A Case of Type 2 Hereditary Angioedema Treated with Acute Exacerbation with Icatibant

서울대학교 의과대학 내과학교실[1], 분당서울대병원 내과[2]

황유경[1,2], 안경민[1,2], 김용현[1,2], 김세훈[1,2], 장윤석[1,2]

Introduction: Hereditary angioedema is a rare autosomal dominant disorder caused by a genetic deficiency or decreased function of C1-esterase inhibitor (INH). It is characterized by swelling of subcutaneous and submucosal tissues of extremities, gastrointestinal tract and upper airways which can be life threatening. Thus early recognition and treatment of disease are important. Short and long-term prophylaxis is used to decrease severity and frequency of attacks. Icatibant is a selective bradykinin B2 receptor antagonist, resulting in earlier treatment of the acute attack and decreased emergency admittance.

Case: The patient was 47-year-old woman, suffered from edema of limbs, swelling of lips, occipital swelling, gastrointestinal edema and abdominal pain for a long time. Her C1-INH activity was decreased and type 2 hereditary angioedema was diagnosed, taking danazol for long term maintenance treatment. Acute exacerbation after chewing the oral mucosal during a meal caused facial edema and visited the emergency room. Her vital signs were stable on presentation. On physical examination, chest was clear to auscultation bilaterally with no wheezing sound. Facial edema and cervical edema, especially severe edema on the left lip and left jaw were found without urticaria or rash. Peripheral blood leukocytes was 12,680/μL, hemoglobin was 13.5 g/dL, platelets was 277,000/μL and C4 was 2.2mg/dL. C1-INH 38.0mg/dL, normal range, but C1-INH activity decreased less than 20%. One hour after subcutaneous injection of Icatibant 30 mg, the symptoms of tightness in the larynx and dyspnea were improved, cervical and facial edema symptoms improved within one hour, and she was discharged the next day.

Discussion: The unpredictability and recurrence of hereditary angioedema attacks could be negative impact on social life and quality of life. And also acute attacks of hereditary angioedema can cause life-threatening emergencies such as laryngeal edema, appropriate measures are needed. In this case shows the timely and proper use of Icatibant on hereditary angioedema type II, could reduce morbidity and mortality.