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Is the Diagnosis of Acute Posterior Multifocal Placoid Pigment Epitheliopathy with Fatal Cerebral Stroke Correct? A Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: To reiterate the fatal cerebral stroke in association with acute posterior multifocal placoid pigment epitheliopathy (AMPEE).
Report of a Case: A previously healthy 9-year-old girl developed acute visual loss in her left eye and was diagnosed to have unilateral acute posterior multifocal placoid pigment epitheliopathy (AMPEE) elsewhere and received systemic steroid treatment with topical steroid drops. She was seen a month later by us as no visual improvement was noted by the family. The patient was hospitalized for a routine systemic work-up as the diagnosis seemed unconvincing. Three days after the hospitalization, she suffered a cerebral stroke and died in the hospital despite extensive treatment in the intensive care unit.
Conclusion: The neurologic changes including cerebral stroke can be seen in cases with AMPEE and high suspicion should be present in clinicians whenever a case with AMPEE diagnosed.

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1. INTRODUCTION

Acute posterior multifocal placoid pigment epitheliopathy (AMPEE) is in the spectrum of white-dot syndrome complex characterized by multiple circumscribed , flat gray-white lesions at the level of the retinal pigment epithelium with an acute onset of visual deterioration, metamorphopsia, and scotomas [1-3]. During the course, initially observed multiple lesions may end up with various sized chorioretinal scars. Neurologic involvement associated with AMPEE is a rare entity and a variety of clinical presentations such as headaches, strokes, cerebral venous sinus thrombosis , seizures and central nervous system vasculitis can be encountered [4-13]. We hereby report a case with possible unilateral AMPEE having a fatal stroke during the course.

2. CASE REPORT

A previously healthy 9-year-old girl experienced sudden visual loss in her left eye and was diagnosed to have unilateral AMPEE at another eye clinic out of town. Oral and topical steroids were commenced without any visual benefit. As no visual improvement was experienced the family demanded a second opinion and she was seen by us a month after the episode. On our examination, visual acuity was 20/20 in OD and light perception in OS. Left afferent pupillary defect was present. There was no cells in the anterior chamber and vitreous in OS while the right anterior segment was normal. The right fundus was normal, and the left optic disc looked palish and posterior pole looked pale with slightly constricted vessels. Her initial fundus pictures were compared to the newly acquired pictures(Figs. 1a and 1b).

Fluorescein angiogram (FA) and optical coherence tomography (OCT) images were also taken (Figs. 2a, 2b and 2c) and fluorescein angiographic pictures were also compared.

We felt that both the initial and recent fundus pictures of the left fundus were not compatible with the initial diagnosis of unilateral AMPEE and left ophtalmic artery occlusion might be in the differential diagnosis. Thereby, she was hospitalized for a meticulous systemic work up. During the systemic evaluation the patient suffered a syncope episode and developed status epilepticus. She was intubated and admitted to the intensive care unit. A magnetic resonance imaging study demonstrated frontal and left temporo-parieto-occipital cytotoxic edema consistent with ischemic infarcts (Figs. 3a and 3b).

She was aggresively treated for brain edema and refractory status epilepticus in the pediatric intensive care unit. Nevertheless, the brain edema progressed to herniation subsequently and the patient died. The family refused an autopsy.

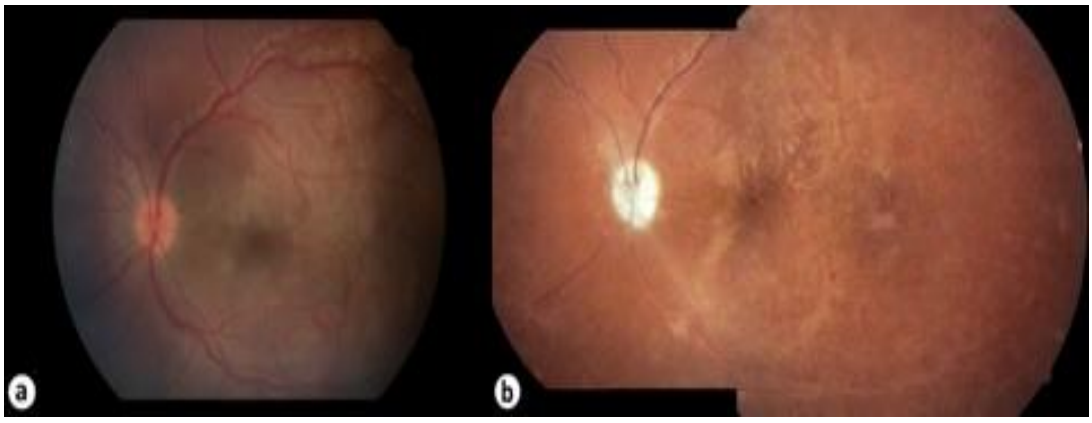

Fig. 1a and 1b. a) Initial color picture showing the pale and slightly swollen optic disc with palish looking posterior pole b) color picture taken a month after the episode showing whitish optic nerve with slight chorioretinal atrophy at the posterior pole

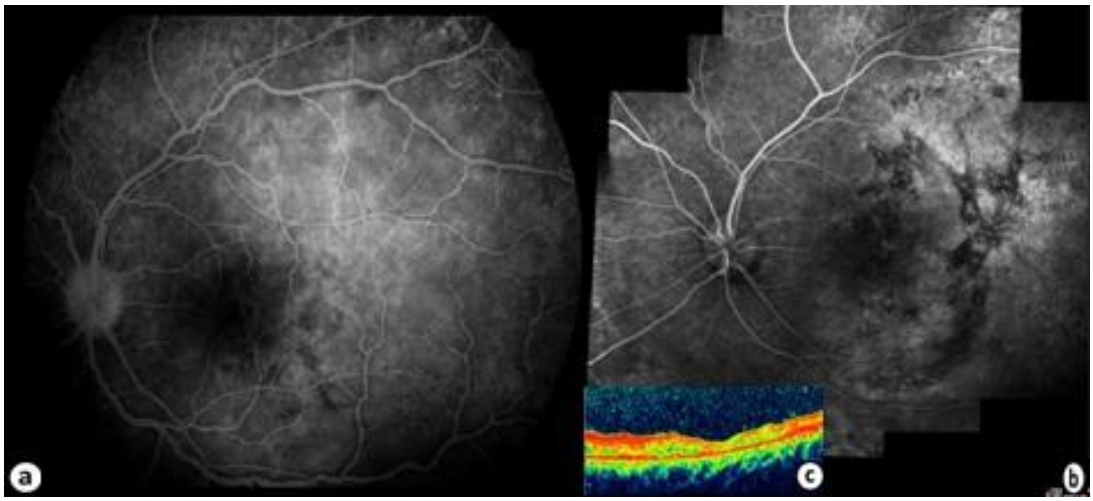

Fig. 2a, 2b and 2c. a) Initial midvenous phase angiogram showing the widespread hyperfluorescence at the deeper layers with disc leaking b) composite midphase angiogram taken a month after the episode showing the widespread window defects at the posterior pole c) thinned retina

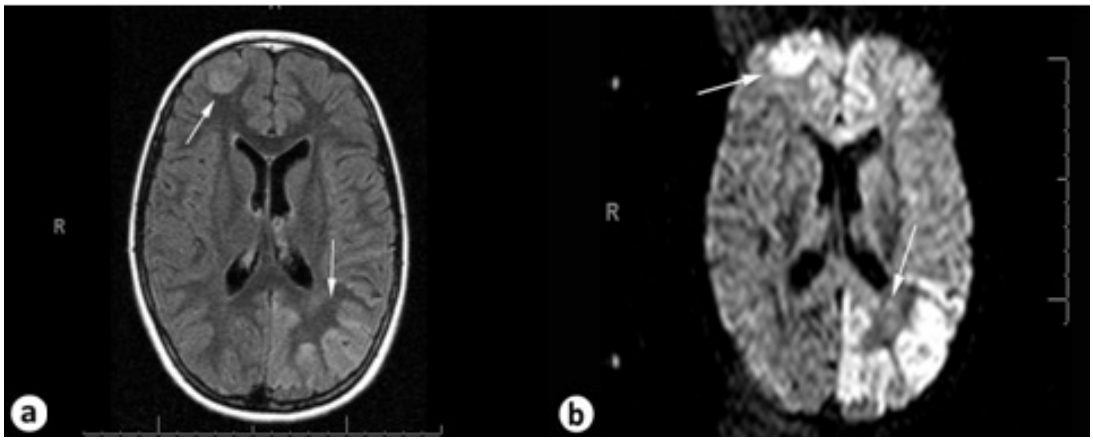

Fig. 3a and 3b. Cranial MRI, FLAIR axial image(a) and diffusion weighted axial (b) images showing bilateral cerebral acute infarction

3. DISCUSSION

Stroke related to AMPEE is most likely to occur simultaneusly or within a few weeks of the ophtalmic disease presentation. So far five cases with AMPEE and cerebral vasculitis with eventual stroke related death were reported [4,5,8,10,13]. Wilson et al. [4] reported a 24-year old man with bilateral AMPEE and he was put on 40 mg/day oral prednisone. Almost a month later, he suddenly collapsed and hospitalized while he was on steroid taper. Radionuclide anterior cerebral perfusion study and EEG demonstrated brain death and the patient died. An autopsy was performed. While there was no evidence of widespread vasculitis, vasculitis changes were demonstrated in medium sized arterioles within the leptomeninges and a few smaller intracerebral vessels. Hammer et al. [5] described a 25-year-old woman who was six months postpartum. She received a diagnosis of bilateral AMPEE and was put on 40 mg/day oral prednisone. Three weeks later she developed hemiplegia. A computered tomographic investigation showed diffuse cerebral infarction and the patient passed away due to cerebral herniation. De Vries et al. [8] reported a 23-year-old white man with bilateral ocular involvement who developed hemiparesis and hyperesthesia three days after the inital eye

examination and receiving the diagnosis of AMPEE. MRI study of the brain showed left medial cerebral artery occlusion and narrowing of the right posterior cerebral artery with infarctions in these vascular territories and the patient died due to herniation. After reviewing the findings in the autopsy, the authors concluded that the cause was focal granulomatous vasculitis affecting the large cerebral arteries. El Sanhouri et al. [10] described a 53-year-old woman with an already established Crohn disease who received a diagnosis of bilateral AMPEE and put the patient on systemic steroid. Prednisone was tapered from 80 mg/day to 20 mg/day over five days. On the sixth day, she developed severe headache and unresponsiveness. She was intubated but three days later she died despite the administration of pulse steroid therapy. Neither MRI nor autopsy could be performed. Very recently, Tsang et al. [13] reported a previously healthy 29-year-old man who had received a diagnosis of bilateral AMPEE following a two month long flu-like symptoms and severe headaches. Approximately ,three weeks later, he developed bilateral cerebral infarction and died 14 days after despite intravenous methylprednisolone and craniotomy for the management of raised intracranial pressure. At autopsy, multiple infarctions of varying ages within a 10 day period were detected.

All above mentioned five patients with AMPEE having a fatal course due to neurological problems had bilateral ocular involvement and their ages varied between 23 and 53 years. Present case was nine years old and had unilateral ocular involvement. Besides, the initial fundus appearance was not compatible with AMPEE according to our clinical judgement. However, as the family refused the autopsy we could not further comment on the diagnosis.

AMPEE should be considered among the causes of stroke and aseptic meningitis in adults. Severe neurological complications are difficult to predict at the time of ocular disease. Neuroimaging or other diagnostic procedures such as lumbar puncture and even cerebral angiography are not necessary for each and every patient with AMPEE unless there is a clue for neurological involvement. It is difficult to predict the potential neurologic complications when the initial diagnosis is established.

4. CONCLUSION

The neurologic changes including cerebral stroke can be seen in cases with AMPEE and high suspicion should be present in clinicians whenever a case with AMPEE diagnosed.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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