Research letter

Population-based prevalence of eosinophilic fasciitis (Shulman syndrome): a capture–recapture study

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Dear Editor, Our knowledge of eosinophilic fasciitis (EF), also known as Shulman syndrome, is limited and its prevalence has not been estimated so far. We conducted a regional survey with the aim of estimating the prevalence of EF in Alsace, a region in the north-east of France.

We retrospectively collected details of individuals with EF from 1 January 1983 to 30 March 2015 among Alsace residents aged 18 years and older, then performed a capture–recapture analysis in 2015 with the prevalent cases.

Potential EF cases were identified through three separate sources: (i) the French hospital discharge database (Programme de Médicalisation des Systèmes d'Information), which provides medical information about all patients discharged from all six public hospitals in the Alsace region using the ICD-10 code M35.4 ('disseminated fasciitis with eosinophilia'); (ii) regional experts involved in the care of patients with autoimmune diseases (using a computerized search of the medical records of patients treated in their departments: internal medicine, n = 21; rheumatology, n = 12; dermatology, n = 7; and haematology, n = 8, from public hospitals in Alsace); (iii) computerized case databases from pathology departments (n = 4) in the Alsace region, (the paper archives of these departments were checked manually when available). All living patients were contacted to obtain informed consent. The study was approved by the ethics review board of Strasbourg's school of medicine and by Commission Nationale Informatique et Libertés.

As there are no validated diagnosis criteria for EF, two authors (L.S. and L.A.) experienced in the diagnosis and management of EF independently reviewed the medical files of all patients with potential EF and validated the diagnosis by consensus, using a combination of clinical, pathological, imaging and biological findings in addition to disease course, with the exclusion of differential diagnoses.

Capture–recapture analysis was used to estimate the total number of missing cases for the year 2015. Dependencies between sources were assessed using the method of Wittes *et al.*[1] The number of cases was estimated using the Chao log-linear model for a closed population.[2] Statistical analyses were performed using R version 3.1.3 package (R Foundation, Vienna, Austria).[3] The prevalence of EF and its 95% confidence interval (CI) was calculated using the EpiTools epidemiological calculators (http://epitools.ausvet.com.au).

Using the three sources, we identified 30 potential EF cases. Following a thorough review of these patients' cases, we validated by consensus 19 EF cases according to compatible clinical, pathological, imaging and biological findings, in addition to disease course. Sixteen of these patients fulfilled the Pinal-Fernandez criteria;[4] the three remaining patients had one major criterion of this classification, but only one minor. The cases of 11 patients were excluded because of differential diagnoses, living in a different region or country (n = 4), refusal to sign the consent form (n = 1), death before study date (n = 2) and missing data (n = 4). Of the validated 19 patients, 13 were identified using the expert databases, 11 using the Programme de Médicalisation des Systèmes d'Information and four using the pathology data. Thirteen of the cases were identified from a single source, three from two sources and three from all three sources, as indicated in Figure 1.

Alsace region had an estimated population of 1·880 million in January 2015 (https://www.insee.fr/fr/statistiques/1893198). We estimated the total number of cases of EF in the Alsace region to be 27 (95% CI 20–63), yielding a 2015 population-based prevalence of 14 (95% CI 10–21) per million inhabitants for EF in Alsace.

Alsace offers ideal conditions for epidemiological studies of rare diseases, as patients are unlikely to seek care outside of the region because of its particular geography. Additionally, the population base is large, resulting in straightforward estimation of the 95% CI of the result.

Some biases, including the lack of standardized criteria for the diagnosis of EF and a retrospective design, may have had an impact on the results of the present study. However, our study has several strengths. The physicians involved had significant knowledge of the disease and are members of national and regional public accredited centres for rare and autoimmune diseases. All EF diagnoses were verified independently by two experts. We also included magnetic resonance imaging results as part of the EF diagnostic strategy.

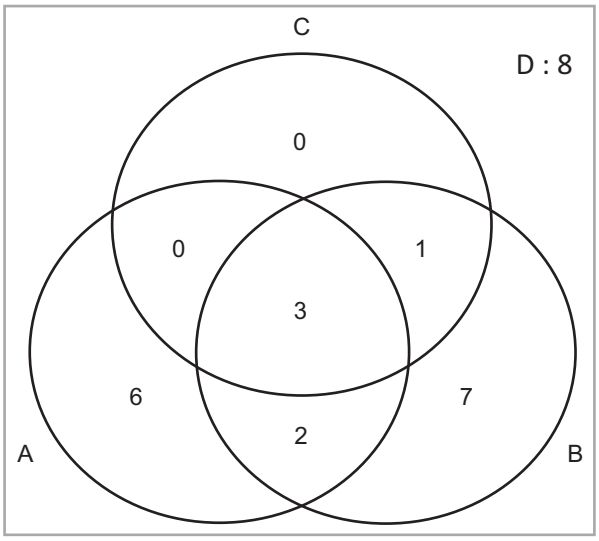

Fig 1. Number of patients with eosinophilic fasciitis ascertained from each of the three sources evaluated and among-source overlaps after exclusions. A: Programme de Médicalisation des Systèmes d'Information; B: expert-identified cases; C: pathology data; D: estimated missed cases.

Reliable diagnosis criteria based on the most recent and noninvasive diagnostic tools should be defined and validated to help clinicians standardize the diagnosis and management of this very rare disease.

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