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RESEARCH ARTICLE

AETIOLOGIES AND PATHOPHYSIOLOGICAL BASIS OF MONOCULAR VISUAL AURA

Chen Yunru

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Abstract

Monocular visual aura is an atypical form of visual disturbance that could be indicative of many different underlying causes, originating in either the vascular or nervous system. The word "aura" is used to refer to migraine aura, but the description of 'scintillations, scotoma and blindness' can also suggest many other aetiologies. There has been a varied use of terminology to describe these visual symptoms, such as "monocular blindness", "monocular visual loss", "phosphenes". This narrative literature review aims to summarise the aetiologies of these monocular visual symptoms and their pathophysiologies. Relevant articles were sourced using databases PubMed and ScienceDirect, and from the bibliography of the articles. 17 articles were selected in total. Monocular visual symptoms were found to be a possible result of benign retinal migraine, as well as vascular and neurological causes. More research is still needed to understand the pathophysiological mechanisms underlying each of the aetiologies.

Introduction:-

Monocular visual auras are visual disturbances specific to only one eye per episode. These auras associated with migraines may be permanent or more often, transient, lasting 5–60 min [1,2]. Persistent auras are defined by the International Headache Society as auras that last for more than a week without radiographic evidence of infarction [3]. During the aura, the affected eye may or may not vary from episode to episode depending on the cause of the aura.

Migraine auras can be in the form of negative or positive symptoms that may or may not occur concurrently. Positive symptoms include scintillations, which are often reported as flashing rays of light, zigzag lightning-like patterns, bright-colored streaks, or halos. Negative symptoms include scotoma ("blind spot") or hemianopsia. Other symptoms can also be visual distortions, blurring, or less commonly, diplopia and metamorphopsia [4]. More elaborate visual auras like the Alice in Wonderland syndrome are rare and occur more often in children and young adults [5]. Most patients who develop visual auras experience only negative rather than positive features alone or in combination with negative ones [2]. Many other disease aetiologies could also result in these symptoms, although they are termed as "monocular blindness", "monocular visual loss" or "phosphenes".

Monocular visual aura may or may not precede or occur alongside other symptoms, such as a headache. Retinal migraines is a benign condition that specifically involves monocular visual auras. ICHD defines retinal migraines as ‘repeated attacks of monocular visual disturbance, including scintillation, scotoma or blindness’, that is not always associated with a headache [6].

Corresponding Author:- Chen Yunru

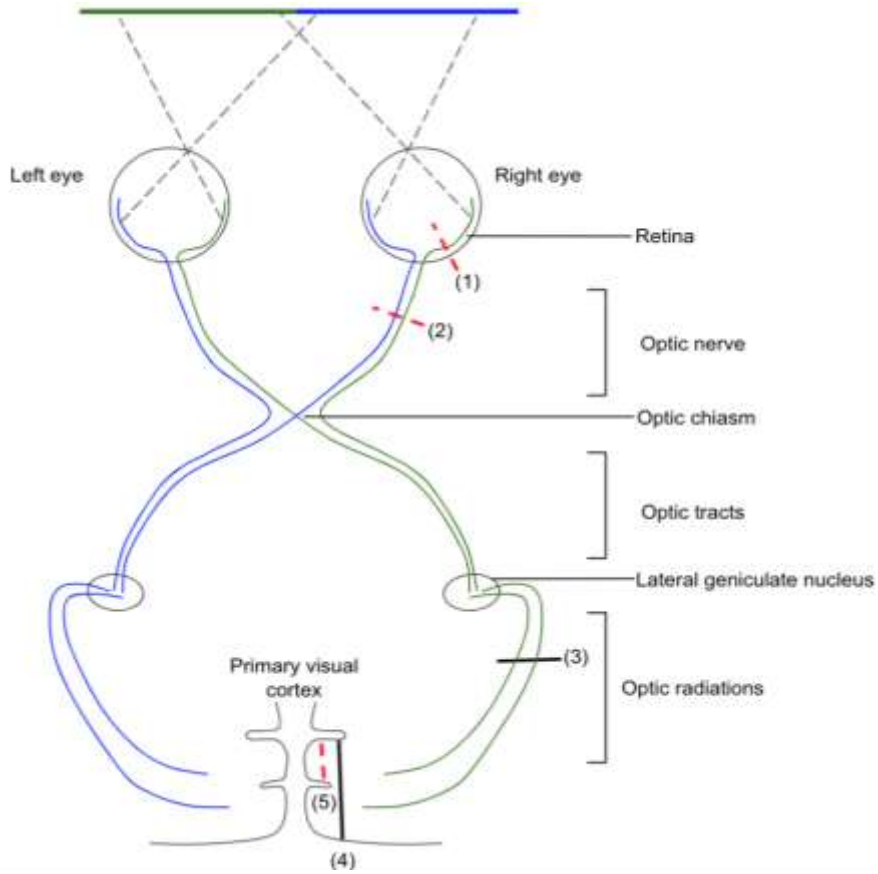

Figure 1:- Diagram of visual pathway.

Lesions occurring along the visual pathway (Figure 1, (3), (4)) or hypoperfusion of any artery supplying blood to these parts of the eye or brain (Figure 1, (1), (2), (5)) can result in monocular visual symptoms. The unilaterality of visual symptoms often indicates that the problem is intraocular [7] (Figure 1, (1)).

Methods:-

The literature review was conducted using two databases: PubMed and ScienceDirect. This review focuses on transient monocular visual auras or blindness. Key terms “Monocular visual aura”, "monocular blindness", "monocular visual loss" and "phosphenes" were included in the search strategy. A total of 26 and 523 articles were found on PubMed and ScienceDirect respectively. Non-English articles (including both abstract and the full text) indexed in the two databases were excluded.

Inclusion Criteria

Articles discussing the pathophysiology of monocular visual auras like unilateral scintillations, scotoma or blindness were shortlisted for review. Relevant articles were also selected from the bibliography of the articles for discussion.

Exclusion Criteria

17 articles were shortlisted for final review. In choosing the articles for final review, the following exclusion criteria were used: systematic reviews and reviews, case reports, and articles not written in the English Language.

Aetiologies and Pathophysiology

Retinal migraine

Monocular visual symptoms can suggest retinal migraine, as a diagnosis of exclusion. Retinal migraine is considered as one of the rare causes of monocular visual auras and may be acephalgic [15]. O'Connor et al. evaluated 61 patients with acephalgic migraine and found that headache is not a necessary feature of migraine attacks. These

patients experienced scintillation and transient hemianopia among other visual symptoms, with only 2 experiencing a headache [16].

Cortical spreading depression (CSD) is a widely accepted model to explain the cause of migraine aura. Based on the 21 quantitative protocols on 3 subjects with migraine auras, Grüsser suggests a mechanism for the aura. He suggests that the aura is initiated by a rise in release of neurotransmitters or focal brain ischemia, which results in local hyperexcitation that then spreads to results in the hyperexcitation of the line of Gennari [17].

A study by Shibata et al. suggested that there are different pathophysiologies in the visual pathway between migraine with aura and without aura. Patients with migraine with aura have increased amplitude of pattern-reversal visual evoked potentials (PVEPs) after attacks, which may be due to CSD [18]. Similarly, another study using visual evoked potentials suggests that the abnormalities in visual pathway may be involved in the pathophysiology of migraine [19]. These studies, however, did not specifically distinguish patients with monocular visual aura.

Interictal visual field deficits in people with migraine are often monocular. Studies suggest that during this interictal period, patients with migraine with aura may have abnormalities in the extrastriate cortex, primary visual cortex and the visual pathway [17,19,20,21].

A study which measured transient retinal and cortical responses to flickering stimuli showed that there was normal retinal processing but abnormal cortical function between migraine attacks [20]. A study by Ditchfield et al. suggests that migraine patients may have abnormalities that extend beyond the primary visual cortex to the extrastriate cortex. Conducted when patients are not experiencing migraine attacks or using medication, patients with migraine generally performed worse on both global motion and global form perception tasks than controls. Since detecting global motion and global form stimuli requires processing of visual stimuli, it is suggested that the poorer performance of migraine patients might be attributed to abnormalities in both the primary and extrastriate visual cortex [21].

A longitudinal study by Sand et al. using visual evoked potentials showed that patients with migraine aura seem to have generally increased visual cortex excitability as compared to patients with migraine without aura and the controls. This increased excitability seems to be detectable from the prodromal phase (defined as within 72 hours before attack), which is evidence of migraine being a result of central nervous system dysfunction [22]. Similarly, two studies by Shibata et al. using PVEPs showed that there is abnormal visual cortex processing in patients with migraine with aura. It is postulated that this excitability of the visual cortex in interictal periods may be crucial in increasing the person's susceptibility to migraine attacks [17,22].

Increased cortical excitability can occur as a result of enhanced cortical excitation or weakened cortical inhibition. A study using binocular rivalry as a measure to determine the applicable mechanism found weak evidence of enhanced cortical excitation in migraine and no evidence of weakened cortical inhibition compared to those without migraine, thus suggesting enhanced cortical excitation to be a more likely cause [24].

Differential diagnosis

Vascular causes

Transient ischemic attack (TIA)

Monocular visual aura caused by TIA are most commonly described as transient monocular visual loss or blindness, or (monocular) amaurosis fugax. Such visual symptoms seem to be uncommonly experienced by patients during the attack. A study of the patient records from 2008 - 2009 in Japan found that only 2.9% (13) out of 444 TIA patients had experienced transient monocular visual loss during the attack [8].

Carotid artery dissection

Monocular visual loss often precedes an ischemic stroke when the internal carotid artery is involved. In a study by Biousse et al., 62% (91) of the 146 patients with extracranial internal carotid artery dissection had visual symptoms ipsilateral to the dissection. 54% (41) of these patients with such symptoms had transient monocular visual auras. Only 31 of the patients with monocular visual auras had associated pain. 23 patients described the auras as positive symptoms like “scintillations” or “flashing lights”, sometimes after changes in posture or exposure to bright lights. 18 patients had a stroke within the first week after the onset of visual symptoms, and 24 had a stroke within the first 2 weeks [9].

Vascular disease

Monocular visual loss can also be caused by acute vascular disease of the choroid, large vessels of the retina or the carotid artery. Rizzo found that 8 patients with monocular visual loss whose cause was otherwise unknown had occlusions either in the retina or choroid [10].

For monocular visual auras caused by vascular causes, ocular or cortical hypoperfusion or occlusion may be the cause of the visual symptoms. Biousse et al. suggests that these symptoms may be related to transient retinal, choroidal, or optic nerve ischemia of embolic or hemodynamic origin. For internal carotid artery dissection, the authors find that the symptoms are more likely of hemodynamic than embolic origin as none of the 76 patients with transient visual loss from ICA dissection had retinal emboli. Patients with monocular visual auras caused by ICA dissection had evidence of ipsilateral ocular and cerebral hypoperfusion suggesting either a reversal of flow in the ipsilateral ophthalmic artery, hypoperfusion in the ipsilateral middle cerebral artery (MCA), or delayed filling of ophthalmic artery and MCA. The authors find that an acute choroidal hypoperfusion seems a more likely explanation for the positive visual symptoms or scintillations than retinal spreading depression [9] (Figure 1, (1)).

Neurological causes

Idiopathic intracranial hypertension (IIH)

Papilledema is frequently caused by IIH. Papilledema can result in increased bulb pressure, retinal ischemia or TIA at the optic nerve, which causes the transient visual obscurations often seen in patients with papilledema [11]. Visual symptoms that present in the form of a sudden onset of transient monocular visual loss is well-associated with TIA [12]. Transient visual obscurations were also frequently reported by patients suffering from IIH, although such symptoms, together with other presenting symptoms like tinnitus, neck pain and obesity are not diagnostic for IHH [13].

The pathophysiology of these visual symptoms in patients with papilledema is currently unknown. The mechanism by which increased intracranial pressure, caused by IHH, results in monocular visual symptoms is also not known.

Lesion

While most retrochiasmal lesions result in homonymous symptoms, the temporal crescent syndrome is the only exception which involves monocular visual defects in the temporal crescent. A study by Landau et al. studied 2 patients with purely monocular visual field defects in the temporal crescent, and found the monocular visual loss to be contralateral to the lesion [14].

Monocular visual loss of the temporal crescent may be caused by damage to the optic radiation, cortical damage or a combination of both. Cortical damage involves a lesion in the primary visual cortex (Figure 1, (4)), which is the most anterior portion that only subserves the temporal crescent of the contralateral eye [14].

Other possible causes of monocular visual auras include ocular diseases like retinal detachment [15].

Diagnosis

Fluorescein angiography has been suggested as an alternative test for vascular disease of the retina or choroid that do not show funduscopic signs. The study by Rizzo suggested the angiogram should be performed with the camera directed at the location of the visual field defect in the affected eye. This could provide more information on the rate and symmetry of blood flow which can help pinpoint the cause of the visual loss [14].

Measurement of visual evoked potential latency and amplitude could be a possible test for the diagnosis of migraine. A study on 41 patients found that N75 and P100 latencies were significantly longer and the amplitudes were higher for the patients with migraine than the control. P100 latency was found to be significantly longer in patients with aura than the patients without aura. The authors found the test to be reliable and accurate in differentiating migraine patients from the control [22].

Conclusion:-

Although monocular visual aura is benign in most cases, care must be taken when eliminating other underlying vascular and neurological causes. While monocular visual symptoms is an atypical presentation that can be quite indicative of specific lesions if the "blind spot" occurs in the temporal crescent, in other cases it requires exclusion of each possible cause to lead to the final diagnosis. Further research on the specific part of the visual pathway that

results in monocular visual symptoms could be done to ascertain the mechanisms by which retinal migraines and its differential diagnosis occur, to aid in the development of more accurate diagnosis for these monocular visual symptoms.

References:-

[1] Handbook of Clinical Neurology 3rd series. (2019). Handbook of Clinical Neurology, v-vi, 511-528. https://doi.org/10.1016/b978-0-444-64029-1.09998-7
[2] Grosberg, B. M., Solomon, S., & Lipton, R. B. (2005). Retinal migraine. Current pain and headache reports, 9(4), 268–271. https://doi.org/10.1007/s11916-005-0035-2
[3] Headache Classification Subcommittee of The International Headache Society The International Classification of Headache Disorders, second edition. Cephalalgia. 2004;24 (suppl. 1): 9–160.
[4] Kunkel R. S. (2005). Migraine aura without headache: benign, but a diagnosis of exclusion. Cleveland Clinic journal of medicine, 72(6), 529–534. https://doi.org/10.3949/ccjm.72.6.529
[5] Geetha Vasudevan, Osman Farooq, Edward Fine (2021). Alice in Wonderland Syndrome With Pediatric Concussion and Synesthesia: A Unique Occurrence (4223), Neurology Apr 2021, 96 (15 Supplement) 4223;
[6] Arnold, M. Headache classification committee of the international headache society (ihs) the international classification of headache disorders. Cephalalgia 2018, 38, 1–211.
[7] Ogun O. (2016). Red flags in neuro-ophthalmology. Community eye health, 29(96), 64–65.
[8] Tanaka, K., Uehara, T., Kimura, K., Okada, Y., Hasegawa, Y., Tanahashi, N., Suzuki, A., Takagi, S., Nakagawara, J., Arii, K., Nagahiro, S., Ogasawara, K., Nagao, T., Uchiyama, S., Matsumoto, M., Iihara, K., Toyoda, K., Minematsu, K., & Japan TIA Research Group 2009-2011 (2014). Features of patients with transient monocular blindness: a multicenter retrospective study in Japan. Journal of stroke and cerebrovascular diseases: the official journal of National Stroke Association, 23(3), e151–e155.https://doi.org/10.1016/j.jstrokecerebrovasdis.2013.09.017
[9] Biousse, V., Touboul, P. J., D'Anglejan-Chatillon, J., Lévy, C., Schaison, M., & Bousser, M. G. (1998). Ophthalmologic manifestations of internal carotid artery dissection. American journal of ophthalmology, 126(4), 565–577. https://doi.org/10.1016/s0002-9394(98)00136-6
[10] Rizzo, Joseph F. (1993). Occult Retinal and Choroidal Vascular Disease. Ophthalmology, 100(9), 1407–1416. doi:10.1016/S0161-6420(93)31468-5
[11] Jensen, R. H., Radojicic, A., & Yri, H. (2016). The diagnosis and management of idiopathic intracranial hypertension and the associated headache. Therapeutic advances in neurological disorders, 9(4), 317–326. https://doi.org/10.1177/1756285616635987
[12] Tuna, M. A., & Rothwell, P. M. (2021). Diagnosis of non-consensus transient ischaemic attacks with focal, negative, and non-progressive symptoms: population-based validation by investigation and prognosis. The Lancet, 397(10277), 902–912. doi:10.1016/s0140-6736(20)31961-9
[13] Radojicic, A., Vukovic-Cvetkovic, V., Pekmezovic, T., Trajkovic, G., Zidverc-Trajkovic, J., & Jensen, R. H. (2019). Predictive role of presenting symptoms and clinical findings in idiopathic intracranial hypertension. Journal of the neurological sciences, 399, 89–93. https://doi.org/10.1016/j.jns.2019.02.006
[14] Landau, Klara; Wichmann, Werner; Valavanis, Anton (1995). The Missing Temporal Crescent. American Journal of Ophthalmology, 119(3), 345–349. doi:10.1016/s0002-9394(14)71178-x
[15] Lal, V., Jogi, V., Mehta, S., Gupta, A., & amp; Singh, P. (2016). More clinical observations on migraine associated with monocular visual symptoms in an Indian population. Annals of Indian Academy of Neurology, 19(1), 63. https://doi.org/10.4103/0972-2327.168628
[16] O'Connor, Patrick S.; Tredici, Thomas J. (1981). Acephalgic Migraine. Ophthalmology, 88(10), 999–1003. doi:10.1016/S0161-6420(81)80027-9
[17] Grüsser O. J. (1995). Migraine phosphenes and the retino-cortical magnification factor. Vision research, 35(8), 1125–1134. https://doi.org/10.1016/0042-6989(94)00187-q
[18] Shibata, K., Osawa, M., & Iwata, M. (1997). Pattern reversal visual evoked potentials in classic and common migraine. Journal of the neurological sciences, 145(2), 177–181. https://doi.org/10.1016/s0022-510x(96)00258-4
[19] Boylu, E., Domaç, F. M., Koçer, A., Unal, Z., Tanridağ, T., & Us, O. (2010). Visual evoked potential abnormalities in migraine patients. Electromyography and clinical neurophysiology, 50(6), 303–308.
[20] Nguyen, B. N.; McKendrick, A. M.; Vingrys, A. J. (2012). Simultaneous retinal and cortical visually evoked electrophysiological responses in between migraine attacks. Cephalalgia, 32(12), 896–907. doi:10.1177/0333102412453953
[21] Jennifer A. Ditchfield; Allison M. McKendrick; David R. Badcock (2006). Processing of global form and motion in migraineurs. , 46(1-2), 141–148. doi:10.1016/j.visres.2005.09.014

[22] Trond Sand; Nikita Zhitniy; Linda R. White; Lars Jacob Stovner (2008). Visual evoked potential latency, amplitude and habituation in migraine: A longitudinal study. , 119(5), 1020–1027. doi:10.1016/j.clinph.2008.01.009
[23] K. Shibata; K. Yamane; M. Iwata; S. Ohkawa (2005). Evaluating the effects of spatial frequency on migraines by using pattern-reversal visual evoked potentials. , 116(9), 2220–2227. doi:10.1016/j.clinph.2005.05.015
[24] F Wilkinson; O Karanovic; HR Wilson (2008). Binocular rivalry in migraine. , 28(12), 1327–1338. doi:10.1111/j.1468-2982.2008.01696.x.