

Table 1 Blood test at admission.

Peripheral blood		Biochemical study				Urine analysis
Haemoglobin	11.6 g/dL	Total protein	5 g/dL	Lactate dehydrogenase	418 IU/L	Protein++
Haematocrit	56.2%	Albumin	2.7 g/dL	GOT	9 IU/L	
MCV	85.0 fL	Urea	12 mg/dL	GPT	13 IU/L	
				Glucose	95 mg/dL	
Platelets	$10 \times 10^4/\mu\text{L}$	Creatinine	0.9 mg/dL			
ESR	20 mm/h	Uric acid	6.7 mg/dL			
Leukocytes	14 380/ μL	Alkaline phosphatase	110 IU/L			

ESR: erythrocyte sedimentation rate; GOT: glutamic oxaloacetic transaminase; GPT: glutamate-pyruvate transaminase; MCV: mean corpuscular volume.

nervorum, causing transient ischaemia that would lead to nerve palsy.⁶ In our patient, who had no history suggesting an underlying systemic disease, presenting preeclampsia and no fever, intracranial hypertension, or underlying intracranial lesion on MR images, together with the complete resolution of symptoms postpartum, suggest preeclampsia-related sixth nerve palsy.

References

- Lambert G, Brichant JF, Hartstein G, Bonhomme V, Dewandre PY. Preeclampsia: an update. *Acta Anaesthesiol Belg*. 2014;65:137–49.
- Thamban S, Nama V, Sharma R, Kollipara PJ. Abducens nerve palsy complicating pregnancy. *J Obstet Gynaecol*. 2006;26:811–2.
- Barry-Kinsella C, Milner M, McCarthy N, Walshe J. Sixth nerve palsy: an unusual manifestation of preeclampsia. *Obstet Gynecol*. 1994;83:849–51.
- Thurtell MJ, Sharp KL, Spies JM, Halmagyi GM. Isolated sixth cranial nerve palsy in preeclampsia. *J Neuro-Ophthalmol*. 2006;26:296–8.
- Park CM, Kim SY. Abducens nerve palsy in preeclampsia after delivery: an unusual case report. *J Obstet Gynaecol Res*. 2007;33:543–5.
- Vallejo-Vaz AJ, Stiefel P, Alfaro V, Miranda ML. Isolated abducens nerve palsy in preeclampsia and hypertension in pregnancy. *Hypertens Res*. 2013;36:834–5.

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Usefulness of brain SPECT imaging in the study of recurrent hypersomnia: Kleine-Levin syndrome[☆]

Utilidad del SPECT cerebral en el estudio de la hipersomnia recurrente: síndrome de Kleine-Levin

Dear Editor:

Kleine-Levin syndrome is an infrequent neuropsychiatric disorder that manifests with recurrent, self-limited episodes of



hypersomnia, normally accompanied by behavioural alterations (hypersexuality, irritability, and aggressiveness) and such cognitive alterations as confusion and hallucinations. It presents in the second decade of life, with a higher incidence in young men (4:1). Episodes last from 1 to 2 weeks, with complete remission of symptoms. Episodes may occur several times per year, separated by asymptomatic periods.¹

We present the case of a 17-year-old patient who, after symptoms of increased bowel activity, nausea, and low-grade fever, began to present a marked increase in somnolence, sleeping up to 20 hours per day; he was admitted to the internal medicine department. During his hospital stay, he underwent a blood test, head CT scan, brain MRI scan, chest and abdomen CT scan, EEG, polysomnography, and waking EEG; none revealed significant findings. The multiple sleep latency test revealed a mean sleep latency above 10 min (14 min) and did not show REM sleep in any sleep attempt. The CSF study showed a mildly elevated protein level. Considering a possible emotional origin, the psychiatry department assessed the patient on 3 occasions, with no

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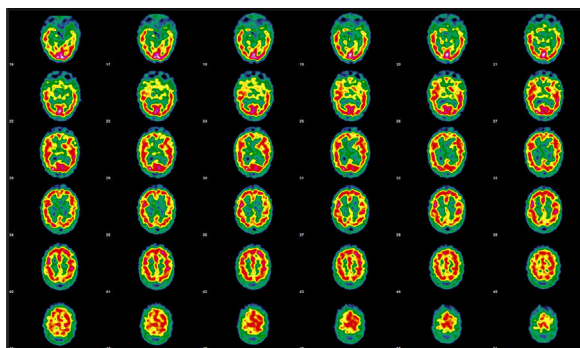


Figure 1 Initial brain perfusion SPECT performed after the administration of 925 MBq of ^{99m}Tc -HMPAO. Representative transaxial tomographic slices are shown. Qualitative assessment detected irregular cortical function in both hemispheres, with more marked hypoactivity in the anterior and orbitofrontal regions, and in the right mesial temporal region. The left hemisphere showed defects in the orbitofrontal, temporobasilar, and mesial regions. Asymmetry is observed in the basal nuclei, with decreased activity on the right side; the cerebellum is unaffected.

abnormal findings and no dysfunction in the school or family setting.

Since the patient did not meet criteria for narcolepsy or idiopathic hypersomnia, and considering the history of gastrointestinal involvement and high CSF protein level, he was discharged with an initial diagnosis of post-infectious encephalitis, not ruling out other secondary origins of the symptoms. We started empiric outpatient treatment with prednisone on a tapering schedule, starting at 70 mg and decreasing by 10 mg/week.

Several days later, he was admitted to the neurology department due to excessive hours of daytime sleep (up to 20 hours per day). Considering his tendency to sleep, hyperphagia, and behavioural alterations during wakefulness, and the presence of a gastrointestinal infection as the trigger factor for the symptoms, he was diagnosed with recurrent encephalitis. A brain perfusion SPECT scan revealed a cortical activity pattern not corresponding with the patient's age; the alterations observed were compatible with clinical suspicion of Kleine-Levin syndrome (Fig. 1).

After a significant clinical improvement, the patient was discharged with treatment with modafinil (100 mg/day); on some days he continued to experience mild excessive daytime sleepiness, although to a lesser degree than during admission.

Two months later he once more developed symptoms of hypersomnia, which improved when the dose of modafinil was increased (200 mg/day).

During the 2-year follow-up, the patient presented a further 3 episodes of similar characteristics. A SPECT study performed 2 years after symptom onset revealed normal uptake in the frontal area and persisting alterations in the temporal region (Fig. 2).

Kleine-Levin syndrome is a rare disease whose diagnosis is based on clinical symptoms.² To date, no objective complementary tests able to confirm its diagnosis have been developed. Our patient presented successive episodes of

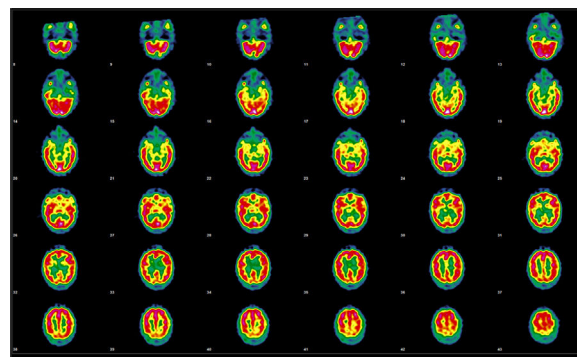


Figure 2 Follow-up brain perfusion SPECT performed after the administration of 925 MBq of ^{99m}Tc -HMPAO, 20 months after the initial study. Representative transaxial tomographic slices are shown. Irregular cortical function is observed in both hemispheres, with defects of higher intensity in the orbitofrontal and temporal regions and small, diffusely distributed irregularities in the remaining areas of both hemispheres; no alterations are observed in the basal nuclei or cerebellum. Compared with the previous studies, an improvement is observed in the defects in the orbitofrontal and temporal regions, with normal uptake in the anterior frontal region.

self-limited hypersomnia, with no significant findings in any of the complementary tests performed during admission.

Aetiology of Kleine-Levin syndrome remains unknown and clinical symptoms suggest the possibility of a hypothalamic-pituitary dysfunction caused by such trigger factors as head trauma, aseptic meningoencephalitis, or Prader-Willi syndrome. Onset during adolescence, symptom recurrence, and infections as frequent trigger factors suggest an autoimmune aetiology; furthermore, an association between Kleine-Levin syndrome and *HLA-DR1* has been observed.³ The trigger factor (gastroenteritis) and the age of onset in our patient coincide with descriptions in the literature.

Differential diagnosis should include recurrent major depression, bipolar depression, and recurrent neurotic or organic hypersomnias, which are mainly caused by intraventricular tumours in the third or fourth ventricle.⁴

To rule out a personality disorder as the origin of the symptoms, it is essential to perform a psychological study during the clinical episode and the asymptomatic period. No psychiatric disorder was observed in our patient.

Functional imaging has been shown to be useful for studying the pathophysiology of the condition.⁵

During the symptomatic phase, SPECT typically reveal hypoperfusion in the uni- or bilateral temporal region, frontotemporal region, and basal ganglia.⁶ Such dysfunction may constitute a diaschisis phenomenon, favoured by frontal hypoactivation derived from diencephalic dysfunction. In our case, SPECT revealed hypoperfusion in the right basal ganglia, both temporal regions, and the right frontotemporal region.

In interictal periods, results from the studies performed suggest that SPECT findings usually normalise, but temporal alterations persist even after clinical alterations have disappeared. Findings from the second SPECT study were pathological, although the patient presented no symptoms; this is consistent with the results reported by Gabrieli et al.³

We conclude that brain perfusion SPECT should be considered during episodes of recurrent hypersomnia in adolescents to rule out a possible associated Kleine-Levin syndrome.

References

1. Pedrós Roselló A, Hoyo Rodrigo B, López Gomáriz E. Síndrome de Kleine-Levin: análisis clínico comparativo. A propósito de dos casos. *Psiquiatr Biol*. 2011;18:124–6.
2. Huang YS, Guilleminault C, Kao PF, Liu FY. SPECT findings in the Kleine-Levin syndrome. *Sleep*. 2005;28:955–60.
3. Gabrieli RDE. Cognitive neuroscience of human memory. *Annu Rev Psychol*. 1998;49:87–115.
4. Peraíta-Adrados R. Síndrome de Kleine-Levin: aportación diagnóstica de la SPECT cerebral. *Rev Neurol*. 2003;36:559.
5. Arias M, Crespo-Iglesias JM, Pérez J, Requena-Caballero I, Sesar-Ignacio A, Peleteiro-Fernández M. Síndrome de Kleine-Levin: aportación diagnóstica de la SPECT cerebral. *Rev Neurol*. 2002;35:531–3.
6. Vigren P, Engström M, Landtblom AM. SPECT in the Kleine-Levin syndrome, a possible diagnostic and prognostic aid? *Neurology*. 2014;5:178.

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Relationship between Virchow–Robin spaces and Alzheimer disease: a case report[☆]

Relación de los espacios de Virchow–Robin con la enfermedad de Alzheimer: a propósito de un caso

Dear Editor:

Efforts have been made in recent years to evaluate whether the presence of dilated Virchow–Robin spaces (dVRS) on MRI studies, which had previously been considered incidental, may play a role in such diseases as stroke, multiple sclerosis, cerebral amyloid angiopathy, and Alzheimer disease (AD), and whether dVRS cause any symptoms.

We present the case of a 74-year-old woman with no relevant clinical history, who was assessed due to a 3-year history of progressive cortical cognitive decline with anterograde amnesia, aphasia, agnosia, apraxia, and loss of initiative to perform instrumental activities of daily living, scoring 23/30 on the Mini-Mental State Examination (Folstein, 1975), and presenting mild parkinsonism with reduced arm swing during gait and resting tremor of the right hand. No falls or visual hallucinations were reported. A blood test ruled out treatable causes, and a brain CT scan revealed subcortical hypodense lesions in both hemispheres (Fig. 1). A brain MRI scan showed dilatation of



the Virchow–Robin spaces, together with cortical fronto-temporo-parietal atrophy (Fig. 2). In view of these findings, we ruled out hyperhomocysteinaemia and vasculitis and diagnosed probable AD with parkinsonism of vascular origin.

Virchow–Robin spaces are perivascular, interstitial fluid-filled spaces surrounding cerebral vessels passing from the subarachnoid space through the brain parenchyma; they are considered a drainage route for residual metabolites of parenchymatous activity to the subarachnoid space for removal. They are typically located on the lenticulostriate arteries in basal ganglia (type I), perforating arteries of the corona radiata and semioval centres (type II), and brainstem (type III).¹ They are considered to be dilated when their diameter is equal to or larger than 1 mm. Larger spaces may be detected on CT studies, but MRI provides more



Figure 1 Brain computed tomography scan showing hypodense lesions in the subcortical white matter in both frontal lobes.

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