Sudden Onset of Strabismus in Adults- Is It a Life-Threatening Sign?

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ABSTRACT

The overall incidence of strabismus is higher in adults; approximately 1% of children have strabismus, while the estimation among adults is 4% but may run as high as 5.6%. The four types of new-onset strabismus in adults most often found were paralytic, convergence insufficiency, small-angle hypertropia, and divergence insufficiency. The stroke should be suspected in the sudden onset of paralytic strabismus. The most prevalent type of manifest strabismus found on stroke survivors was exotropia. Tumors, myasthenia gravis, thyroid eye disease are common causes of gradual-onset strabismus. Thorough examinations and proper referral are needed.

Keywords: Diplopia, sudden-onset strabismus

Introduction

Strabismus is defined by a disorder of ocular alignment characterized by directional deviation of one eye relative to the other. Although commonly recognized among children, adults also develop strabismus secondary to various conditions, including trauma, surgical procedures, thyroid dysfunction, cranial nerve palsies, or other neurologic diseases.1 The overall incidence of strabismus is higher in adults. Approximately 1% of children has strabismus, while the estimation on the prevalence of adult strabismus is 4% but may run as high as 5.6%.2 If the percentage of 4% prevalence is actual, 1 in 25 adults may develop strabismus and about 10.8 million adults with strabismus in Indonesia. Despite the occasional recurrence of a childhood problem, many adult strabismus cases are entirely new entities.

Discussion

Martinez-Thompson, et al, explained the four types of new-onset strabismus in adults most often found in society were paralytic, convergence insufficiency, small-angle hypertropia, and divergence insufficiency.3 Paralytic strabismus consists of third, fourth, and sixth nerve cranial palsy, internuclear ophthalmoplegia, and myasthenia gravis. Convergence insufficiency happen when a double vision appears while reading at near and absence of double vision at a distance. Small-angle hypertropia is diagnosed when there is a comitant hypertropia and was managed using prisms of ≥3PD with no evidence of oblique muscle dysfunction. Divergence insufficiency is defined when symptoms of diplopia appear at distance vision and absent at near vision. Based on a cohort study conducted by Martinez-Thompson, et al; the most common subtype was paralytic strabismus, reaching as high as 44% of all strabismus patients noted.3

One of the paralytic causes which should be highly suspected in sudden onset of strabismus is stroke. A study conducted by Rowe and VIS Group UK showed the most prevalent type of manifest strabismus found on stroke survivors was exotropia.3 About 77% of stroke patients had a convergence impairment, and exotropia was found in 72% of patients; the incidence was equivalent in the right- or left-sided strokes. The study also showed that around 48% of patients did not have diplopia despite manifest strabismus and 11% were asymptomatic. Paroxysmal exotropia of the left eye as the only symptom of atypical manifestation of transient ischemic attack has been reported.5 A retrospective study on patients with stroke or traumatic brain injury found that the highest oculomotor impairment rate was cranial nerve III palsy strabismus.1 Other than strabismus, denervation to the superior levator palpebra, ciliary body, pupillary constrictor, superior rectus, inferior rectus, medial rectus, and inferior oblique extraocular muscles oculomotor nerve injury is revealed as ptosis, gaze palsy, and mydriasis.5 Stroke patients with oculomotor palsy often have other neurological findings such as...
hemiparesis or cerebellar dysfunction. Solitary oculomotor nerve palsy commonly hinted at other mechanisms such as compression or diabetic microvascular injury.6

Figure 1. Exotropia OS during TIA4

Figure 2. Internuclear ophthalmoplegia22

Internuclear ophthalmoplegia is a disorder of ocular movement manifested as adduction inability of the ipsilateral eye with nystagmus of the abducting eye. Convergence is spared in INO and is a critical diagnostic element in discerning INO from a third nerve palsy.7 Tumors might not present as a sudden onset of strabismus, but it proves to be a concerning issue in diagnosing an underlying cause; the slow manifestation of strabismus with tumor etiology may only be recognized and addressed only when it has reached a significant disturbance.

Another common case of sudden strabismus appears in patients with myasthenia gravis. The first complaint expressed by myasthenia gravis patients is usually droopy eyelid and diplopia and inability to align both eyes properly. The most commonly affected muscle in myasthenia gravis is the medial rectus, followed by the superior rectus.14 Incomitant strabismus, external ophthalmoplegia may mimic motor cranial nerve palsies, but pupils are never involved in myasthenia gravis.15 Ocular myasthenia gravis can mimic any comitant or incomitant strabismus ranging from nerve palsies, gaze palsies, unilateral or bilateral internuclear ophthalmoplegias to even complete ophthalmoplegia.16 These are usually bilateral and asymmetric, insidiously progressing over weeks to months.

Tumors like meningioma and microvascular ischemia are common causes of sixth nerve palsies, especially in elderly with vascular risk factors.12 Hypophysis adenoma, tumor at the petrous apex, cranial fracture, orbital infection, and progressive thrombophlebitis due to sphenoid sinusitis have been reported to cause abducens nerve palsy.8 Tumors might not present as a sudden onset of strabismus, but it proves to be a concerning issue in diagnosing an underlying cause; the slow manifestation of strabismus with tumor etiology may only be recognized and addressed only when it has reached a significant disturbance.

Figure 3. Nine gaze positions on a patient with healthy eyes.19

Hong-Joon Kim, et al, reported a case of paralytic strabismus resulting from pontine hematoma during puerperium.9 Symptoms improved with herbal medicine and acupuncture treatment. Pregnant or puerperal women are reported to have an increased risk of stroke compared to non-pregnant women. In hospital-based and community-based reports, the incidence of ischemic strokes associated with pregnancy or puerperium varies considerably from 4.3 to 210 per 100,000 deliveries.10 Pregnancy-related stroke must be differentiated from a stroke in elderly patients due to its difference in pathophysiological etiologies, mainly due to hypercoagulability, such as venous stasis.11 Recurrence rates and also diagnostic and therapeutic methods may affect the fetal health status. Maternal physiological alterations occur during pregnancy due to the variations of the hormonal status involving the hemostatic and hemodynamic systems.12 whether this adaptation could affect ischemic stroke risk is still unclear, and the relationship is likely complex.

Thyroid orbitopathy may present with various signs, including restriction of one or more extraocular muscles, eyelid fullness, poor convergence, incomplete and infrequent blinking, absent creases in the forehead on superior gaze, resistance to pulling down the retracted upper lid.16 The most common muscles involved in thyroid orbitopathy are the inferior and medial rectus muscles, followed by superior and medial rectus muscles.17 Although exotropia is a more common finding with thyroid-associated orbitopathy; convergence insufficiency has been described. Pseudo fourth nerve palsies have been described with thyroid-associated orbitopathy.16 Exotropia in patients with thyroid-associated orbitopathy should alert physicians on the possibility of concurrent myasthenia gravis.18

It is essential to detect and refer strabismus to a specialist as soon as possible. Physical examination starts by checking ocular alignments.19 This is done by comparing light reflexes from both corneas (Hirschberg Test). In the primary position, reflexes should be in a symmetrical position. Observe whether there are head tilt tendencies in a specific direction during the examination. Do a cover test to confirm strabismus. Examine both
oculomotor movements to determine the location of the anatomical defect.19

Ask the onset of strabismus, and patients report both eyes often crossed since child period. Note if there is also an exophthalmia which indicates Grave’s disease. Chronic exophthalmia is usually associated with a congenital craniofacial anomaly. Enophthalmia is often associated with Duane syndrome or an old blowout fracture. Complete NIII paralysis with incomplete regeneration with lagophthalmia can be seen in severe Grave’s disease. Plagiocephaly and superior oblique muscle can cause facial asymmetry, which can also cause strabismus.20 Evaluate monocular movements. Medial epicanthal may cause pseudostrabismus. In general, alternating strabismus is incompatible with amblyopia. Strong unilateral fixation should alert the physician that there is a reduced visual acuity in the non-fixated eye. Strabismus angle variation often occurs in uncorrected refractive errors, anisometropia, or nystagmus compensation syndrome.20 Check patient’s visual acuity. Check whether the strabismus is latent, intermittent, or manifest through the cover test.

Sensory deficiencies such as cataracts, refractive error, or amblyopia must be addressed since the sensory function is needed to maintain eye alignment and prevent postsurgical drift. In adults with acquired strabismus from trauma or cranial nerve palsy, a period of observation for spontaneous improvement is often indicated. Small deviations may be managed with prism glasses in adults, but more significant variations usually require eye muscle surgery or botulinum toxin injection.21

Never ignore any systemic symptoms such as blood pressure, muscle weakness, body parts weakness. Run workup tests to eliminate differentials and establish diagnoses. Refer patients to the respective field of specialty according to the patient’s need. The complexity of strabismus stresses the need for clinicians to determine the cause, which could turn into a lifesaving decision.

Summary

Strabismus is defined by a disorder of ocular alignment characterized by directional deviation of one eye relative to the other. The overall incidence of strabismus is higher in adults than in children, where the estimation of the prevalence is 4% but may run as high as 5.6%. Four types of new-onset strabismus in adults most often found in society were paralytic, convergence insufficiency, small-angle hypertropia, and divergence insufficiency. Stroke patients with strabismus are most often found with exotropia and a high cranial nerve III palsy strabismus. Strabismic stroke patients occasionally show a form of internuclear ophthalmoplegia (INO). Pregnant or puerperal women also reported having an increased risk of stroke compared to non-pregnant women. Tumors like meningioma and microvascular ischemia are common causes of sixth nerve palsies, especially in elderlies with vascular risk factors. Ocular myasthenia gravis can mimic any comitant or incomitant strabismus ranging from nerve palsies, gaze palsies, unilateral or bilateral internuclear ophthalmoplegias to even complete ophthalmoplegia. It is essential to detect and refer strabismus to a specialist as soon as possible. Thorough physical examination is needed, and physicians need to note never to ignore any systemic symptoms.

REFERENCES

1. Liu GT, Volpe NE, Galetta SL. Neuro-ophthalmology: Diagnosis and management. 2nd Ed. China: Elsevier; 2010.
2. Martinez-Thompson JM, Diehl NN, Holmes JM, Mohney BG. Incidence, types, and lifetime risk of adult-onset strabismus. Ophthal. 2014;121(4):877-82.
3. Rowe F, VS group UK. The profile of strabismus in stroke survivors. Eye (Lond). 2010;24(4):682-3.
4. Wang X, Yu Y, Zhang W, Meng H. Transient ischemic attack characterized by external strabismus of the left eye. Medicine (Baltimore). 2018;97(24):11150.
5. Clifftreda KJ, Kopano N, Rutner D, Suchoff IB, Han ME, Craig S. Occurrence of oculomotor dysfunctions in acquired brain injury: A retrospective analysis. Optometry-J Am Optom Assoc. 2007;78(4):135–61.
6. Pula JH, Yuen CA. Eyes and stroke: The visual aspects of cerebrovascular disease. Stroke and Vasc Neur. 2017;2(4):210-20.
7. Rowe FJ, Wright D, Brand D, Jackson C, Harrison S, Maan T. Profile of gaze dysfunction following cerebrovascular accident. Ophthalmol. 2013;2013:1-8. 264604
8. Khandker N, Schmerler D, Mahajan S, Strbian D, Serra A. Value of eye movement examination in aiding precise localization in stroke. Stroke 2014;45:157–9.
9. Kim HJ, Kim YK, Kang SY, Kim JI, Yoon JW, et al. A case report of one patient with paralytic strabismus resulting from pontine hematoma during puerperium. Korean J Oriental Intern Med. 2004;25(3):640-7.
10. Zott ED, Giossi A, Volonghi I, Costa P, Padovani A, Pezzini A. Ischemic stroke during pregnancy and puerperium. Stroke Res Treat. 2011; 2011: 606780
11. Gear KE, Bushnell CD. Stroke and pregnancy: Clinical presentation, evaluation, treatment and epidemiology. Clin Obstet Gynecol. 2013;56(2):350–9.
12. Prasad S, Volpe NJ. Paralytic strabismus: Third, fourth, and sixth nerve palsy. Neuro Clin. 2010;28:803–33.
13. Ayberk G, Ozveren MF, Yildirim T, Erkan K, Cakir EK, Kocak A. Review of a series with abducens nerve palsy. Turk Neurosurg. 2008;18(4):366-73.
14. Nair AG, Patil-Chhablani P, Venkatramani DV, Gandhi RA. Ocular myasthenia gravis: A review. Indian J Ophthalmol. 2014;62(10):985–91.
15. Noel MC, Burkat CN, Jirawuthiworavong GV, Marct MM, Noel MC, Yen MT. Myasthenia gravis. [Internet] [cited 2020 July 2]. Available from: https://eyewiki.aao.org/Myasthenia_Gravis
16. Ing EB, Roy H. Thyroid-associated orbitopathy. [Internet]. 2019 [cited 2020 June 23]. Available from: https://emedicine.medscape.com/article/1218444-overview#4
17. Şahlı E, Gündüz K. Thyroid-associated ophthalmopathy. Turk J Ophthalmol. 2017;47(2):94–105.
18. Vargus ME, Warren FA, Kupersmith MJ. Exotropia as a sign of myasthenia gravis in dysthyroid ophthalmopathy. Br J Ophthalmol. 1993;77(12):822–3.
19. Helveston EM, Moodley A. How to check eye alignment and movement. Comm Eye Health. 2019;32(107):55.
20. Noorden GK, Helveston EM. Strabismus: A decision making approach. United States: Mosby-Year Book; 2010.
21. Levin LA, Albert DM. Ocular disease: Mechanism and management e-book. Saunders Elsevier, 2010.
22. Johnson B. Site of lesion of internuclear ophthalmoplegia – medicine mcq. [Internet] [cited 2020 July 1]. Available from: https://pgblazer.com/site-of-lesion-of-internuclear-ophthalmoplegia-medicine-mcq/
23. Ayberk G, Ozveren MF, Yildirim T, Erkan K, Cay EK, Kocak A. Review of a series with abducens nerve palsy. Turk Neurosurg. 2008;18(4):366-73