News Release

Title

Establishment of in-hospital clinical network for patients with neurofibromatosis type 1 in Nagoya University Hospital

Key Points

・We determined the minimal irradiance and dose of UV-LED needed to produce sufficient 25(OH)D in mice.

・ Low energy irradiation of narrow-range UV-LED was effective in improving osteoporotic and sarcopenic changes in the presence of vitamin D deficiency in a senescence-accelerated mouse model.

・ Low energy irradiation of narrow-range UV-LED did not induce severe skin damages.

Summary

Prof. Yoshihiro Nishida at Department of Rehabilitation Medicine, Dr. Kunihiro Ikuta at Department of Orthopaedic Surgery, and Prof. Norio Ozaki at Department of Psychiatry, Nagoya University Hospital, established in-hospital clinical network for patients with neurofibromatosis type 1 in Nagoya University Hospital, and reported the progress of this clinical network.

Neurofibromatosis type 1 (NF1) is a genetic multisystem disorder. Clinicians must be aware of the diverse clinical features of this disorder in order to provide optimal care for it. We have set up an NF1 in-hospital medical care network of specialists regardless of patient age, launching a multidisciplinary approach to the disease for the first time in Japan.

From January 2014 to December 2020, 246 patients were enrolled in the NF1 patient list and medical records. Mean age was 26.0 years ranging from 3 months to 80 years. The number distribution was higher as the first visit age was lower. There were 107 males (41%) and 139 females. After 2011, the number of patients has increased since the year when the medical care network was started. Regarding orthopedic signs, scoliosis was present in 60 cases (26%), and bone abnormalities in the upper arm, forearm, and tibia in 8 cases (3.5%). Neurofibromas other than cutaneous neurofibromas were present in 90 cases (39%), and MPNST in 17 cases (7.4%).

We launched a multidisciplinary NF1 clinic system for the first time in Japan. For patients with NF1, which is a hereditary and systemic disease associated with a high incidence of malignant tumors, it will be of great benefit when the number of such clinics in Japan and the rest of Asia is increased.

Research Background

Neurofibromatosis type-1 (NF1) is a hereditary disease that affects 1 in 2,500-3,000 people regardless of gender or ethnicity. There is a marked phenotypic variability and a unpredictable lifetime course. Complications in adulthood include central nervous system tumors, malignancy, cognitive deficits and vasculopathy. Plexiform neurofibromas affect up to 60% of individuals with NF1, can cause pain and disfigurement, and may undergo malignant change. Malignant peripheral nerve sheath tumor (MPNST) is the most common malignancy associated with NF1 with a lifetime risk of 5.9–15.8%. There is a decreased life expectancy of 8–20 years in NF1 patients.

Thus, for NF1 patients who have various symptoms and a high incidence of malignant tumors that have a great influence on the prognosis of life, it is inappropriate to treat them in a single department. In the United States, the Neurofibromatosis Clinic Network was established by the Children's Tumor Foundation in 2007 to standardize and raise the level of clinical care for neurofibromatosis nationwide. On the other hand, in Japan, there was no NF1 clinic that treated with multidisciplinary, so NF1 medical treatment was performed only by a specialist in a single department generally for one symptom. There were many problems such as delays in medical treatment for other symptoms in NF1 caused by medical treatment in a single department.

In order to solve these problems, we have built and promoted an in-hospital NF1 medical care network with multiple departments and occupations since 2014. The purpose of this study is to introduce the process of launching this NF1 medical care network and its progress.

Research Results

In January 2014, we started the NF1 in-hospital medical care network (Figure 1). For NF1 patients who had their first visit before that and had been continuously treated, we requested each department to provide medical care as an NF1 network.

From January 2014 to December 2020, 246 patients were enrolled in the NF1 patient list and medical records. Excluding the three patients whose age at the first visit was unknown, mean age was 26.0 years ranging from 3 months to 80 years. The younger the age at the first visit, the higher the patient number distribution (Figure 2). There were 107 males (41%) and 139 females. Looking at the number of first-time patients by age group by gender, the number of males in their 20s and 30s was markedly lower than that of females（Figure 3）. Divided by 5 years, the number of first-time NF1 patients is increasing markedly. Especially after 2011, the number of patients has increased since the year when the medical care network was started（Figure 4）。Excluding 15 patients whose family history is unknown, familial NF1 was 101 (44%), and non-familial was 130, with non-familial slightly predominant.

Regarding orthopedic signs, there was a description that was investigated in 230 patients. Scoliosis was present in 60 cases (26%), and bone abnormalities in the upper arm, forearm, and tibia were observed in 8 cases (3.5%). Neurofibromas other than cutaneous ones were found in 90 cases (39%) and MPNST in 17 cases (7.4%)（Figure 5）.

Research Summary and Future Perspective

For the first time in Japan, we established an NF1 medical care network with multiple departments and multiple occupations. By comprehensively treating NF1 patients through this network, it will be possible to accurately deal with various symptoms. In addition, if such NF1 clinics are established all over Japan, the quality of medical care will be improved, which will be a great advantage for NF1 patients and their families. It will also contribute to the registry of patients with neurofibromatosis type 1, which is a designated intractable disease, and to grasp the actual situation.

Publication

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