Study finds Hong Kong acquired hemophilia A patients are double that of Caucasian populations

June 7 2024

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A research team led by Dr. Albert Sin Chun-fung from the Department of Pathology, School of Clinical Medicine, LKS Faculty of Medicine of the University of Hong Kong (HKUMed) has achieved a significant

breakthrough in characterizing territory-wide cohorts of acquired hemophilia A (AHA) patients in Hong Kong and identifying their prognostic factors. This discovery provides valuable insights for improving survival rates among patients with this rare but fatal blood disorder.

The analysis revealed that the annual incidence of AHA cases in Hong Kong is twice as high as that observed in Caucasian populations. There is a higher mortality rate in the older demographic in local AHA patients, mainly because of severe infections resulting from immunosuppressive therapy. The study was published in *Thrombosis Research*.

Acquired hemophilia A (AHA) is a rare bleeding disorder caused by the production of autoantibodies that destroy factor VIII, an important coagulation factor for blood clotting. Patients with AHA experience severe bleeding and organ damage. The incidence of AHA in Caucasian cohorts is about one per million per year. Its rarity and rapid onset of catastrophic bleeding make diagnosis particularly difficult.

While most studies have been conducted on Western populations, there is a lack of research on East Asian patients. Given the inherent differences in the hemostatic systems between Caucasians and East Asian patients, the clinical characteristics of AHA in East Asian patients are different from those in Caucasians. Therefore, there is an urgent need to conduct a comprehensive study on this disorder in the East Asian patient population.

The research team analyzed data from the medical records of 165 AHA patients diagnosed between 2012 and 2021 in local public hospitals. This comprehensive analysis encompassed demographic information, medical history, drug records and outcomes.

Researchers estimated that the incidence of AHA among the local

population is 2.4 cases per million per year, more than double the rate reported for Caucasian populations. The analysis revealed that Hong Kong AHA patients are predominantly elderly with multiple medical comorbidities.

The median age of diagnosis is 80, which is older than the patient populations reported in previous studies. The mortality rate is 55.2% compared to 25% in Caucasian cohorts. About half (49.5%) of these deaths were caused by severe infections due to immunosuppressive therapy required to manage the condition.

The team found that diagnosis is often delayed, with a median of seven days taken to establish the condition. They found that age is the key determining factor in mortality. Advanced age at diagnosis is associated with a higher mortality rate, as well as a lower chance and longer time to achieve remission.

Understanding of AHA in East Asian patient populations is limited, as comprehensive studies involving cohorts of AHA for East Asian patients are lacking. AHA remains an under-recognized condition both among the general public and general medical practitioners.

This study revealed a higher incidence of AHA compared with Western regions and discovered unique characteristics of Chinese AHA patients. The patient group comprised mainly elderly patients with multiple medical comorbidities, presenting difficulties in managing the condition by immunosuppression, a crucial therapeutic strategy to treat AHA.

"This study highlights the unique epidemiological and clinical features of AHA in the local Chinese population. These are clearly vulnerable patients who require specialized, age-appropriate care," said Dr. Albert Sin Chun-fung, Clinical Assistant Professor, Department of Pathology, School of Clinical Medicine, HKUMed, who led the research.

"But the reality is that diagnosis is often delayed, and hospital stays are excessively long with a median length of 25 days, placing a heavy burden on our public health care system."

These findings highlight the urgency of raising awareness and providing education about AHA among the general public and general medical practitioners to ensure timely recognition and appropriate treatment of this condition in these patient groups.

Dr. Sin emphasized that the lack of awareness can lead to delayed diagnosis and suboptimal management of this potentially serious autoimmune disorder. Additional studies are also necessary to develop better treatment strategies that minimize the risk of immunosuppression and shorten hospital stays.

More information: Chun-fung Sin et al, Characteristics and outcome of a territory-wide cohort study of patients with acquired hemophilia A in Hong Kong, *Thrombosis Research* (2023). DOI: 10.1016/j.thromres.2023.11.025

Provided by The University of Hong Kong