CLINICAL PRESENTATION OF AUTOINFLAMMATORY DISEASES IN CHILDREN, ADOLESCENTS AND YOUNG ADULTS: A LATIN AMERICAN MULTICENTER STUDY

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BACKGROUND

Multicenter studies of autoinflammatory diseases (AID) are scarce in Latin American population. Given the importance of different disease prevalence and phenotypes worldwide, particularly between children and adolescents/young adults (AYA), we aimed to compare accrued signs and symptoms, laboratorial findings and treatments in AID patients grouped by age at diagnosis of disease and sex.

METHODS

This is an observational, descriptive study of a Latin American (LA) registry through a designed web system for data storage, collected between 2015 and 2018. The case-report forms were translated into Portuguese and Spanish languages, including demographic, clinical, laboratorial, genetic features and treatment.

RESULTS

We included 152 patients, being 51.3% male and 75% Caucasians. The median disease onset age was 2.1 years (0–15.6 years), median diagnosis age 6.9 years (0–21.9 years); 111 (73%) were children (0–9 years) and 41 (27%) AYA (10–21 years). Periodic fever, aphthous stomatitis, pharyngitis, and adenitis syndrome (PFAPA) was the most frequent diagnosis, followed by chronic non-bacterial osteomyelitis (CNO) and familial Mediterranean fever (FMF). Periodic fever, aphthous stomatitis, pharyngitis, and adenitis syndrome was seen more often in young children, and CNO in older patients, with female predominance. The FMF prevalence was higher in boys. Further analysis comparing children and AYA revealed that some clinical features, such as fever, cervical adenitis, aphthous stomatitis, pharyngitis and lymphadenopathy, were more frequent in children; and arthralgia, arthritis, bone pain, osteomyelitis, osteitis and aseptic arthritis were more frequent in AYA.

CONCLUSION

Differences related to accrued signs and symptoms, laboratorial findings and treatments in AID patients were observed between the LA population grouped by age at diagnosis of disease and sex. Children had predominantly PFAPA manifestations at presentation; whilst AYA had more musculoskeletal involvement.

KEYWORDS

Autoinflammatory disease, Children, Adolescents, Periodic fever, Familial Mediterranean fever.