Editorial

Hereditary angioedema-Perspective of Bangladesh

Hereditary angioedema (HAE) is a rare, potentially life-threatening genetic disorder characterized by recurrent, non-pruritic swelling of the skin, mucosa, and internal organs which is usually misdiagnosed as allergic reaction. It is due to C1-inhibitor deficiency or dysfunction and bradykinin-mediated vascular permeability. Although pathophysiology has been clarified and expanded diagnostic and therapeutic options, HAE remains unrecognized, frequently misdiagnosed, and undertreated in low- and middle-income settings.

The true prevalence of HAE in Bangladesh is unknown. According to global estimates (1:50,000–1:100,000)[1], the country is likely to harbor several hundred affected individuals. The case identification is obscured due to unawareness among primary care providers about non-histaminergic angioedema. The confirmatory laboratory testing (C4, C1-inhibitor level and function) is available only at certain centers of the country. On the other hand lack of family screening and genetic testing limits the subtyping of HAE[2].

HAE patients are usually treated with antihistamines and corticosteroids, which actually do no remedy for the patients. This approach is mostly due to unawareness of the disease by the primary health care provider and sometimes specialist doctors also.

The first line treatment is targeted acute therapies (plasma-derived or recombinant C1-inhibitor concentrate, bradykinin B2 receptor antagonist icatibant, kallikrein inhibitors) are largely unavailable. Severe attacks can be managed by Fresh frozen plasma, but again it requires special settings[3].

Prophylactic options include long-term C1-INH, lanadelumab, or oral kallikrein inhibitors. But these are also out of reach for most patients. Lastly androgenic anabolic agents are used despite adverse effects and monitoring needs.

To tackle any emergency situation, any emergency preparedness like individualized action plans, carry-on medication for acute attacks is not in practice[4].

HAE should be included in the national rare disease registries. Development of laboratory services for C4 and C1-INH assays in regional centers, plus referral networks is necessary. Access to medicine should be ensured and life-saving agents should be made available at public hospitals. National clinical guidelines and patient action plan templates should be developed and disseminated for awareness build up and training the primary health care providers.

Patient support group should be established and encouraged to go for family screening to identify affected relatives. A national HAE registry can be created to document epidemiology, outcomes, promote local research partnerships to evaluate cost-effective diagnostic and therapeutic models applicable for Bangladesh.

Finally, recurrent angioedema without urticaria, recurrent abdominal pain with unexplained swelling, or a family history of angioedema should go for HAE screening. Emergency departments should be equipped with proper airway management strategies and available therapies for management of HAE.

Reduce diagnostic delay, escape the avoidable deaths by ensuring proper emergency management at the time of acute attack, and improving quality of life for people with HAE-all these should be ensured by proper use of available limited resources in Bangladesh.

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