Optic nerve sheath fenestration for visual rehabilitation in moyamoya disease

Atanu Barh, Bipasha Mukherjee, S. Ambika[1]

Access this article online

Quick Response Code:

Website: www.saudijophthalmol.org

DOI: 10.4103/1319-4534.310410

Abstract:

A 13-year-old female presented with decrease in vision in both eyes with headache and vomiting for 15 days. Her visual acuity was perception of light with inaccurate projection in the right eye and counting fingers close to face in the left eye. Pupils were nonreactive. Fundus showed bilateral disc edema. Magnetic resonance angiography showed an attenuated caliber of the left internal carotid artery with occlusion of the left middle cerebral artery with collaterals, characteristic of moyamoya disease. The patient underwent right optic nerve sheath fenestration, following which her vision improved to 6/36 in the right and 6/24 in the left eye. Moyamoya disease is an occlusive disease of the cerebral vasculature most commonly seen in the Japanese. Children usually present with ischemic events. In the literature, visual symptoms secondary to raised intracranial tension in moyamoya disease are not well described. Ophthalmologists should be aware of this disease and the treatment options for salvaging vision.

Keywords:

Moyamoya disease, optic nerve sheath fenestration, papilledema

Introduction

Moyamoya disease is a chronic, occlusive cerebrovascular disease of unknown etiology characterized by stenosis of the supraclinoid portion of the internal carotid arteries with the formation of an abnormal vascular network at the base of the brain.[1,2] The term "moyamoya" is a Japanese expression that means something hazy like a puff of cigarette smoke drifting in the air.[2] It has a bimodal distribution of age of onset, with the first peak in childhood and a second peak in the fourth decade of life.[3] Symptoms are either related to ischemic changes or due to intracranial hemorrhage. Children are mostly affected by ischemic changes in the form of motor disturbances, whereas adults are more prone to develop hemorrhagic episodes characterized by altered sensorium.[4,5] Ocular involvement in moyamoya disease is uncommon and mostly due to ischemic changes in the form of anterior ischemic optic neuropathy or retinal vascular occlusion and congenital disc anomalies.[6-9] Optic nerve involvement secondary to raised intracranial tension (ICT) associated with moyamoya disease has not been reported in the literature. We report a rare case of rapidly progressive optic neuropathy due to severe papilledema in a young patient with unilateral moyamoya disease.

Case Report

A 13-year-old female presented with occipital headache with recurrent episodes of vomiting for 1 month. She developed sudden-onset binocular double vision followed by a gradually progressive diminution of vision in both the eyes over the last 15 days. There was no history of fever or convulsion. On examination, she was conscious, co-operative, and oriented. Her visual acuity in the right eye was perception of light with an inaccurate projection of rays and in the left eye was counting fingers close to face. She had sluggishly reacting pupils in both eyes. Intraocular pressure and anterior segment examination were normal in both the eyes. Fundus examination revealed Magnetic resonance (MR) imaging of brain and orbit showed bilateral prominent perioptic cerebrospinal fluid (CSF) space [Figure 2a] with flattening of the posterior

Departments of Orbit, Oculoplasty, Reconstructive and Aesthetic Services and [1]Neuro-Ophthalmology, Sankara Nethralaya, Medical Research Foundation, Chennai, Tamil Nadu, India

Address for correspondence:
Dr. Bipasha Mukherjee, Department of Orbit, Oculoplasty, Reconstructive and Aesthetic Services, Sankara Nethralaya, Medical Research Foundation, 18, College Road, Chennai - 600 006, Tamil Nadu, India.
E-mail: beas003@yahoo.co.uk

Submitted: 10-Sep-2019
Revised: 21-Apr-2020
Accepted: 04-Jun-2020
Published: 27-Feb-2021

How to cite this article: Barh A, Mukherjee B, Ambika S. Optic nerve sheath fenestration for visual rehabilitation in moyamoya disease. Saudi J Ophthalmol 2020;34:223-6.

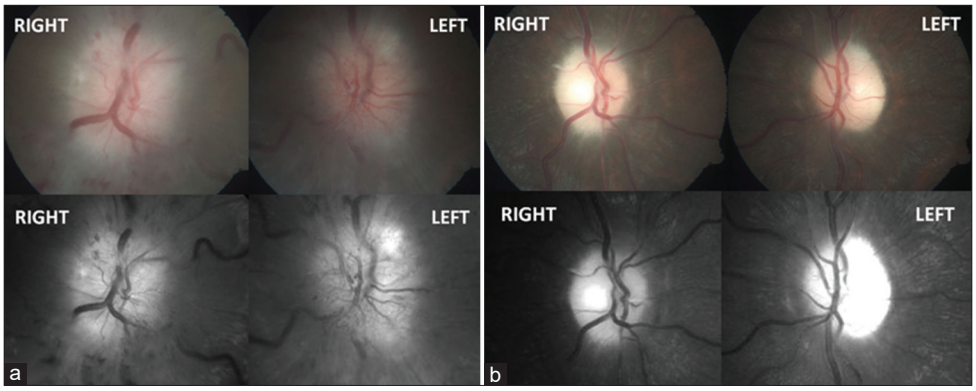

Figure 1: (a) Bilateral florid disc edema at presentation. (b) Significant resolution of bilateral disc edema with the appearance of disc pallor at 1-month follow-up

sclera and tortuous optic nerve. There was no evidence of stroke, granuloma, or space-occupying lesion. MR angiogram revealed attenuated caliber of the left internal carotid artery with occlusion of the left middle cerebral artery (MCA) along with collateral formation [Figure 2b and c], suggestive of left unilateral moyamoya disease. MR venogram ruled out venous sinus thrombosis. Digital subtraction angiography further confirmed the diagnosis of moyamoya disease by detecting the left ICA supraclinoid segment occlusion with the presence of moyamoya blush along the basal ganglia [Figure 3]. Evidence of bilateral stenosis of transverse-sigmoid sinus junction established the diagnosis of raised intracranial pressure causing papilledema. Lumbar puncture revealed raised CSF pressure and normal biochemical analysis. CSF revealed a high opening pressure of 75 cmH2O with normal morphology. Serum antinuclear antibody screening was negative. She was positive for antiphospholipid antibody IgM and negative for anticardiolipin and serum beta-2 glycoprotein. Hemoglobin electrophoresis was within the normal limit, and serum sickling test was negative.

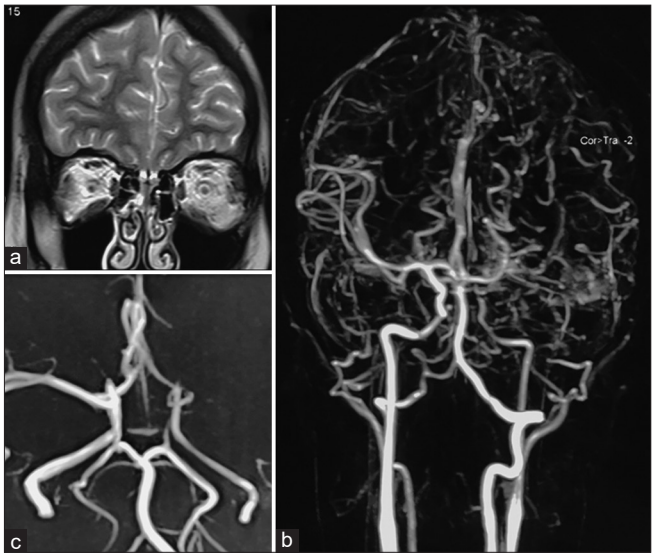

Figure 2: (a) Bilateral prominent perioptic cerebrospinal fluid space. (b) Stenosis of the supraclinoid portion of the left internal carotid artery. (c) Occlusion of the left middle cerebral artery

The patient was initially treated conservatively with intravenous mannitol, oral acetazolamide, and topiramate by neurologists. Systemic conditions including headache and vomiting subsided with the treatment, but vision and papilledema remained unresponsive. In view of the rapidly deteriorating vision, the patient was planned for urgent right optic nerve sheath fenestration (ONSF) under general anesthesia. The right eye was preferred due to the already compromised visual status compared to the left eye. A small window of optic nerve sheath was removed and sent for histopathological examination which revealed fibrocollagenous dural sheath with focal myxoid changes. Postoperative period was uneventful, and the patient was discharged on oral acetazolamide and aspirin. She was advised to consult with a rheumatologist due to high levels of serum IgM antiphospholipid antibody.

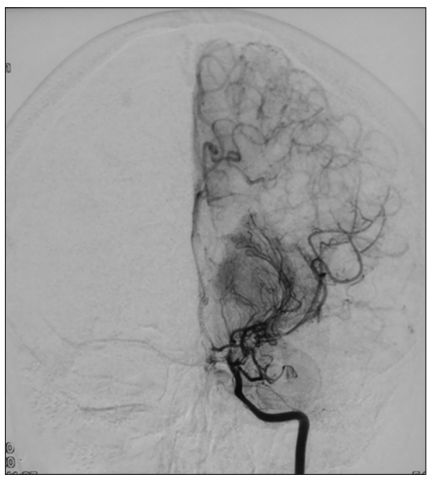

Figure 3: "Moyamoya" blush around the basal ganglia

At 1-month follow-up, the patient showed significant improvement in visual acuity which improved to 3/60 in the right eye and 6/24 in the left eye. Fundus examination revealed significant resolution of bilateral optic disc edema with pallor [Figure 1a and b]. Humphrey visual fields though

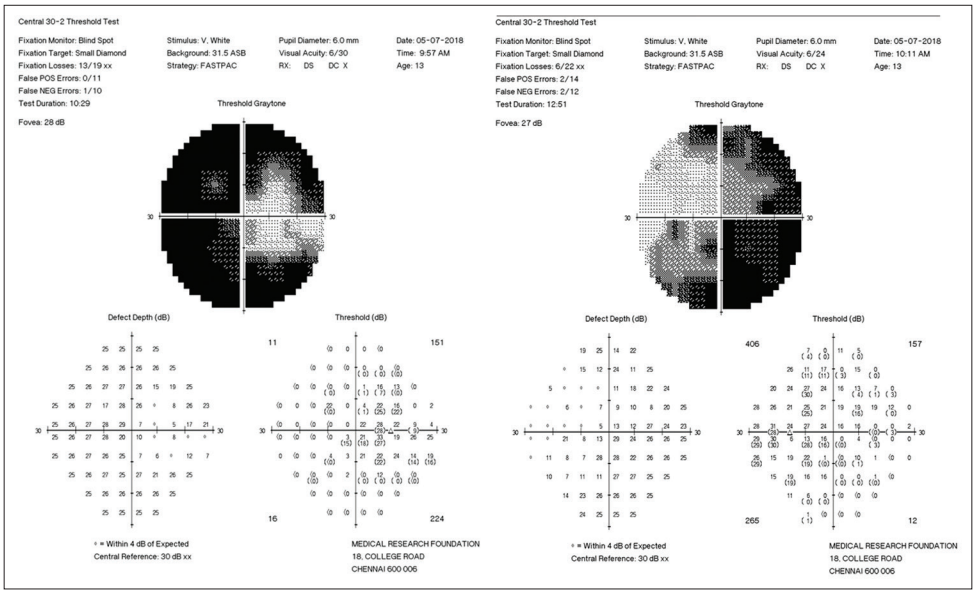

Figure 4: Humphrey 30-2 visual fields showing bilateral constricted visual fields

showed [Figure 4]. She underwent anastomosis of the superficial temporal artery to the MCA (STA-MCA) for cerebral revascularization 2 months after ONSF. Postoperatively, she had recurrent episodes of CSF leak from the operated site which failed to resolve after repeat suturing and lumbar puncture. Finally, she was managed by lumboperitoneal shunt 1 month after STA-MCA anastomosis.

At 6-month follow-up from the first surgery, her vision improved to 6/36 in the right eye and remained stable at 6/24 in the left eye. Her neurological symptoms improved significantly. She was further advised to continue oral anticonvulsant and aspirin with regular consultation with neurologists and ophthalmologists.

Discussion

Moyamoya disease is a spontaneous progressive steno-occlusive disease of terminal portions of the internal carotid arteries or the proximal areas of the anterior or the middle cerebral arteries. The characteristics of moyamoya vessels are the abnormal vascular networks in the arterial territories near the occlusive or stenotic lesions. Clinical symptoms are mostly related to chronic cerebral hypoperfusion. Most of the children present with transient ischemic attack or cerebral infarction, whereas about half of affected adults develop spontaneous intracranial hemorrhage, most often in the basal ganglia and thalamus.[4] In this case, the patient presented with symptoms of raised ICT including headache, vomiting, and false localizing signs, i.e., bilateral abduction limitation due to stretching of the sixth nerve over the petrous bone. The cause of raised ICT in moyamoya disease is not clear. Intracranial and intraventricular hemorrhage can produce hydrocephalus which in turn causes raised ICT.[10,11] In our case, raised ICT was probably due to idiopathic intracranial hypertension. Although ocular involvement is uncommon in moyamoya disease, few works of literature describe some isolated ophthalmic manifestations mostly due to chronic hypoperfusion in the form of retinal venous and arterial occlusion and ischemic optic neuropathy.[6-8] Other rare ophthalmic associations include morning glory disc anomaly, optic nerve hypoplasia, micro-ophthalmos, retinochoroidal coloboma, and nystagmus.[9] Rapidly progressive vision loss due to cerebral hyperperfusion has not been described in any literature. In this present case report, the patient developed profound vision loss within a month of the onset of the symptoms due to severe papilledema. Prompt surgical intervention in the form of ONSF surgery restored significant functional vision in this case.

Although rare, a patient with moyamoya disease can present with profound vision loss. Ophthalmologists should be well aware of this disease as early intervention has a significant effect on quality of life by restoring functional vision.

Acknowledgments

We would like to thank Dr. Suresh Bapu, Director and Consultant Neurosurgeon, Department of Neurosurgery, SRM Institute for Medical Sciences, Chennai, India.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Nishimoto A and Takeuchi S. Abnormal cerebrovascular network related to the internal carotid arteries. J Neurosurg 1967;29:255-60.
2. Suzuki J, Takaku A. Cerebrovascular "moyamoya" disease showing abnormal net-like vessels in base of brain. Arch Neurol 1969;20:288-99.
3. Suzuki J, Kodama N. Moyamoya disease: A review. Stroke 1983;14:104-09.
4. Fukui M, Kono S, Sueishi K, Ikezaki K. Moyamoya disease. Neuropathology 20 Suppl: S61-4, 2000.

5. Kuroda S, Houkin K: Moyamoya disease: Current concepts and future perspectives. Lancet Neurol 2008;7:1056-66.
6. Das D, Handique SK, Bhattacharjee H, Buragohain SK, Bharali G, Deka AC, *et al*. Moyamoya disease with neuro-ophthalmic manifestations: a rare case report. Eye Brain. 2010;2:63-65.
7. Kumar MA, Ganesh BA. CRAO in Moyamoya disease. J Clin Diagnos Res : JCDR 2013;7:545-7.
8. Güçlü H, Gurlu VP, Ozal SA, Esgin H. A moyamoya patient with bilateral consecutive branch retinal vein occlusion. Neuro-Ophthalmol 2016;40:93-6.
9. Bakri SJ, Siker D, Masaryk T, Luciano MG, Traboulsi EI. Ocular malformations, moyamoya disease, and midline cranial defects: A distinct syndrome. Am J Ophthalmol 1999;127:356-7.
10. Rafay MF, Smith SE, Dirks P, Armstrong D, deVeber GA. Hemorrhage predisposing to cerebral infarction in children with moyamoya disease. Pediatr Neurol 2006;34:400-4.
11. Patra S, Purkait R, Sinhamahapatra T, Misra A. Cerebral infarction following intracranial hemorrhage in pediatric Moyamoya disease – A case report and brief review of literature. Ann Indian Acad Neurol 2012;15:60-2.