REGULAR ARTICLE

blood advances

Incidence of acquired pure red cell aplasia: a nationwide epidemiologic analysis with 2 registry databases in Japan

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Key Points

- A nationwide epidemiologic study for acquired PRCA identified 1055 patients, an incidence rate of 1.06 patients per million per year.
- The median age was 73 years old with female predominance (1.5:1), and 69% of the PRCA was idiopathic.

Acquired pure red cell aplasia (PRCA) is a rare syndrome characterized by anemia with reticulocytopenia and a marked reduction in erythroid precursors. Given its rarity, the true incidence is largely unknown, and epidemiological data representing the general population, with a description of the full spectrum of etiologies, are scarce. An epidemiological study on PRCA in Japan conducted 30 years ago estimated the annual incidence as 0.3 per million. To update the data and investigate the incidence and demographics of PRCA, we conducted a nationwide epidemiological study using the Japanese Society of Hematology (JSH) Hematologic Disease Registry, a hematologic disease registration database managed by the JSH and the Diagnosis Procedure Combination (DPC) study data available at a website of the Ministry of Health, Labor, and Welfare (MHLW) of Japan. A total of 1055 patients with newly diagnosed acquired PRCA were identified between 2012 and 2019, and the average annual incidence was calculated at 1.06 (95% confidence interval [CI], 0.83-1.28) per million. The median age was 73 (range, 18-99) years. The female-to-male ratio was 1.5:1, and the female predominance was most prominent in the child-bearing age group. Sixty-nine percent of acquired PRCA was idiopathic. The incidence of PRCA was approximately 20% of that of aplastic anemia (AA) during the same period. Approximately 0.98 patients per million per year (95% CI, 0.89-1.07) required hospitalization for the treatment of PRCA. These results are expected to contribute to the discussion of resource allocation for PRCA in the aging population in many countries, including Japan.

Introduction

Acquired pure red cell aplasia (PRCA) has been defined as a bone marrow disorder characterized by anemia, reticulocytopenia, and erythroid hypoplasia.[1,2] It presents as cytopenia with a single lineage but shares some clinical features with other bone marrow failure syndromes and autoimmune cytopenias. Nosologically, PRCA is classified either into a congenital entity, such as Diamond-Blackfan anemia, or acquired PRCA. The latter may be associated with various underlying medical conditions and diseases,

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The full-text version of this article contains a data supplement.