondary type of erythromelalgia, they frequently suffer from proliferated myeloblastic disease and the remaining conditions consist of hypertension, diabetes mellitus and systemic lupus erythematosis. Regarding drug therapy, aspirin was reported to be effective. Even though nonsteroidal antiinflammatory drugs (NSAIDs), anti-serotonin derivative β-brokers were used, their effectiveness were reported to be not stable. The first patient was diagnosed with idiopathic erythromelalgia, which was because the patient was not suffering from any concomitant disease except for chronic hepatitis. The second patient was diagnosed as having idiopathic erythromelalgia based on the absence of concomitant

disease. Etiology of erythromelalgia appears to be acceleration of prostaglandin compound, metabolic disturbance of serotonin, disturbance of vasomotor system and the presence of arterial-venous shunt due to hypoxia. More research is required to achive vailed and definitive theories.[4] There are some reports of successful treatment with thoracic epidural blockade for patients suffering drug-resistant erythromelalgia.[5] Because sympathetic nerve blockade contribute to reduction of hyperhidrosis, dilatation of vessels, and management of sensory system of the sympathetic nerve, it has been employed to treat hyperhidrosis, Raynaud's disease, arteriosclerotic obliterans and causalgia. Davis et al.[6] reported that three of six patients who underwent thoracic epidural blockade for erythromelalgia showed the reduction of symptoms. As for efficacy of sympathetic nerve blockade in erythromelalgia, it may improve circulation, leading to the reduction of pooled blood in the peripheral capillaries, which resulted in the improvement of burning pain. In the treatment of erythromelalgia, either thoracic epidural blockade or satellite ganglion blockade were considered reliable preoperative evaluation to determine whether thoracic sympathectomy is adequate. While a recurrence rate of sympathetic nerve blockade for erythromelalgia needs more research, 0% for hyperhidrosis and 50 % for Raynaud's disease syndrome were reported by Kalgaard et al.[7] In the present cases, thoracic sympathectomy remarkably reduced patient complaints, however, the second case suggested the presence of regeneration of sympathetic nerve because of incomplete denervation by electrocautery. In our experience on thoracoscopic sympathectomy for palmer hyperhidrosis, two of fifty cases (4%) showed recurrence of symptoms. In Case 2, a coagulation technique by electrocautery was employed because of the presence of intrapleural adhesions. Therefore, sympathetic blockade was performed on the current cases with a cutting technique based on the prior experience. Thoracoscopic surgery offered minimally invasive thoracic sympathectomy for erythromelalgia in patients presented. We came to the conclusion that thoracic sympathectomy by thoracoscopic surgery could offer effective treatment for patients, particularly with drug-resistant disease.

Conclusion

We reported two successful cases suffering from erythromelalgia who achieved permanent reduction of symptoms by thoracoscopic sympathectomy.

REFERENCES

1. Mitchell SW. On a rare vaso-motor neurosis of the extremities, and on the maladies with which it may be confounded. Am J Med Sci 1878; 76: 17–36.
2. Smith LA, Allen EV. Erythermalgia (erythromelalgia) of the extremities: A syndrome characterized by redness, heart, and pain. Am Heart J 1938; 16: 175–88.
3. Catchpole BN. Erythromelalgia. Lancet 1964; 42: 909–11.
4. Landry GJ, Edwards JM, Porter JM. Current management of Raynaud's syndrome. Adv Surg 1996; 30: 333–47.
5. Shiga T, Sakamoto A, Koizumi K, Ogawa R. Endoscopic thoracic sympathectomy for primary erythromelalgia in the upper extremities. Anesth Analg 1999; 88: 865–6.
6. Davis MD, O'Fallon WM, Rogers RS 3rd, Rooke TW. Natural history of erythromelalgia: Presentation and outcome in 168 patients. Arch Dermatol 2000; 136: 330–6.
7. Kalgaard OM, Seem E, Kvernebo K. Erythromelalgia: A clinical study of 87 cases. J Intern Med 1997; 242: 191–7.