Opis choroby *

Definicja

A rare non-histaminic angioedema characterized by potentially life-threatening episodes of edema of subcutaneous and/or mucosal tissues without urticaria, caused by excessive consumption of C1 esterase inhibitor (C1-INH) in the context of lymphoproliferative or autoimmune diseases. Patients typically present in the fourth decade of life or later and without a family history of angioedema. Clinical manifestation includes nonpitting edema of the skin predominantly involving the face, but also the limbs or genitals, as well as abdominal pain due to involvement of the gastrointestinal mucosa, and severe edema of the upper airway and oral mucosa. Laboratory examination shows low C1-INH activity and low C3, C4, and C1q levels. Autoantibodies to C1-INH are frequently detectable.

Dane

Klasyfikacja
Choroba

Synonimy
Acquired angioneurotic edema with C1 inhibitor deficiency
Nabyty obrzęk angioneurotyczny z niedoborem C1Inh
Nabyty obrzęk angioneurotyczny z niedoborem inhibitora C1
Acquired angioneurotic edema with C1Inh deficiency

Kod ORPHA
528663

Kod OMIM
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Kod ICD10
D84.1

Kod ICD11
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*Źródło

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