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당뇨 환자에서 발생한 안면부 털곰팡이증에 대한 증례

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Fatal Facial Mucormycosis on a Diabetic Patient: A Case Report

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Purpose: Mucormycosis generally occurs in patients with uncontrolled diabetes mellitus and immunocompromised conditions. It is rare, but once a patient is infected with it, it can occur as a rapidly extending, aggressive, and life-threatening rhinocerebral infection with a high mortality rate.

Methods: A 70-year-old female patient had a 40 years of history of adult onset diabetes mellitus. She presented herself with severe right hemifacial pain, swelling, and weakness for 3 days and was admitted to ENT. On a facial computed tomography (CT) scan, it was found that her infection extended from her inferior temporal scalp to her submental area and involved her submandibular, masseter, prevertebral, parapharyngeal, retropharyngeal, and pharyngeal mucosal space and pansinusitis. Through endoscopic sinus surgery, mucormycosis was confirmed via histologic examination.

Results: Despite empiric antibiotics and amphotericin B administration, the patient was in a septic condition and in a coma. The patient's family wanted to withdraw her life support, and the patient expired.

Conclusion: Mucormycosis is very rare, but is one of the disastrous complications of uncontrolled diabetes mellitus. Suspicion of its occurrence, based on identified risk factors, and its rapid diagnosis can enhance the chance of its cure.

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Key Words: Mucormycosis, Diabetes complication

I. INTRODUCTION

Mucormycosis, also known as zygomycosis, is an opportunistic infection and a very aggressive and fatal. Its most common pathogens are Rhizopus, Mucor, and Absidia.[1] They are found in soil, animal faeces, or decaying plant materials.[2] Humans are normally resistant to the disease because Mucorales exist in the environment. The factors that predispose humans to infection from these organisms include poorly controlled diabetes, iron or aluminum overload, an immunosuppressive condition, desferoxamine therapy, and renal disease.[1-6] With the increasing prevalence of diabetes and immunosuppressive conditions, mucormycosis is now considered an important fungal infection.

This case report describes the authors' experience of the disease in a patient with uncontrolled diabetes mellitus. It includes a review of literature.

II. CASE

A 79-year-old female patient presented herself with sudden right facial swelling and pain, right hemifacial palsy, and dizziness for 3 days. She presented to black eschar and necrotic change, right hemiface (Fig. 1).

The patient had had diabetes mellitus for 40 years, with poor glycemic control. A laboratory examination revealed a white blood cell count of $19.69 \times 10^3/\mu L$, an erythrocyte sedimentation rate of 84 mm/hr, C-reactive protein of 38.4 mg/dL, blood urea nitrogen of 52.0 mg/dL, creatinine of 2.53 mg/dL, hemoglobin A1c of 10.4%, and fasting blood sugar of 224 mg/dL. The urine was positive for ketones. An arterial blood gas analysis showed a pH of 7.3, an arterial CO_2 pressure of 20.5 mmHg, and plasma CO_2 of 10.7 mmol/L. The patient's brain CT was normal, but her facial CT showed that the infection extended from her inferior temporal scalp to her submental area and involved her submandibular,

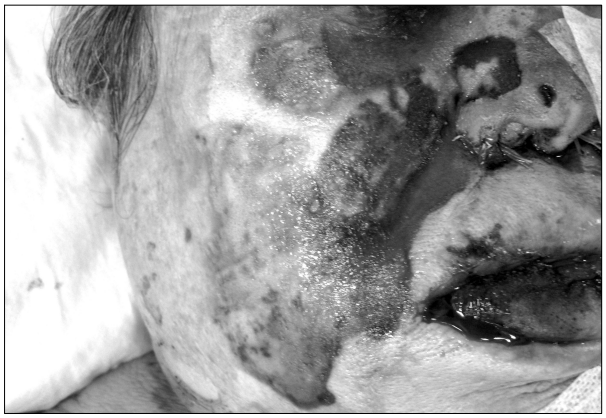

Fig. 1. On hospital day 8, a 79-year-old female patient presented to black eschar and necrotic change, right hemiface.

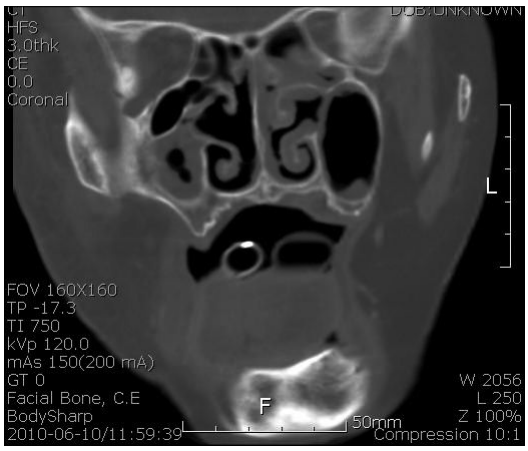

Fig. 2. Preoperative computed tomographic scan showing infection was extended from inferior temporal scalp to submental area with involving pansinusitis.

masseter, prevertebral, parapharyngeal, retropharyngeal, and pharyngeal mucosal space. Pansinusitis was diagnosed (Fig. 2). Internal carotid artery stenosis was also shown. The patient was admitted into the Intensive Care Unit and underwent endoscopic sinus surgery. After the surgery, her biopsy and culture showed broad, nonseptate hyphae with right-angle branching, characteristic of mucorales (Fig. 3). Liposomal amphotericin B was administered to her intravenously, and intravenous fluid and insulin were used to correct her hyperglycemia and ketoacidosis. The patient's general condition worsened, however, and visual loss appeared. The authors planned for debridement, but the Department of Internal Medicine recommended the delay of the operation. Despite all medical efforts, the patient expired due to septic shock and multi-organ failure on hospital day 16.

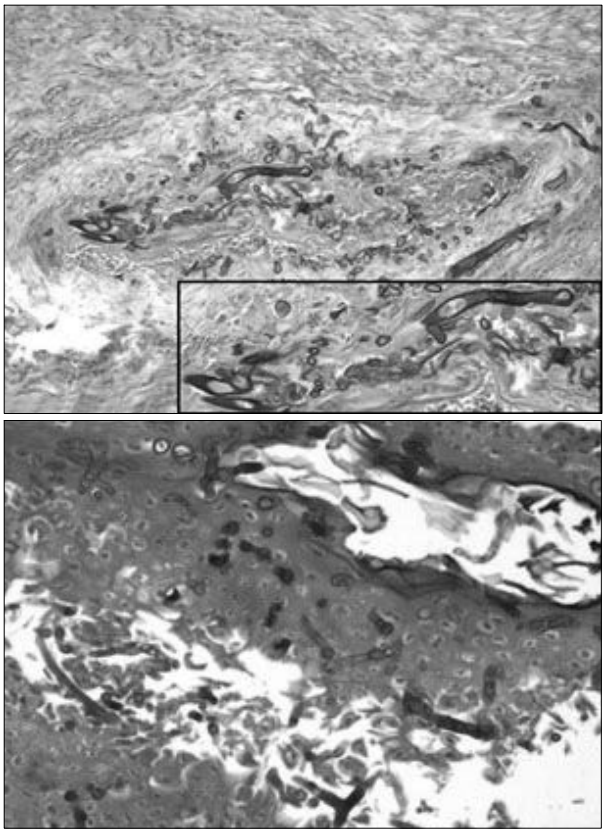

Fig. 3. (Above) The tissue from nasal cavity shows many broad nonseptate fungal hyphae in the vessel wall as well as within the thrombus in the lumen (inset). (Below) Skin biopsy also reveals the totally necrotic and denuded epidermis with many irregularly branching fungal hyphae. Morphologically consistent with Mucormycosis.

III. DISCUSSION

Mucormycosis, or more appropriately known as zygomycosis, is a rare and potentially fatal fungal infection. Mucorales fungi have great affinity to arteries and grow along the internal elastic lamina, causing thrombosis and infarction.[2] Based on the clinical presentation and site of the mucormycosis, it can be divided into six categories: (1) rhinocerebral, (2) pulmonary, (3) cutaneous, (4) gastrointestinal, (5) disseminated, and (6) miscellaneous.[2,5] Adam et al.[7] analyzed 116 cases of mucormycosis in his patients and in literature. The most common type is the rhinocerebral type (39%), followed by the pulmonary type (22%), the disseminated and cutaneous types (16%), the gastrointestinal type (4%), and the miscellaneous type (3%).

Mucormycosis usually occurs in people with poorly controlled diabetes mellitus (especially those with ketoacidosis), solid malignancies, iron overload, and extensive

burns, and in those who are taking glucocorticosteroid agents or neutropenia related to hematologic malignancies.[1-6] A recently identified important clinical feature of mucormycosis is that people with elevated available serum iron are more susceptible to it.[2] Neutropenia, systemic acidosis, and hyperglycemia are other significant risk factors.[2,5] Up to 60~80% of people with mucormycosis have diabetes mellitus, and half of those have diabetic ketoacidosis at the time of their infection.[3]

Symptoms that may suggest mucormycosis in susceptible individuals include multiple cranial nerve palsies, unilateral periorbital facial pain, orbital inflammation, eyelid edema, blepharoptosis, proptosis, acute ocular motility changes, internal or external ophthalmoplegia, headache, and acute vision loss.[2] Contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) are useful, but they present no symptoms of mucormycosis, except for sinusitis.[8] There are no reliable serologic, polymerase-chain-reaction-based, or skin tests for mucormycosis.[2] Therefore, it should be diagnosed via a tissue biopsy and a frozen section.[2,6] Aggressive early surgical debridement and simultaneous intravenous amphotericin B administration are mandatory.[1,2,4,6] Liposomal amphotericin B is more effective for mucormycosis patients, because it has fewer side-effects, is less nephrotoxic, and can be more concentrated in tissues with increased capillary permeability.[2,6] In selected patients, hyperbaric oxygen therapy may play a role.[2,6] Despite of early debridement and appropriate intravenous liposomal amphotericin B administration, mortality rates are high.[7]

High suspicion of this fatal fungal infection in immunocompromised patients is the first step in its treatment. Then early and adequate diagnosis and treatment, including local debridement of all necrotic tissue and simultaneous application of amphotericin B, should have a favorable outcome.

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