Gleich syndrome: what we know so far

Maria Moroianu-Brînzei[1], Cătălina Cernat[1], Rama Boustani[1], Mihaela-Ruxandra Udrea[1], Otilia-Luminița Manolache[1], Selda Ali[1,2], Roxana Silvia Bumbăcea[1,2]

1.Spitalul Clinic de Nefrologie 'Dr. Carol Davila' București;

2. Universitatea de Medicină și Farmacie 'Carol Davila' București

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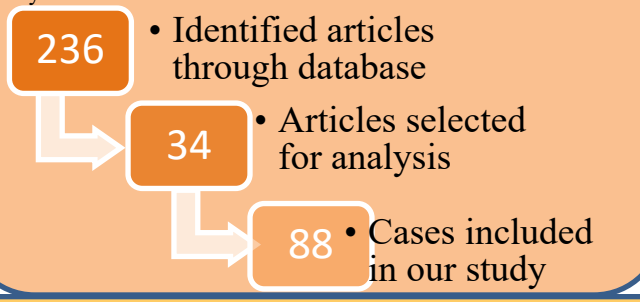
Conferința Națională DE ALERGOLOGIE ȘI IMUNOLOGIE CLINICĂ
2-4 iunie 2022 | Hotel Crowne Plaza, București

Background: Gleich's syndrome, also known as episodic angioedema with hypereosinophilia, is a rare disease characterized by recurrent episodes of angioedema, urticaria and eosinophilia. It may also be accompanied by fever and weight gain. Symptomatology appears at regular time intervals and the evolution varies from spontaneous resolution to requirement of biologics.

Methods: We conducted a systematic search of the literature in an online medical database (PubMed) using "Gleich's syndrome" and "episodic angioedema with eosinophilia" as keywords.



Results: We identified 34 articles published until december 2021, with a total of 88 cases of Gleich syndrome. Throughout these cases, recurrent angioedema with hypereosinophilia were the most commonly reported clinical manifestations. Twenty-seven patients also developed urticaria during angioedema episodes. Twenty-three patients had a history of atopy (genetic tendency to develop allergic diseases) and were diagnosed in the past with: allergic rhinitis, allergic conjunctivitis, food allergy, and seven patients had concurrent asthma. All patients who received first-line treatment with oral corticosteroids showed a favorable outcome, except for two who needed biological therapy with omalizumab and mepolizumab, respectively.

Clinical manifestations	Number of patients
Angioedema with hypereosinophilia	88
Urticaria	27
Atopy	23
Asthma	7

Conclusion: Gleich syndrome defined by episodic angioedema with hypereosinophilia is a benign entity, different from hereditary or acquired angioedema with or without urticaria and primary hypereosinophilia. The activity periods of the disease can remit spontaneously, and the condition is kept under control by the administration of oral corticosteroids in most cases. Cases refractory to corticosteroid therapy have been successfully treated by adding biologic therapy. Defining a set of criteria for the diagnosis of the disease is necessary for the recognition of Gleich syndrome.

References

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