Description of Hypereosinophilic syndrome (HES) and subtypes in North America

Aims

Hypereosinophilic syndrome (HES) is a group of rare hematologic disorders in which eosinophils are overproduced for protracted periods of time resulting in organ damage. Different subtypes of HES have been identified but little is known about their similarities or differences. This review aims to describe clinical characteristics of HES subtypes based on reported cases in the literature.

Methods

A PubMed targeted literature search from January 2000 to March 2020 was conducted to identify publications, in English, of the following subtypes: idiopathic (HES), myeloproliferative (HIES), lymphoid (L-HES) and chronic eosinophilic leukemia, not otherwise specified (CEL-NOS). Information from 170 publications with individual case data worldwide was reviewed. Among them, 79 individuals (11% HES, 25 HIES, 21 L-HES, 1 CEL-NOS) were reported in North America (Canada and USA) and have been summarized here.

Results

From the selected articles (Figure 1) the following information was collected:

- Author, country and year of publication
- Age and gender, HES subtype and blood eosinophil count at diagnosis
- Organ affected, symptoms/diagnosis of presentation
- Treatment, maintenance therapy, follow-up time and mortality

Table 1. Main treatment reported

Conclusions

Differences in clinical manifestations and treatment between subtypes were observed for optimal management of HES patients. It is important to differentiate these subtypes and provide an early treatment in an effort to limit the organ damage observed.

Table 2. Five most commonly reported signs and/or symptoms*

*Signs and/or symptoms are presented as reported in the articles, and categories are not mutually exclusive.

Table 3. Main organ involvement by HES subtype

* Included for data extraction (n=231 publications)

Figure 1. PRISMA flow chart

Figure 2. Organ involvement by HES subtype

Table 1. Age, gender and eosinophil counts at diagnosis by HES subtype

HES subtype | Age, years | median (range) | Male | Female | Absolute eosinophil count | Percentage
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HES, NOS (N=21) | 46.6 (18-66) | 20 (04) | 26 (61) | 6700 (360-17600) | 19% | 26%
HIES (N=6) | 46.7 (17-47) | 21 (08) | 5 (41) | 30 460 (6 250-21 600) | 26% | 33%
L-HES (N=10) | 46.3 (16- 79) | 10 (0) | 6 (0) | 610 (6 200-52 600) | 38% | 55%
CEL-NOS (N=1) | 46 (11) | 1 (00) | 0 (0) | 6 (200) | 55% | 55%

* Normal white blood cell count: 4 500–10 000 cells/µL. Normal differential blood cell count: eosinophils 0%–5% of total white blood cells. 5% threshold for HES diagnosis.

* Time interval from treatment to first tumor event in months.

* Signs and/or symptoms proportioned as reported in the articles, and categories are not mutually exclusive.

* Provided by the author. The abstract was not available in the published article.

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