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Original Article

The diagnosis and treatment of hereditary angioedema patients in Japan: A patient reported outcome survey

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C1-INH C1 inhibitor
HAE Hereditary angioedema
pdC1-INH plasma-derived C1 inhibitor concentrate

ABSTRACT

Background: The rate at which patients are accurately diagnosed with hereditary angioedema (HAE), as well as diagnosed patients access to modern treatments differs greatly among countries. Moreover, the severity and burden of HAE on patients have been reported mostly on the basis of physician-reported surveys. To gain insight into the real-world conditions of patients with HAE through a patient-reported survey in Japan and identify any unmet needs.
Methods: A questionnaire was distributed to 121 patients with HAE via a Japanese HAE patient organization during 2016–2017. Responses were collected from 70 patients (57.9%) and subjected to analysis.
Results: The average periods from the initial appearance of symptoms (e.g. edema) to a HAE diagnosis was 15.6 years (min–max, 0–53). Patients visited an average of 4.6 different departments until receiving a definitive diagnosis. The average age at the first visit was 25.6 years (3–73) and at diagnosis 32.8 years (0–73). Patients reported an average of 15.7 (0–100) attacks per year, but only 53.1% of attacks were treated. The days of hospitalization due to severe attacks was 14.3 (0–200) before diagnosis, but these declined to 4.3 (0–50) after diagnosis. In the treatment for attacks, 82% of the patients were treated with the plasma-derived C1 inhibitor concentrate, and 69% of the patients reported experiencing a therapeutic effect.
Conclusions: There is a long gap between first attack and diagnosis of HAE, and the number of non-treated attacks is high in Japan. Steps are needed to improve the diagnostic and treatment environments to address these issues.

Introduction

Hereditary angioedema (HAE) is a genetic disorder that results in the unregulated increase of bradykinin leading to edema. It is classified into HAE type I, HAE type II and HAE with normal C1-inhibitor (C1–INH) based on serum levels of C1–INH protein and function.[1] With a small but growing number of patients

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日本における遺伝性血管性浮腫の治療の現状および問題点について報告しています。

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