Clinical Reasoning:
Two see or not two see—Is it really double vision?

SECTION 1
A 57-year-old right-handed woman presented to the emergency department with complaints of double vision and intractable nausea that began abruptly 2 days earlier. Her visual symptoms were characterized as seeing overlapping or separate horizontally or diagonally displaced objects. She had no history of headaches or stroke. Her cerebrovascular risk factors included hypertension, type II diabetes, coronary artery disease, and cigarette smoking. Her medications included clopidogrel, lisinopril, paroxetine, and oxycodone. Her family history was notable for late-onset ischemic heart disease in her parents with no first-degree relatives with early vascular disease. On examination, her blood pressure was 158/101 mm Hg, pulse rate was 87 bpm, and she was afebrile. She was alert and fully oriented. Her attention, recall of recent events, and general fund of knowledge were normal. Her speech was fluent and nondysarthric. Cranial nerve examination was notable for no dysconjugacy or nystagmus, but double vision predominately in the horizontal plane, in all directions of gaze. The diplopia persisted with monocular vision in each eye, and did not improve with a pinhole test. The degree of diplopia waxed and waned during the examination, with visual field extinction tests being difficult to perform reliably. Her pupils were equal with bilateral hippus. Visual fields were full to confrontation. Direct funduscopy revealed normal optic discs. She had a mild right hemiparesis with mild right arm and leg drift, but no facial asymmetry. There was mild hypesthesia over her right arm and leg and appendicular ataxia in her right arm that was worse with eyes open. She did not have extinction to double simultaneous sensory stimuli. Gait evaluation was deferred during her initial examination.

Questions for consideration:
1. What is the significance of the presence of diplopia in both eyes, with either eye closed?
2. What more could be elicited from the history and examination to help characterize the problem?
SECTION 2
The patient had the abrupt onset of bilateral monocular diplopia. Monocular diplopia affecting one eye is commonly related to an intraocular process such as a refractive error, cataract, or a macular disorder. Bilateral monocular diplopia due to a refractive cause is unusual because simultaneous acute ocular pathology in both eyes is unlikely. Perception of sight, including the subjective experience of seeing double, involves multifaceted higher-order cognitive processing. Perplexing visual symptoms such as the overlapping images that this patient reported, combined with her motor and sensory abnormalities, raised concern for a central process. The abrupt onset of her symptoms and her risk factor profile suggests a cerebrovascular cause. A more detailed examination of cortical visual perception was then performed.

Additional examination findings. The patient was unable to describe events in the Cookie Thief image, used in the NIH Stroke Scale, or identify complex overlapping figures. She was unable to recognize pictures of famous faces or well-known landmarks; however, she was able to recognize family members and hospital staff. The patient could read text and identify solid colors presented on a tablet computer, but had difficulty reading numbers on Ishihara color plates.

The patient later elaborated that the double vision involved persisting images or trails left by objects, more so in her left visual field. Most of these symptoms improved over hours and largely resolved by the following day.

Question for consideration:
1. How may the patient’s visual deficits be concisely defined or labeled?
Palinopsia is defined as an abnormal perception of visual stimuli in the visual field after stimulus removal, but sometimes experienced as multiple images or trails as an object is moved in the visual field. These phenomena are termed visual trailing or as cerebral diplopia or polyopia when a patient reports seeing 2 or more duplicated images arranged in ordered rows or columns after fixation on an object. This can be due to ocular disease, but tends to localize to lesions involving the primary visual cortex, optic radiations (more commonly right-sided), or optic tract. It also can be caused by a range of conditions such as toxicity from medications or illicit drugs, seizures, migraine, or traumatic brain injury. In this patient, additional history and examination confirmed the visual complaint was consistent with palinopsia.

Visual agnosia encompasses impairment in perception (apperceptive agnosia) and recognition (associative agnosia) of visually presented objects, not due to a deficit in vision. Prosopagnosia is a subtype of associative visual agnosia with impaired recognition of familiar faces that tends to localize to lesions of the right fusiform gyrus. This patient had difficulty recognizing pictures of famous landmarks and faces. She had no apparent visual field deficit or extinction to double simultaneous stimuli. She had no trouble naming common objects or recognizing family or staff. Her difficulty recognizing pictures of faces and famous places, but not with other visual stimuli, or recognizing people in person when richer perceptual cues were present, suggests an associative visual agnosia.

Simultanagnosia is a type of apperceptive visual agnosia, commonly defined as the inability to perceive multiple objects at the same time, typically associated with parieto-occipital lesions. It may be more accurately defined as the inability to perceive and decode the scene as a whole whereas the ability to identify discrete elements is not impaired, i.e., “seeing the forest but not the trees,” or also perhaps a tree but not the forest.

Simultanagnosia was identified when the patient could not identify overlapping objects in pictures or decode complex visual scenes, whereas she had no difficulty naming objects presented in simple pictures. Simultanagnosia that occurs with impaired reaching or grasping under visual guidance (optic ataxia) and impaired gaze fixation (oculomotor apraxia) define Bálint syndrome. This syndrome tends to localize to the occipito-parietal junction, involving dorsal, spatial visual associative pathways, sparing the ventral, semantic connections. Bálint syndrome may occur with bilateral middle cerebral artery/posterior cerebral artery watershed infarcts. Although this patient had transient right upper limb ataxia that was worse with her eyes open, there was neither convincing optic ataxia nor oculomotor apraxia.

Question for consideration:
1. To what areas do the patient’s symptoms localize?
SECTION 4

A CT scan of the brain revealed only an area of hypointensity in the right cerebellar hemisphere. CT angiogram showed marked atherosclerotic disease in the vertebrobasilar system, but no arterial occlusion. A diffusion-weighted MRI was obtained because of concern for a parietal or parieto-occipital cortical stroke (figure). This showed scattered lesions, consistent with multiple emboli to the bilateral posterior cerebral artery (PCA) territories, involving both primary and visual associative areas. Small pontine perforator and right superior cerebellar artery (SCA) distribution strokes were also present.

DISCUSSION

This case demonstrates the clinical challenge of localizing a subjective complaint of double vision, which may take many forms and can localize to the eyes, oculomotor systems, visual pathways, and as in this patient, central structures involved in conscious visual perceptual processes. A careful history and examination was key. Bilateral monocular diplopia or any other unusual visual symptoms should lead to testing of higher order visual function, which may be quickly and easily performed during the initial evaluation. Portable tablet computers have removed the inconvenience of performing such examinations in clinical settings. This is a paradigm shift in the behavioral examination, elements of which may now be rapidly performed at the point of care.

In this case, an MRI brain scan confirmed an etiologic diagnosis of stroke with ischemic lesions involving portions of the inferior temporal and occipital lobes bilaterally, including small infarcts adjacent to the primary visual cortex (lesions 2 and 4), as well as infarcts in the brainstem and cerebellum. Palinopsia without visual field defects can occur with lesions in the posterior visual pathways. Interestingly, visual hallucinations have been described with brainstem lesions, and this patient also had a small pontine infarction. The phenomena of Lhermitte peduncular hallucinosis are typically described as well-formed, bright, colorful shapes, patterns, or images. Palinopsia, as occurred in this patient, does not fit well with the previous descriptions of peduncular hallucinosis, nor would her visual agnosia or simultanagnosia. The brainstem and cerebellar lesions can, however, account for her mild right-sided sensory abnormalities, right hemiparesis, and right arm ataxia (lesions 10–12).

Although the patient’s symptoms were present for several days at the time of her initial presentation and she was not a candidate for acute intervention, a high index of suspicion, including querying for visual or...
ataxic symptoms, is needed to improve the chance of rapid clinical diagnosis. Seizures, migraine, trauma, and hallucinogenic drugs or toxins need to be considered in the differential diagnosis. A primary psychiatric cause should not be diagnosed without a thorough evaluation for a structural abnormality. Approaches to identifying this problem include having the patient describe the Cookie Thief picture. Overlapping figures is a useful bedside screen for visual perception problems in a multitude of clinical settings. Figures can be obtained from the article by Giannakopoulos et al., and are an excellent addition to examination aids carried by the neurologist.

Anecdotally, the famous faces images we have long carried (e.g., George Bush, Princess Diana) were famous during their era, but can mystify younger patients. To keep pace with the fading of fame, a clinically useful, if not validated, set of currently famous (or infamous) faces and places can be obtained on the Internet. Use of electronic media also allows easy portability and rapid access to specialized assessments such as the overlapping figures, the NIH Stroke Scale, and other measures that can be useful in specific acute settings.

This patient illustrates how commonly available technology (the computer tablet) can aid clinical diagnosis, and together with modern imaging, lead to a better understanding of the potential causes of transient clinical symptoms and findings.

AUTHOR CONTRIBUTIONS
Richard Ronan Murphy: concept and design, critical revision of manuscript for intellectual content. Abdullah Al Sawaf: acquisition of clinical examination findings. Danny R. Rose, Jr.: critical revision of manuscript for intellectual content. Larry Goldstein: critical revision of manuscript for intellectual content. Charles D. Smith: critical revision of manuscript for intellectual content.

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