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ORIGINAL ARTICLE

Skew Deviation: Case Report and Review of the Literature

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ABSTRACT

Objective: To present a patient with a sudden onset ocular tilt reaction (OTR) and review recent knowledge and evolving insights of the underlying pathophysiological mechanisms of skew deviation and OTR. Methods: A middle-aged hypertensive man who had previously suffered stroke with good recovery presented with suddenonset double vision, slurred speech, ataxia, and a head tilt. Romberg test was positive. The patient denied having disturbances of visual acuity, eye pain, or recent trauma. The right eyeball was pushed upward. The patient complained of double vision in any gaze direction. Movements of the extraocular muscles (EOMs) in the horizontal plane were normal, whereas vertical version and convergence were not possible. We administered a Hess-Lancaster test, cover test, fundoscopic examination, Parks-Bielschowsky three-step test, upright-supine test, brain magnetic resonance imaging (MRI), transcranial doppler (TCD) ultrasonography, electrocardiogram (ECG), Holter monitor (24 h), and echocardiography. Results: The Hess-Lancaster test showed superior rectus muscle and inferior obliguus muscle palsy to the left and rectus inferior muscle and superior obliguus muscle palsy to the right. The right eyeball fell behind when looking downward and the left eyeball when looking upward. Cover alternating test was positive from vertical, R/L. Examination of the ocular fundus showed incyclotorsion of elevated right eye and excyclotorsion of depressed eye. The Parks-Bielschowsky three-step test was negative. A brain MRI with gadolinium revealed a small zone of diffusion restriction in the medial portion of the right cerebral peduncle and right thalamus. There was a gradual improvement in the patient's neurological status following treatment. Conclusion: Skew deviation, a not uncommon clinical condition, should be promptly recognized when binocular vertical diplopia cannot be interpreted by trochlearis and oculomotor nerve lesion, myasthenia gravis, or orbital pathology. Maddox rod, cover test, Parks-Bielschowsky three-step, and other tests should help to establish the diagnosis. The prognosis depends on etiology, but it is commonly favorable; the majority of patients recover spontaneously after less than a year. More invasive management options should be discussed thereafter.

Keywords: Neuro-ophthalmology, ocular tilt reaction, skew deviation, strabismus, stroke, vertical double vision

INTRODUCTION

For more than a century, Hertwig-Magendie sign or skew deviation have been used to denote acquired vertical strabismus caused by supranuclear posterior fossa pathology.^{1–5} Some suggest that this was the transitory eye condition that Michelangelo suffered while painting the Sistene Chapel in Rome in 1510.⁶ Although for many years wrongly assumed to be of nonlocalizing value, evolving neuroopthalmological and neurotological insights have proved the opposite.⁷

Pathological conditions that affect the brain stem, cerebellum, or peripheral vestibular system may initially present (among other neurological signs) with skew deviation.^{1,2,7} Magendie and, later,Hertwig were the first to describe this condition in animals in 1824. Stewart and Holmes reported vertical strabismus in a patient with a cerebellar tumor and, later on, following gunshot wounds to the head. Keane was the first to propose an imbalance of the otolith-ocular reflex as a possible cause of skew deviation, and

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asymmetric disruption of otolithic projections to oculomotor and trochlear nuclei as the underlying pathological mechanism, and stressed the possible role of polysynaptic pathways via the cerebellum. Brandt and Dietrich described ipsiversive skew deviation as a localizing sign of a pontomedullary lesion, while contraversive skew deviation was found to implicate more rostral, pontomesencephalic lesions.^{2,7–9}

In humans, achieving normal oculomotor activity is dependent upon a complex interplay between complex supranuclear circuitry, brainstem nuclei, cranial nerves III, IV, and VI, their respective neuromuscular junctions, target muscles, and otolith organs that consist of the utricle and saccule.¹⁰ The utriculo-ocular reflex is responsible for detection of horizontal head translation and static head tilt. It transmits information to the midbrain, vertically acting ocular motoneurons, as well as to the interstitial nucleus of Cajal (INC). An imbalance of underlying pathways can cause asymmetric reduction of the translational vestibulo-ocular and ocular counter-roll responses and result in skew deviation. ^{11,12}

TABLE 1. Conditions presented with skew deviation.

Condition	Diagnosis
Neurogenic Otogenic	Stroke
	Brain hemorrhage
	Traumatic brain injury
	Unruptured intracranial aneurysms
	Demyelinating lesion
	Brain tumor
	Abscess
	Raised intracranial pressure
	Hashimoto encephalopathy
	Infection (Ramsey-Hunt syndrome, central
	nervous system cryptococcosis)
	Paraneoplastic encephalomyelitis
	Creutzfeldt-Jakob disease
	Epilepsy
	Syringobulbia
	Arnold-Chiari malformation
	Spinocerebellar ataxia types 3 and 6
	Neurosurgical procedures after resection of
	the cerebellopontine angle tumor, deep
	brain stimulation (DBS)
	Acute unilateral vestibular organ lesion
	Vestibular neuritis
	Post-surgery complication after
Testerne all and all all a	labirinthectomy, vestibular nerve surgery
Internal medicine	Hepatic coma
	Transient complication of cardiac
	catheterization (ischemic strokes not
David in david	detected on neuroimaging)
Drug-induced	Carbamazepine
	 Consequent to oral or intravenous admin- istration of diphenylhydantoin and amitriptyline
	• Intratympanic gentamicin therapy for the treatment of episodic vertigo

Both peripheral and central conditions may present with skew deviation^{1,2,5,7,13–30} (Table 1).

The most common cause of skew deviation is brainstem stroke. Additional forms, such as paroxysmal skew deviation (noise induced due to perilymph fistula—Tullio phenomenon), alternating skew deviation, and transient neonatal skew have been recognized.⁷

Classical skew deviation usually occurs as a part of ocular tilt reaction (OTR), a clinical syndrome characterized by a complex interplay of perceptual (tilt in subjective vertical), ocular motor (tonic or paroxysmal ocular torsion; and skew deviation), and postural (head tilt) manifestations.⁷

OTR and skew deviation are usually ipsiversive with peripheral and pontomedullary lesions, and contraversive with pontomesencephalic lesions; i.e., if the brain lesion occurs prior to crossing the pathway, the lower eye will be on the same side as the lesion, and if the lesion is higher in the brain (after pathways crossing), the higher eye will represent the side of the lesion. In thalamus lesions, the tilts of the subjective visual vertical may be contraversive or ipsiversive.^{7,31} By convention, the term hypertropia is used (regardless of the lesion site) to denote the higher eye, regardless of which is at fault. Three different types of skew deviation are described when they occur as a part of OTR: upward deviation of both eyes (lesion site in utricle; e.g., Tullio phenomenon due to a subluxated stapes foot plate), hypertropia of one eye (lesion is in the dorsolateral medulla oblongata), and simultaneous hypertropia of one eye and hypotropia of the other eye (lesion is in the midbrain tegmentum).³¹ The Parks-Bielschowsky three-step test is commonly used to differentiate between these two conditions. The first step should determine which eye is hypertropic in the primary position (the uncovered eye moves down as the other is covered). The second step should focus on whether the hypertropia increases on right or left gaze, while the third step serves to determine whether the hypertropia increases on right or left head tilt. For clinical purposes, we further divide it according to eye deviation in each lateral field of gaze: comitant (deviation stays the same in each lateral field of gaze); incomitant (hyperdeviation increases in one lateral field of gaze); and laterally alternating skew deviation (hyperdeviation of the abducting eye in each lateral field of gaze).⁹

In some cases, skew deviation can clinically mimic trochlear nerve lesion (especially in the case of laterally alternating skew deviation, which can be difficult to distinguish from bilateral fourth nerve lesion), and it could be difficult to differentiate between these two conditions. In the first case,, other focal neurological signs (astasia, dysarthria, dysmetria) can be present and brain magnetic resonance imaging (MRI) usually reveals a posterior fossa lesion. This is usually not the case with unilateral peripheral trochlear nerve palsy.⁵

Indirect ophthalmoscopy and optical coherence tomography (OCT) retinal nerve fiber layer plots were found to be useful in objectively quantifying the degree of ocular cyclotorsion. While in a trochlear nerve lesion the fundus of the hypertropic eye is excyclorotated, in skew deviation it is incyclorotated and the fundus of the hypotropic eye is excyclorotated.³² In both trochlear nerve lesion and skew deviation, the head is usually tilted contralateral to the side of the hypertropic eye. In the first case, this represents a compensatory mechanism (to minimize hypertropia), while, in skew deviation, it reflects an underlying pathologic mechanism where it strives to realign the eyes and head to a tilted position which the brain perceives as vertical.^{5,7} Thus, skew deviation is associated with a tilt in the subjective visual vertical; i.e., a misconception of vertical.³ This is a basis of the upright-supine test, which is highly specific in differentiating skew deviation from other causes of vertical strabismus.³³ It is thought that, when changing from an upright to a supine position, the orientation of the utricles changes from earth-horizontal to earth-vertical, this leads to a saturation or reduction in the overall afferent activities of the utriculo-ocular reflex, minimizing any asymmetry of the reflex. This leads to the reduction of torsion and vertical misalignment in skew deviation. The test is considered to be positive if vertical deviation decreases by $\geq 50\%$ from the upright to supine position.⁵

The ocular tilt reaction and skew deviation usually resolve spontaneously over weeks to months.⁷ As symptomatic relief, an eye patch and prism may be used. Botulinum toxin and surgical correction are usually effective in more resistant, long-term cases.^{34–36}

CASE REPORT

A 59-year-old man with a long history of high blood pressure and hyperlipidemia, who previously suffered an ischemic brainstem stroke, presented with sudden onset of diplopia, speech disturbances, and gait impairment (drift to the left). He denied having any changes of visual acuity, eye pain, or recent trauma. Neurological examination revealed dysarthria, and a slightly tilted head towards the left shoulder with lowering of the chin. The right eyeball was positioned more upwards compared to the somewhat lowered left eye (vertical misalignment-strabismus). It fell behind when the patient was requested to look downwards and laterally, whereas the left eye fell backwards when the patient was requested to look upwards and laterally. The patient complained of diplopia in any gaze direction. In the Romberg test,

the patient fell to the left side and, while performing a tandem walk, a drift to the left was detected.

Movements in the horizontal axis were intact, while the patient could not perform vertical version and convergence. The Hess-Lancaster test showed superior rectus muscle and inferior obliquus muscle palsy on the left and rectus inferior muscle palsy and superior obliquus muscle palsy on the right. The Parks-Bielschowsky three-step test was negative. A cover alternating test was + from vertical, R/L. Examination of the ocular fundus showed incyclotorsion of the elevated right eye and excyclotorsion of the depressed eye. An upright-supine test was positive.

A brain MRI revealed a small zone of diffusion restriction in the medial portion of the right cerebral peduncle and right thalamus, consistent with impaired posterior cerebral artery irrigation and acute ischemia, as well as diffuse chronic postischemic lesions periventricular, sub-cortically, and in the brainstem. A previous MRI performed between hospitalizations found only chronic vascular lesions supra and infratentorially.

Transcranial doppler (TCD) ultrasonography revealed no significant changes in haemodynamics (stenosis/vasospasm), other than diffuse atherosclerotic changes consistent with age.

An electrocardiogram (ECG) as well as Holter monitor (24 h) detected sinus rhythm and showed non-specific T-wave abnormalities as well as periodical arrhythmias with three isolated supraventricular extrasystoles (SVES). The echocardiography did not disclose a cardiac source of emboli. There was a partial, gradual improvement in the patient's neurological status following treatment.

On a regular follow-up visit (two months after hospital discharge), the patient did not report any visual disturbances, and OTR recovered spontaneously, with normal neurological examination.

DISCUSSION

We present a patient who developed sudden-onset OTR: right-sided skew deviation, head tilt to the left shoulder, and tilt of the subjective visual vertical. The Parks threestep test was negative and the upright supine test was positive. One must bear in mind the possibility that OTR mimics, for example, a trochlear nerve lesion when examining patients who present with head tilt and eye vertical misalignment, in which the Parks-Bielschowsky threestep test is usually positive, the fundus of the hypertropic eye excyclorotated, and the upright-supine test is negative.⁵ In our patient, fundoscopy showed incyclotorsion of the elevated right eye and excyclotorsion of the depressed eye, which corroborates skew deviation as an underlying mechanism. Furthermore, the upright supine test was positive. A brain MRI revealed a small zone of diffusion restriction in the medial portion of the right cerebral peduncle and right thalamus, which is consistent with clinical presentation. Other neurological signs (dysarthria, ataxia) are suggestive of skew deviation as an underlying pathological mechanism of vertical misalignment.⁵

Finally, symptoms usually resolve spontaneously (over weeks to months), as was the case with the presented patient.⁷

DECLARATION OF INTEREST

The authors report no conflicts of interest with this study. The authors alone are responsible for the content and writing of this article.

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