Small Fiber Neuropathy Underlies Erythromelalgia

BY JANE SALODOF MCNEIL
Southwest Bones

Park City, Utah — Small fiber neuropathy plays an important role in erythromelalgia, a rare and mysterious skin condition characterized by red, hot, and painful extremities. Dr. Mark D.P. Davis said at a clinical dermatology seminar sponsored by Medicis.

Thermoregulatory sweat testing and other neurologic evaluations of 32 erythromelalgia patients showed that people with the syndrome do not sweat in all or part of their bodies, he reported.

Dr. Davis, dermatology professor at the Mayo Clinic in Rochester, Minn., illustrated his talk with diagrams of various patterns of impaired sweat function, from the new study. These included regional and distal, multifocal, global, and distal areas where the patients did not sweat.

“Sweat glands are innervated by small nerves,” he explained. “If they are dysfunctional, you may not sweat normally.”

Recent papers documenting SCN9A mutations in an inherited form of erythromelalgia confirmed the neuropathic basis of the condition. “Sweat glands are innervated by small nerves,” Dr. Davis said. “If they are dysfunctional, you may not sweat normally.”

The prevalence of neuropathy and vasculopathy in erythromelalgia suggests that “in other flushing conditions it is conceivable that the same thing is going on,” he said. He cited lupus and brachioradial pruritus as infectious and itching skin disorders, respectively, that involve both nerves and skin.

The first description of erythromelalgia was published in 1878. Dr. Davis said. In 2000, he and his colleagues presented a natural history of the disorder based on 148 patients seen between 1970 and 1994 at the Mayo Clinic (Arch. Dermatol. 2000;136:330-6). None had other diagnoses to explain their symptoms. The patients’ average age was 56 years, with a wide range of 5-93 years and average follow-up of 9 years. The majority, 72%, were female.

Two-thirds of the patients presented with abnormalities in the affected limb. These included cyanosis (49%), acrocyanosis (10%), ulcers (6%), and reticular skin pattern (5%).

Half the patients were unable to walk long distances or stand for long periods. About 14% were unable to keep a job and 13% were unable to drive. Functional impairment was so debilitating that 3% required a wheelchair and 2% were bed-bound.

“Sweat glands are innervated by small nerves,” he explained. “If they are dysfunctional, you may not sweat normally.”

Recent papers documenting SCN9A mutations in an inherited form of erythromelalgia confirmed the neuropathic basis of the condition. “Sweat glands are innervated by small nerves,” Dr. Davis said. “If they are dysfunctional, you may not sweat normally.”

The prevalence of neuropathy and vasculopathy in erythromelalgia suggests that “in other flushing conditions it is conceivable that the same thing is going on,” he said. He cited lupus and brachioradial pruritus as infectious and itching skin disorders, respectively, that involve both nerves and skin.

The first description of erythromelalgia was published in 1878. Dr. Davis said. In 2000, he and his colleagues presented a natural history of the disorder based on 148 patients seen between 1970 and 1994 at the Mayo Clinic (Arch. Dermatol. 2000;136:330-6). None had other diagnoses to explain their symptoms. The patients’ average age was 56 years, with a wide range of 5-93 years and average follow-up of 9 years. The majority, 72%, were female.

Two-thirds of the patients presented with abnormalities in the affected limb. These included cyanosis (49%), acrocyanosis (10%), ulcers (6%), and reticular skin pattern (5%).

Half the patients were unable to walk long distances or stand for long periods. About 14% were unable to keep a job and 13% were unable to drive. Functional impairment was so debilitating that 3% required a wheelchair and 2% were bed-bound.

“Sweat glands are innervated by small nerves,” he explained. “If they are dysfunctional, you may not sweat normally.”

Recent papers documenting SCN9A mutations in an inherited form of erythromelalgia confirmed the neuropathic basis of the condition. “Sweat glands are innervated by small nerves,” Dr. Davis said. “If they are dysfunctional, you may not sweat normally.”

The prevalence of neuropathy and vasculopathy in erythromelalgia suggests that “in other flushing conditions it is conceivable that the same thing is going on,” he said. He cited lupus and brachioradial pruritus as infectious and itching skin disorders, respectively, that involve both nerves and skin.

The first description of erythromelalgia was published in 1878. Dr. Davis said. In 2000, he and his colleagues presented a natural history of the disorder based on 148 patients seen between 1970 and 1994 at the Mayo Clinic (Arch. Dermatol. 2000;136:330-6). None had other diagnoses to explain their symptoms. The patients’ average age was 56 years, with a wide range of 5-93 years and average follow-up of 9 years. The majority, 72%, were female.

Two-thirds of the patients presented with abnormalities in the affected limb. These included cyanosis (49%), acrocyanosis (10%), ulcers (6%), and reticular skin pattern (5%).

Half the patients were unable to walk long distances or stand for long periods. About 14% were unable to keep a job and 13% were unable to drive. Functional impairment was so debilitating that 3% required a wheelchair and 2% were bed-bound.