

Acute reversal of vision metamorphopsia: report of two cases and literature review

Metamorfopsia aguda com inversão da visão: relato de dois casos

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Abstract

Background and Purpose: Acute reversal of vision metamorphopsia (RVM) is a rare, paroxysmal, sudden and transient vertical inversion of vision, also known as floor-on-the-ceiling phenomenon. Its pathogenesis is uncertain, but it may occur due to a transient impairment of vestibular projections to the parietal lobes. The purpose of this manuscript is to report two new cases of RVM with very distinct features. **Cases description:** 1) A 60 year-old man with a right paramedian pontine stroke developed episodes of upside-down reversal of vision during transition from awake to sleep time (hypnagogic acute reversal of vision metamorphopsia) that subsided after treatment with gabapentin; and 2) A 34 year-old woman who had a brief episode of acute reversal of vision metamorphopsia and was diagnosed with systemic lupus erythematosus. She was also diagnosed with a pseudotumor cerebri. **Conclusions:** In summary, acute reversal of vision metamorphopsia is rare and can be secondary to transient brainstem dysfunction. To our knowledge, this is also the first report of its presentation as a hypnagogic phenomenon due to a high pontomesencephalic stroke and in a patient with systemic lupus erythematosus.

Keywords: Vision disorders, Metamorphopsia, Brainstem infarction

Resumo

Objetivo: A metamorfopsia aguda com inversão da visão é um distúrbio raro, paroxístico, súbito e transitório da visão, também conhecido com fenômeno “chão no teto”. Sua patogênese é incerta, mas provavelmente se deve a uma disfunção transitória de projeções vestibulares para os lobos parietais. A proposta deste manuscrito é relatar dois novos casos de metamorfopsia com características bem distintas. **Descrição de casos:** 1) Um paciente masculino, de 60 anos, com histórico de infarto pontino paramediano direito que apresentou episódios de inversão completa da visão durante transição do estado de vigília para o estado de sono (metamorfopsia hipnagógica aguda com inversão de visão) e que apresentou melhora do quadro após tratamento com gabapentina e, 2) Uma paciente de 34 anos que apresentou um breve episódio de inversão da visão e teve diagnóstico posterior de lúpus eritematoso sistêmico. Foi diagnosticada também com pseudotumor cerebri. **Conclusões:** Em resumo, a metamorfopsia aguda com inversão da visão é um distúrbio raro e pode ser causado por disfunção transitória do tronco encefálico. Pelo que se sabe, essa é a primeira vez que se descreve a apresentação desse distúrbio como um fenômeno hipnagógico devido a um infarto pontomesencefálico alto e em um paciente com lúpus eritematoso sistêmico.

Palavras-chave: Transtornos da visão, Metamorfopsia, Infarto de tronco encefálico

BACKGROUND AND PURPOSE

Metamorphopsia can be defined as a visual illusion that distorts shape, color, size or angulation of objects. The so-called “Reversal of Vision Metamorphopsia” (RVM) is a rare, paroxysmal, sudden and transient form, described as a 180°, vertical inversion of vision, also known as “floor-on-the-ceiling phenomenon” or “room tilt illusion (RTI)”¹.

Symptoms of inverted vision have been observed in patients with strokes (especially brainstem/cerebellum), brain tumors, head injury, epilepsy, migraine, multiple sclerosis and brainstem injuries^{2,3,4,5}. The mechanisms underlying the phenomenology of RVM are not fully understood, although as previously discussed by some authors it may be a manifestation of an acute central nervous system insult, specifically a manifestation of transient dysfunction of the vestibular projections to the parietal lobes, as a result from a transient failure of the mechanisms mediating reinversion of the images that enter the retina⁶. The favorable

response to anticonvulsants in some cases suggests a possible underlying hyperactivity of cortical or subcortical neurons⁽²⁾.

The purpose of this manuscript is to report 2 new cases of RVM with very distinct features. Part of this study has been reported in abstract form elsewhere⁷

CASES PRESENTATION

Case 1

A 60 year-old man with a history of hypertension and alcoholism was diagnosed with colon cancer and underwent a partial colectomy without colostomy. During his hospital course, he developed sepsis and recurrent infections (including osteomyelitis), requiring inpatient care for a period of five months.

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During his inpatient care, he developed confusion, which consisted of recurrent paranoid ideas about “having receiving a gift and being stolen” that were persistent and occasionally associated with vivid visual hallucinations according to his family. Visual hallucinations improved, but thereafter, he started to complain of recurrent “visual changes in his room” during the transition from the awake to the sleep state. He described them as if “the room had turned 180 degrees upside down”. These episodes lasted for about 2-3 minutes and occurred 4 to 5 times. He denied oscillopsia, sensation of body levitation or any form of out-of body experience.

His neurological exam revealed normal orientation to time and place, intact cranial nerves, brisk reflexes in the upper extremities but was otherwise non-focal (there were also signs of peripheral neuropathy in his legs). A Brain Magnetic Resonance Imaging was ordered and revealed a subacute right paramedian pontine stroke (Figure 1) with significant involvement of the pontine tegmentum at the pontomesencephalic transition. He continued to take clopidogrel for stroke prophylaxis. He had no further episodes after he was started on gabapentin (300 mg BID).

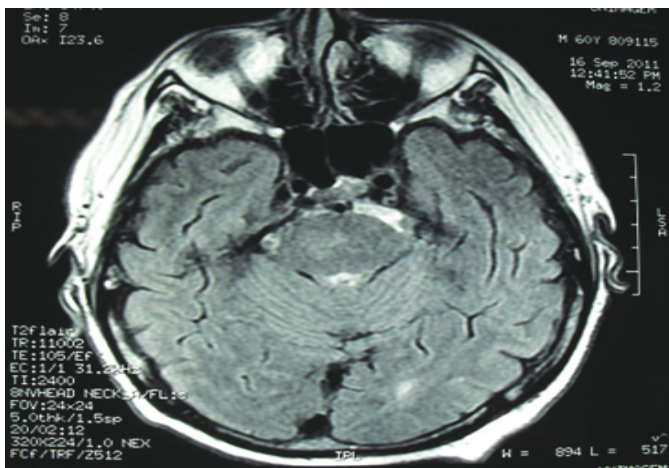


Figure 1 - Brain magnetic resonance FLAIR imaging (MRI), Case 1, highlighting the presence of a subacute right paramedian pontine ischemic stroke (right pontomesencephalic junction) with more prominent involvement of the pontine tegmentum (including the reticular formation).

Case 2

A 34 year-old woman reported that in 1997 she developed strange visual symptoms, right before she was diagnosed with systemic lupus erythematosus (SLE). At that time, she said that she did experience weight loss, hypermenorrhea, anemia, severe joint pain and visual changes (visual scotoma, diplopia and blurred vision) and that right before the diagnosis of SLE was made, two days after she was admitted to a hospital for inpatient investigation (because of an episode of pneumonia), she was in her living room when she suddenly saw the “ceiling on the floor”. According to her, all the objects and furniture that were initially on the floor moved all the way up to the ceiling, and vice versa. She also reported that she closed her eyes (because of fear) and when she opened them, she saw the inferior half of her sister with her feet in the floor (still located on the top of the room), while the superior half

of her body (including her head) followed upon an imaginary line that separated the two halves (the abdomen was turned to the ceiling). She had no further episodes in the subsequent years. However, a few years later, she subsequently developed headaches and was diagnosed with pseudotumor cerebri. Work-up for cerebral venous thrombosis was negative and she was successfully treated with acetazolamide. A Head CT, Brain MRI, MRA and MRV were all negative.

CONCLUSIONS

Our two patients illustrate very distinct features of RVM. Patient one initially had symptoms suggestive of top of the basilar syndrome, that may include visual hallucinations⁽⁸⁾. Although he had a history of alcoholism, the clinical course of the hallucinations and his neurological exam were not consistent with Wernicke or Wernicke-Korsakoff syndrome and he also had vitamin supplements during his hospital course, although thiamine or transketolase levels were not checked. Thereafter, he developed episodes of transient and short-lasting vertical inversion of the vision (RVM) during the transition from the awake to the sleep stage (hypnagogic RVM) and was diagnosed with a brainstem stroke. To our knowledge, this is the first report of hypnagogic RVM associated with ischemia in the pontomesencephalic transition (right paramedian pontine stroke with more prominent involvement of the pontine tegmentum, see Figure 1). There is at least one report of hypnagogic hallucinations due to peduncular hallucinosis, but to the best of our knowledge, hypnagogic RVM has never been reported in patients with pontine strokes.

As it can be seen in table 1, there are a few other cases of brainstem ischemia and brainstem strokes in patients with RVM. None of the reviewed cases were associated with hypnagogic onset. Spells usually lasted from 3 to 60 minutes. Strokes in the territory of the PICA artery with frequent involvement of different parts of the cerebellum were the most commonly reported localization. At least in one of the reports⁽⁹⁾, the pontomesencephalic junction was involved, in the same localization of our patient, although with a less pronounced involvement of the pontine tegmentum. In several prior observations, vertebrobasilar insufficiency (TIAs) was the most commonly reported pattern and patients were frequently treated with i.v. heparin^(2,10). However, as seen in our patient and previous cases, response to gabapentin may also indicate an underlying overactivity of brainstem centers due to the direct effect of the stroke/ischemia or secondary diaschisis.

On the other hand, the neuroimaging of the second patient didn't reveal any lesion that could explain the occurrence of the inversion of the vision, although Brain MRI was only performed years after the episode of RVM. At that time, she had no other neurological complaints or deficits, but was later diagnosed with pseudotumor cerebri. To our knowledge, this is the first report of occurrence of the “floor-on-ceiling” phenomenon in a patient with SLE. She also had no significant history of migraine, condition that has been previously associated with RVM⁽¹⁾ Brainstem lesions and RVM were also described in patients

with multiple sclerosis⁵, brain tumors¹¹ as well as after surgeries for treatment of the treatment of acoustic neurinomas⁴ and seizures³. In our previous study of RVM, in a patient with repeated episodes of RVM and old paramedian pontine stroke² the video EEG also did not disclose any abnormality, which may further reinforce that the phenomenon is due to subcortical (brainstem) rather than direct cortical involvement.

In summary, RVM is a rare disorder of visual processing, which in most instances is secondary to brainstem disease. Structural abnormalities cannot be demonstrated in all cases (patient 2), but the association of RVM with hypnagogic features suggests the role of pathways that run in the high pontomesencephalic reticular formation near the brainstem centers that regulate sleep patterns in humans.

Table 1 - Literature Review of patients with brainstem ischemic strokes and reversal visual metamorphopsia (RVM) who underwent Brain imaging (Brain magnetic resonance imaging except as noted).

Author	Ref	Lesion localizaton	Duration and Associated symptoms
Charles	12	L PICA stroke	30 min; vertigo, vertebral dissection
de Pablo-Fernández	13	R PICA stroke (occlusion)	60 min; vertigo and ataxia
Gondim	2	Old R paramedian pontine	60 min spells, vertigo & body levitation
Horga-Hernández	14	R PICA stroke	3 min; truncal ataxia, R eye hypotropia
Mehler	10	N=3, TIAs? Head CT only	Variable; syncopal spells, ataxia, tremor
Nowaczewska	9	R high pons+L cerebellum	Vertigo, prior episode of body levitation
River	6	Variable (N=3)	Strokes in brainstem or cerebellum
Slavin	15	Wallenberg syndrome?	Ectatic vertebral artery
Stracciari	16	R PICA stroke	20 min (x3), R occipital headache

CT: computerized tomography, L: left, PICA: posterior inferior cerebellar artery, R: right, TIAs: transient ischemic attacks

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