Seizure 2004; **13:** 250–253
doi:10.1016/S1059-1311(03)00194-8

Single-photon emission computed tomography in a patient with ictal metamorphopsia

KYOUNG HEO[†], YANG JE CHO[†], SEUNG-KOO LEE[‡], SUN AH PARK[†], KYU-SIK KIM[†] & BYUNG IN LEE[†]

Departments of [†]Neurology and [‡]Diagnostic Radiology, Yonsei University College of Medicine, Seoul, South Korea

Correspondence to: Kyoung Heo, M.D., Department of Neurology, Yonsei University College of Medicine, Severance Hospital, Shinchon-dong 134, Seodaemoon-ku, Seoul 120-752, South Korea.
E-mail: kheo@yumc.yonsei.ac.kr

Metamorphopsia is a type of visual illusion, which has been reported as a rare ictal manifestation. The patient presented with a simple partial status epilepticus characterised by continuous facial metamorphopsia, intermittently accompanied by elementary visual hallucinations or other types of visual illusions. Subtraction single-photon emission computed tomography images showed an increased perfusion in the ventrolateral aspect of the right temporo-occipital junction (middle and inferior occipital, and inferior temporal gyri). The result suggests that the anatomical substrate involved in the generation of ictal facial metamorphopsia is located in the visual association areas at the right temporo-occipital junction.

Key words: metamorphopsia; visual illusion; epilepsy; status epilepticus; single-photon emission computed tomography (SPECT).

INTRODUCTION

Visual illusions are defined as misperceptions of real external stimuli. The images involved depend on the type of visual disorder, axis, distance, size, shape, motion, number of images, extinction, and memory[1]. Metamorphopsia, a disorder affecting the perception of shape, refers to changes in the form and contour of visually perceived entities. Metamorphopsia is a rare manifestation of seizures[2–9]. Herein, we describe a patient who experienced a state of continuous metamorphopsia representing a simple partial status epilepticus, which was investigated by single-photon emission computed tomography (SPECT) studies.

CASE REPORT

A 56-year-old right-handed woman was admitted due to recurrent flickerings and continuous metamorphopsia. Seven years prior to her admission, she had developed a hypertensive cerebral hemorrhage in the right occipital region, when she experienced episodes consisting of flickering, a sensation of objects being nearer than they really are, and macropsia. The episodes were short-lived and disappeared spontaneously. Four months prior this admission, she developed repeated flickerings in her left visual field and sensations of objects being either nearer or farther than they really are, which were followed by headache in the right occipital region. Brain magnetic resonance imaging (MRI) revealed a focal cerebromalacia at the right temporo-occipital junction. Valproate was prescribed, which controlled the seizures. She was also found to have severe chronic renal failure, hypertension and diabetes mellitus. About 2 months later, she stopped taking valproate due to the development of epigastric discomfort and nausea. However, she did not develop seizures.

Ten days before her admission, when looking at human faces, she developed continuous visual illusions of faces, which were distorted and swollen, appearing grotesque (metamorphopsia). Four days later, repetitive flickerings occurred in her left or whole visual field, which she perceived as moving towards her. These episodes usually lasted for a few minutes,

1059–1311/$30.00 + 0.00

and recurred 15–30 times per day. On many occasions, objects in front of her appeared to be either nearer or farther away, or to rock from side to side. None of these events were accompanied by loss of consciousness.

On admission, her blood pressure was 110/70 mmHg. Neurological examination was normal except for a dense left homonymous hemianopsia, which she had not been aware of. Her metamorphopsia was restricted to the perception of human faces. Bodies, below the

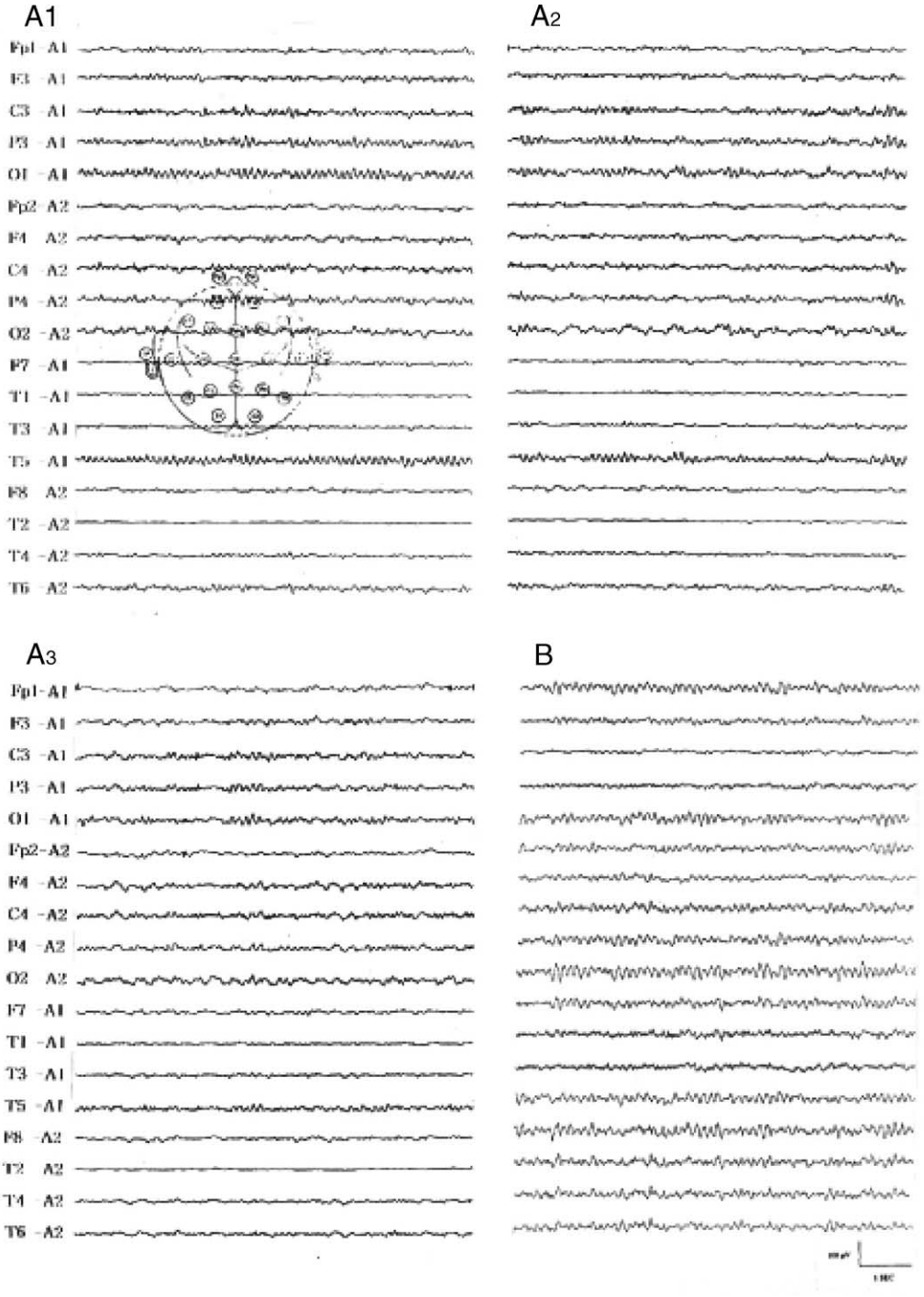

Fig. 1: (*A*) Main EEG features (eye-closing state) during the episode of ictal metamorphopsia, demonstrating poorly developed alpha activity in the right occipital region (A_1) and frequent bursts of rhythmic slow waves with various field distribution, intruding into the right occipital region ($A_{2,3}$); when the EEG demonstrated poorly developed alpha activity, Tc-ECD was injected for SPECT study. (*B*) EEG 1 day later, when metamorphopsia disappeared, showing well developed alpha activity in the right posterior cerebral region.

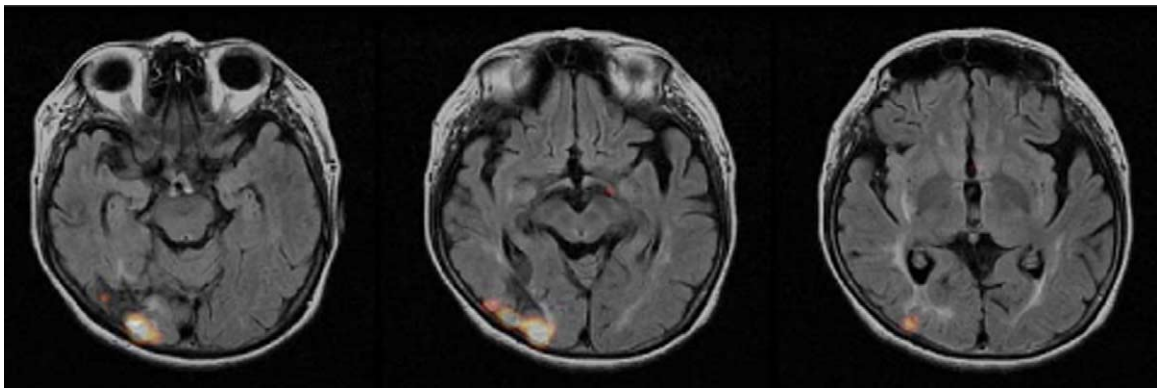

Fig. 2: The subtraction SPECT images coregistered with MR images demonstrating a region of increased blood flow in the ventrolateral aspect of the right temporo-occipital junction (middle and inferior occipital, and inferior temporal gyri) adjacent to the cerebromalacia.

level of the face, animals and other objects appeared normal. She did not have prosopagnosia. Laboratory investigation showed hemoglobin 64 g/l, hematocrit 0.19, BUN 68 mg/dl, serum creatinine 7.6 mg/dl, and blood sugar 125 mg/dl. Brain MRI revealed no interval change. She was treated with valproate. On the second day, she was in a continuous state of metamorphopsia, but developed less frequent episodes of flickering. On the third day, the frequency of the episodes of flickering was further decreased, but the metamorphopsia was persistent and continuous. The EEG, recorded for a period of 40 minutes, demonstrated poorly developed alpha activity in the right occipital region, which was frequently (>20 times) intruded by rhythmic slow waves lasting for 5–20 seconds with little evidence of evolving changes (Fig. 1A). The patient reported only continuous metamorphopsia during the EEG recording. When the EEG demonstrated poorly developed alpha activity, the technithium-99m ethyl cysteinate dimmer (Tc-ECD) was injected for SPECT study. On the fourth day, the metamorphopsia disappeared and the EEG showed well developed and symmetric posterior alpha activity (Fig. 1B).

Antiepileptic drugs treatment was discontinued due to the occurrence of adverse effects, which was followed by relapse of similar episodes. However, they had disappeared as soon as she began to have a regular peritoneal dialysis. Her left homonymous hemianopsia was still present but she could recognize crude stimuli, such as finger shaking. Interictal SPECT study was performed a month after ictal SPECT, when the patient did not report any visual symptoms except preexisting hemianopsia.

SPECT study

740 MBq of Tc-ECD was given intravenously as a radiotracer. The brain images were obtained 2 hours after tracer injection on a brain-dedicated annular crystal gamma camera (Digital Scintigraphic Incorporation, Waltham, MA) equipped with low-energy, high-resolution parallel-hole collimators[10]. Paired ictal and interictal transaxial images were normalised to the mean counts, and the interictal image was subtracted from the ictal image. Ictal hyperperfusion of subtracted SPECT was considered significant only when regional cerebral blood flow difference in each pixel of brain SPECT image between ictal and interictal states was greater than two standard deviations of the distribution of the subtraction pixel intensities. The subtraction images were coregistered to MR images with previously defined transformation parameters by using the Analyze AVW 3.0^{TM} (Biomedical Imaging Resource, Mayo Foundation, Rochester, MN, USA). The results showed a region of increased blood flow in the ventrolateral aspect of the right temporo-occipital junction (middle and inferior occipital, and inferior temporal gyri) adjacent to the cerebromalacia (Fig. 2).

The SPECT study was approved by the Ethics Committee of Severance Hospital, and the patient gave written informed consent.

DISCUSSION

Ictal metamorphopsia has been found to be associated with occipital, temporal, and parietal lesions or epilepsies, especially on the right side, and frequently accompanied by other symptoms including other types of visual illusions[2–9]. In 1949, Critchley[11] suggested that a release or distortion of the multitudinous visual associations might alter the appearance of an object and produce metamorphopsia. However, the anatomical localisation of cerebral regions responsible for the generation of ictal metamorphopsia has not been clearly established yet. This might be due to the poor

spatial resolution of EEG, discrepancy between the epileptogenic focus and ictal symptomatogenic region, and the presence of other associated symptoms. Juhász *et al.*[8] reported a patient who developed focal status epilepticus with continuous numbness in the left arm and face, accompanied by frequent visual illusions, including metamorphopsia. Brain MRI showed an increased signal in the right temporo-parietal area on T2 weighted images related to status epilepticus, corresponding to the area of increased perfusion as revealed by ictal SPECT. The localisation of the cerebral region responsible for metamorphopsia was complicated by the fact that their patient also experienced focal somatosensory symptoms, which might contribute to the MRI and SPECT findings of parietal involvement.

In the present study, the patient showed facial metamorphopsia only for a prolonged period, although preexisting hemianopsia seemed to be aggravated due to ictal or postictal effect, and elementary visual hallucinations or other types of visual illusions occurred intermittently, probably related to ictal spread. This enables us to make a more accurate determination of the anatomical area involved in the provocation of metamorphopsia. We observed an increased blood flow in the ventrolateral aspect of the right temporo-occipital junction, in the SPECT study performed during a period of continuous facial metamorphopsia. Although the scalp EEG did not demonstrate characteristic ictal discharges, it showed persisting focal abnormalities, corresponding to the area of increased perfusion, which normalised after the cessation of metamorphopsia.

Interestingly, our patients had metamorphopsia involving face only. Recent investigations using functional imaging techniques proposed that the right fusiform gyrus is primarily involved in face perception[12, 13]. Also, a small lesion involving this region could cause agnosia for faces (prosopagnosia)[14] and prosopagnosia may be associated with metamorphopsia[15]. However, the area of hyperperfusion on SPECT in our patient did not involve the fusiform gyrus, although ictal involvement of the fusiform gyrus could not be excluded completely only by SPECT finding. Our patient did not have prosopagnosia. It seems unlikely in our patient that facial metamorphopsia and prosopagnosia express the same underlying disorder. Her seizures had disappeared without medication as soon as she began to have a regular peritoneal dialysis. Uremic condition could lower seizure threshold in an excitable tissue due to old cerebral hemorrhage and provoke seizures.

In summary, a dysfunction in the visual association areas, particularly in the ventrolateral aspect of the right temporo-occipital junction, caused by seizure activity, appears to be the source of metamorphopsia.

REFERENCES

1. Kölmel, H. W. Visual illusions and hallucinations. *Baillière's Clinical Neurology* 1993; **2**: 243–264.
2. Critchley, M. Types of visual perseveration: "palinopsia" and "illusory visual spread". *Brain* 1951; **74**: 267–299.
3. Mullan, S. and Penfield, W. Illusions of comparative interpretation and emotion. *Archives of Neurology and Psychiatry* 1959; **81**: 269–284.
4. Bender, M. B., Feldman, M. and Sobin, A. J. Palinopsia. *Brain* 1968; **91**: 321–338.
5. Ida, Y., Kotorii, T. and Nakazawa, Y. A case of epilepsy with ictal metamorphopsia. *Folia Psychiatrica et Neurologica Japonica* 1980; **34**: 395–396.
6. Nass, R., Sinha, S. and Solomon, G. Epileptic facial metamorphopsia. *Brain Dev* 1985; **7**: 50–52.
7. Young, W. B., Heros, D. O., Ehrenberg, B. L. and Hedges, T. R. III Metamorphopsia and palinopsia. Association with periodic lateralized epileptiform discharges in a patient with malignant astrocytoma. *Archives of Neurology* 1989; **46**: 820–822.
8. Juhász, C., Scheidl, E. and Szirmai, I. Reversible focal MRI abnormalities due to status epilepticus. An EEG, single photon emission computed tomography, transcranial Doppler follow-up study. *Electroencephalography and Clinical Neurophysiology* 1998; **107**: 402–407.
9. Bien, C. G., Benninger, F. O., Urbach, H., Schramm, J., Kurthen, M. and Elger, C. E. Localizing value of epileptic visual auras. *Brain* 2000; **123**: 244–253.
10. Lee, J. D., Kim, H. J., Lee, B. I., Kim, O. J., Jeon, T. J. and Kim, M. J. Evaluation of ictal brain SPET using statistical parametric mapping in temporal lobe epilepsy. *European Journal of Nuclear Medicine* 2000; **27**: 1658–1665.
11. Critchley, M. Metamorphopsia of central origin. *Transactions of the Opthalmology, Society of the UK* 1949/1950; **69**: 111–121.
12. Haxby, J. V., Horwitz, B., Ungerleider, L. G., Maisog, J. M., Pietrini, P. and Grady, C. L. The functional organization of human extrastriate cortex: a PET-rCBF study of selective attention to faces and locations. *Journal of Neuroscience* 1994; **14**: 6336–6353.
13. Kanwisher, N., McDermott, J. and Chun, M. M. The fusiform face area: a module in human extrastriate cortex specialized for face perception. *Journal of Neuroscience* 1997; **17**: 4302–4311.
14. Wada, Y. and Yamamoto, T. Selective impairment of facial recognition due to a haematoma restricted to the right fusiform and lateral occipital region. *Journal of Neurology, Neurosurgery, and Psychiatry* 2001; **71**: 254–257.
15. Hecaen, H. and Angelergues, R. Agnosia for faces (prosopagnosia). *Archives of Neurology* 1962; **7**: 24–32.