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Garcin syndrome caused by rhino-orbito-cerebral mucormycosis

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Abstract

Total paralysis of all or near all unilateral cranial nerves indicates Garcin syndrome. Garcin syndrome caused by Rhino-Orbito-Cerebral Mucormycosis (ROCM) complicating with cerebral infarction has been reported rarely. ROCM is an opportunistic fungal infection with high morbidity and mortality rates despite treatment due to acute angioinvasive infections. We report the case of a 69-year-old female, with a history of type 2 Diabetes Mellitus (DM), who presented Garcin syndrome caused by ROCM.

Keywords: Garcin syndrome; Multiple cranial nerve palsy; Rhino-Orbito-Cerebral Mucormycosis (ROCM).

Abbreviations: CT: Computed Tomography; DM: Diabetes Mellitus; ROCM: Rhino-Orbito-Cerebral Mucormycosis.

Description

A 69-year-old DM female presented right ocular pain with diplopia and fever for 2 days. Neurological examination revealed dilated pupil of right eye with fixed gaze of right eye. CT showed mucus accumulation in paranasal sinus and gas bubbles in the right eyeball (Figure 1A). Two days later, eschar formation over rhino-orbital region with ptosis, impaired sensation, loss of nasolabial fold, absence of movement and sensation on palate and uvula, and deterioration of consciousness. Massive infarction of right middle cerebral artery and swollen scalp were detected on repeated CT (Figure 1B). Fungus culture from nostril yielded Rhizopus spp. Biopsy revealed angioinvasive artery and necrotic debris with fungal hyphae (Figure 1C). Multiple cranial nerve (II, III, IV, V, VI, VII, IX, X) palsy indicated Garcin syndrome in ROCM with intravenous amphotericin B. She died from multiple organ failure 6 days later.

ROCM, an opportunistic fungal infection of zygomycetes, led to a variety of infections in immunocompromised patients. The angioinvasive characteristics of fungi result in infarction and necrosis of infectious tissues, finally black eschars in the nasal mucosa and palate [1,2]. Biopsy analysis of the necrotic tissue is first recommended for diagnosis of ROCM. CT and MRI are suggested to detect the destruction of periorbital tissues and identify the extent, and vascular thrombosis [1,2]. Total paralysis of all or nearly all unilateral cranial nerves indicates Garcin syndrome. The most causes are primary tumors or metastases of skull base and rhinopharynx, and basal meningitis [1,2]. Infections account for 1% of multiple cranial neuropathies in 979 patients. Three of 50 patients with 8 or more impaired cranial nerves had infections [3]. Early diagnosis of ROCM combined with early intravenous amphotericin B and surgical debridement is recommended to improve the survival rate as well as correction of underlying predisposing factors [1,2].

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Figure 1: Dilated pupil with fixed gaze of the right eye was examined on neurological test (1A, upper panel). Swollen soft tissue over the right fronto-orbital region and mucus accumulation in paranasal sinuses gas bubbles in anterior aspect of right eyeball (white arrow in 1A, lower panel). Deteriorated erythema with eschar over the right orbital area and nostril (1B, upper panel). Massive low density in right middle cerebral artery territory (black arrow in 1B, lower panel) and swollen subcutaneous soft tissue were detected on CT. Pathologic picture of nasal biopsy showed angioinvasive artery with necrotic debris (magnitude 400X, white arrow in 1C, upper panel) and fungal hyphae (magnitude 400X, white arrow in 1C, lower panel).

Declarations

Conflict of interest declaration: None.

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