Sequential Blindness in a Diabetic Patient: Unusual Presentation of Rhino-Orbito-Cerebral Mucormycosis

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INTRODUCTION

Mucormycosis is a rare opportunistic fungal infection and can involve various anatomical areas. Rhino-orbito-cerebral mucormycosis (ROCM) can be seen in patients with immunosuppression (particularly common with poorly-controlled diabetes mellitus). Classic inflammatory clinical symptoms and signs, such as facial pain and swelling followed by ocular inflammation with varying degree of visual impairment and ocular motility disturbance, are nearly always present to suggest an underlying infectious process. ROCM can be rapidly fatal if not recognized early and treated promptly.

CASE REPORT:

A 54-year-old diabetic complained of several days of left sided facial pain followed by sudden left eye blindness. He was afebrile without swelling or palpable tenderness over the face or sinuses. Ophthalmologic consultant confirmed severe visual loss with diabetic retinopathy, without signs of ocular inflammation. Diagnosis of posterior ischemic optic neuropathy was made. Brain MRI was unremarkable aside from non-specific ethmoid and sphenoid sinus wall thickening. The patient's presentation was thought to be secondary to giant cell arteritis, and steroids/acyclovir were started despite a ESR and a negative temporal artery biopsy.

Two weeks later he was readmitted for sudden right eye blindness. Despite a complaint of pain over the left maxillary sinus, there was no swelling or erythema and no clinical signs of ocular inflammation. Left oculomotor and abducens nerve palsies were noted without facial numbness. Spinal tap was nondiagnostic with 3 WBC/mm3, a protein of 43 mg/dL and a elevated glucose level of 157 mg/dL, consistent with poorly controlled diabetes. Stains and cultures were

Fig. 1 CT scan: Aneurysm of the left ICA

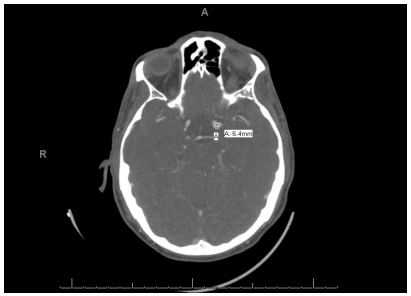


Fig. 2 A coronal section w/ hematoma

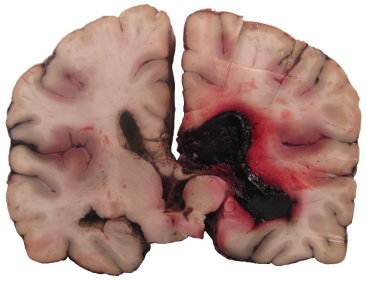


Fig. 3 Aneurysmal ruptured area (H&E)

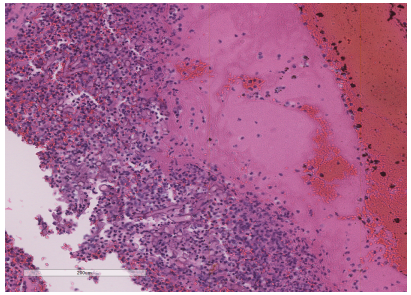


Fig. 4 Aneurysmal ruptured area (GMS)

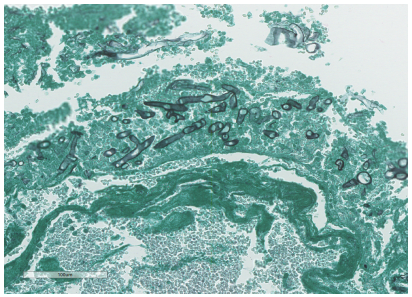


Fig. 5 Aneurysmal ruptured area (PAS)

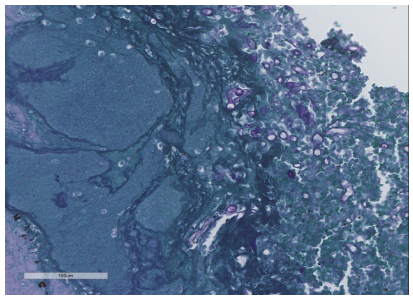
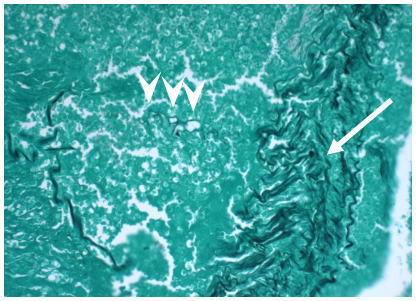


Fig. 6 Aneurysmal ruptured area (EVG)



Endoscopic sphenoid sinus examination disclosed normal appearing mucosa and several random sphenoid sinus wall biopsies revealed no obvious fungal bony invasion. Hyphae seen on sinus surface washing was interpreted as nonspecific mucosal colonization. Two days following sphenoid sinus biopsy the patient suddenly became unresponsive and died shortly. Brain CT scan disclosed aneurysm of the left internal carotid artery (Fig 1), measuring 6.4 mm in maximum diameter, not present one week prior, and basilar subarachnoid hemorrhage and intracerebral hemorrhage confirmed at autopsy (Fig 2). Sectioning of the brain revealed a hematoma (measuring 4.0 x 2.5 x 1.1 cm) that encases tissue in the infundibular area, more in the left side. Microscopic examination of the base of the forebrain showed irregularly shaped, non-septate hyphae with right angle branching and oval conidia which tightly adhere to the blood vessel wall (ICA) with extensive hemorrhage, necrosis and multifocal abscesses (Figs 3-6). Hyphae are highlighted by Grocott's methenamine sliver stain (GMS) and periodic acid–Schiff stain (PAS). Mucor invasion into the right internal carotid artery diagnostic of mycotic aneurysmal rupture was confirmed by Elastic Van Gieson (EVG) stain (Fig 6).

CONCLUSION:

This case illustrates ROCM may present with a predominant sequential blindness without obvious inflammatory clinical signs. A high index of suspicion is warranted in this potentially treatable disease even in absence of clinical-laboratory signs of infectious inflammation.

KEY REFERENCES:

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