Neurologist-in-training

The aim of this section is to prepare the neurologist-in-training for the FMH examination, to confront her or him with specific problems of everyday neurological practice and to give him or her updates on recent controversies in clinical neurology.

Contributions and correspondence to Andrea O. Rossetti, Service de Neurologie BH-07, CHUV, CH-1011 Lausanne, andrea.rossetti@chuv.ch

Neurological MCQ

A. Carota

CHUV, Lausanne

1 Which lesion may not induce a Horner's syndrome?

A C7 root **B** C8 root **C** Th1 root **D** Common carotid artery dissection

2 Please indicate the wrong statement:

A Irritation of the C3 root may cause a painful, burning, red ear.
B Damage of the spinal cord at C5 level can induce an inverted ipsilateral styloradial deep tendon reflex.
C Diaphragmatic paresis excludes a lesion at C3–C5 level or root.
D A C5–C6 lesion may elicit an impairment of the M. pectoralis major deep tendon reflex.

3 Which of the following clinical situations best represents a pure L5 root lesion?

A Touch and pinprick sensibility impairment over knee and medial tibial region
B Abolition of the Achilles tendon reflex
C Paresis of the anterior tibialis muscle
D Paresis of the hip abductors
E Tibialis posterior tendon hyperreflexia

4 Which of the following sentences is false?

A Lower extremity radicular pain often irradiates to the foot as well as to the back.
B A patient with clear L5 radicular sensory-motor symptoms and a normal dorso-lumbar MRI should be reassured and does not need myelography.
C A clinical deficit with involvement of L1–L3 roots suggests rather a medical than a mechanical aetiology, such as diabetes, neoplastic, vascular or other causes.
D Characteristically, lower extremity radicular pain often shows a changing anatomical distribution along the corresponding dermatome over time.

5 Which statement is incorrect?

A Dorsal root lesions result in hypoaesthesia often confined to a specific dermatome.
B Lack of sensory loss does not formally exclude the possibility of a lesion affecting a single dorsal root.
C After dorsal root lesions distribution of hyperalgia is generally larger than tactile hypoaesthesia.
D Burning pain after dorsal root lesion is mediated by large myelinated A-alpha fibres.

(For correct answers, see page 99)

Neuroimaging

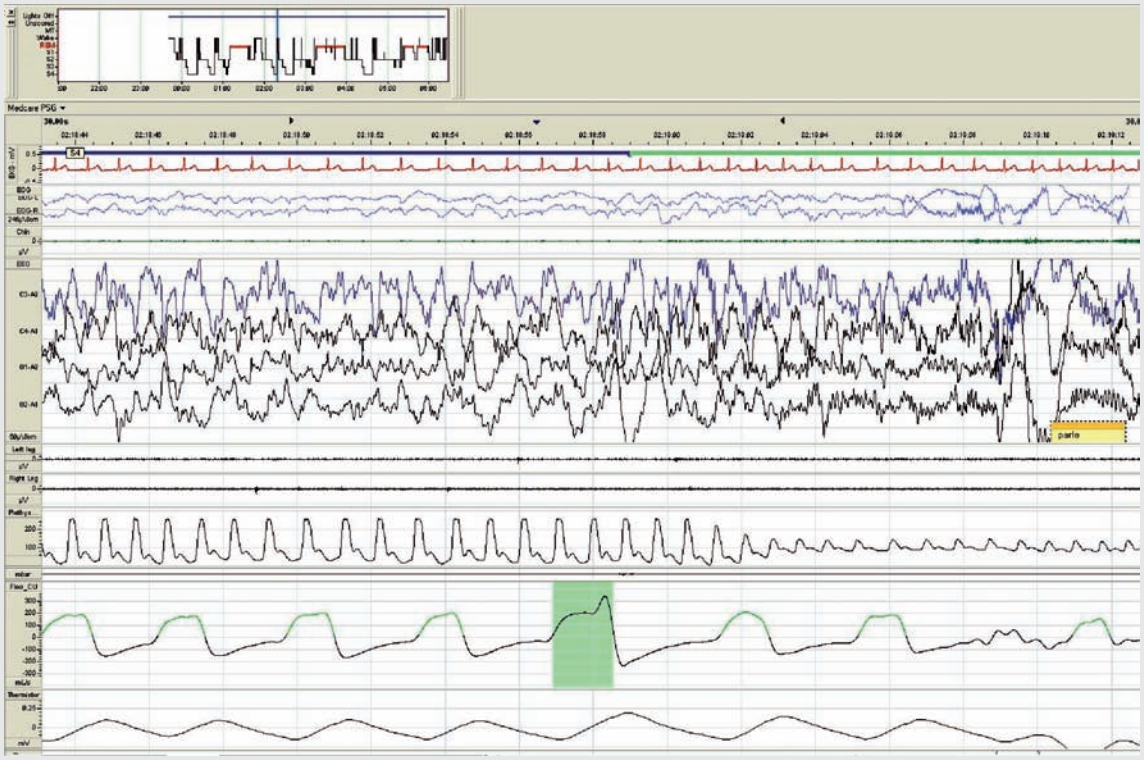
■ **A. O. Rossetti**

CHUV, Lausanne

A 29-year-old man is evaluated at the sleep clinic for recurrent episodes of sleep talking or walking in his apartment. These began at the age of 4, but increased in frequency, occurring now several times a week (one episode per night), rather in the first half of the night. He has only fragmentary memory of these episodes. The patient also complains about increased snoring, especially after having gained weight a few years previously. His brother had sleep talking in his childhood.

Below you find a 30-s screenshot of part of his polysomnographic recording during a typical arousal. On the top right you see the hypnogram, then the EKG line, the electro-oculogram, the chin EMG, the EEG, the legs' EMG, the pulse amplitude (plethysmograph), the nasal airflow, and finally the thermistor signal (placed in front of the mouth). On the right low corner, the yellow box marks talking activity, according to the technologist.

What are your thoughts regarding the nature of these phenomena? Are they epileptic or not?



(For correct answers, see page 99)

Read for you

A. O. Rossetti

CHUV, Lausanne

IVIG in myasthenia gravis and the generalisability of randomised trials

Despite its frequent prescription in common practice, intravenous immunoglobulin treatment (IVIG) for rapid worsening myasthenia gravis has been found in a recent Cochrane database review to lack scientific evidence for its use [1]. In particular, only 5 randomised trials were identified (2 assessing IVIG vs plasma exchange, and 1 each IVIG vs placebo, IVIG vs steroids and 2 different dosages of IG). Therefore, a randomised double-blind clinical trial was performed by a Canadian group, investigating the use of 2 g/kg IVIG (in 2 fractioned doses) versus placebo [2].

The primary outcome was defined as the difference in the change of the Quantitative Myastenia Gravis score (QMG) at day 14. The relatively modest sample size of 52 patients was calculated relying on an older trial using cyclosporine [3, 4].

Formally, it is potentially misleading to proceed in that way since the extent of clinical change might well be different for each specific substance. Nevertheless, the authors were lucky enough to find a just significant result for the primary outcome, with a P of 0.047; whereas the result at 28 days gave a P of 0.055. Of note, a subgroup analysis demonstrated that patients with moderate to severe disease responded better than subjects with mild disease and that patients having undergone thymectomy were also more likely to improve. Now that this trial is available, in the era of "evidence-based medicine", we are informed that IVIG are more effective than placebo for myasthenia gravis.

However, something that is often forgotten, when analysing randomised trials' results, is the *generalisability* of the studies. In fact, in the exclusion criteria of this study [2] we find, among others, respiratory distress requiring ICU admission, severe swallowing difficulty and recent change of steroid dosage. Therefore, this category of patients, who are often encountered in clinical practice and for whom IVIG might represent a therapeutic option (especially when plasma exchange is not feasible), does not have any direct information from this trial regarding the usefulness of IVIG. On the other hand, it would have been difficult from an ethical point of view to recruit patients with severe myasthenia gravis in a trial using placebo. In conclusion, some evidence is available, but, for the moment, this is unlikely to change clinical practice and, what is more important, patients' prognosis.

References

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3 Tindall RS, Phillips JT, Rollins JA, Wells L, Hall K. A clinical therapeutic trial of cyclosporine in myasthenia gravis. Ann N Y Acad Sci. 1993;681:539–51.

4 Barohn RJ, McIntire D, Herbelin L, Wolfe GI, Nations S, Bryan WW. Reliability testing of the quantitative myasthenia gravis score. Ann N Y Acad Sci. 1998;841:769–72.

Answers to MCQ

1 A **2** C **3** D **4** B **5** D

Answers to neuroimaging

The patient's history is interesting in several points. Occurrence of epileptic paroxysms in sleep for 20 years without a diagnosis of epilepsy is rather unusual, as opposed to parasomnias. A consistent percentage of children exhibit at least transitorily non-REM parasomnias, such as sleep terrors, sleep wandering, confusional arousals or sleep talking, mostly without remembering the episodes. There is often a family history as here.

Occurrence – typically a few times per night (classically only once) – of the episodes in the first night's half corresponds to the physiologic clustering of slow-wave sleep (SWS). This is different in epilepsy, where seizures often arise from stage-2 sleep (distributed over the whole night) and may occur in clusters, especially when originating from the frontal lobe. More important, seizures show a consistent tendency to occur with a stereotypical semiology, as opposed to parasomnias.

Prevalence decreases dramatically with age, since less than 5% of the adult population has this kind of parasomnnias. Interestingly, adults sometimes recall parts of the episodes as "flashbacks", as in this case.

Looking at the image, the hypnogram shows arousals directly for stage-3 or -4 sleep (SWS), which represents a typical finding. Furthermore, his arousal, on the EEG tracing, appears peculiar, with persistent delta and superimposed alpha. This is characteristic of non-REM parasomnias, which may be seen as incomplete arousals from deep sleep. The fact that he was heard talking (unintelligibly) reinforces the diagnosis.

In other parts of the recording a moderate sleep apnoea syndrome was observed, which led to several microarousals. These, fragmenting the patient's sleep, are likely to induce an increased SWS pressure, thereby leading to the increase in parasomnia frequency that the subject experienced. The first therapeutic step, in this case, would not be clonazepam administration (that might worsen SAS), but rather prescription of a mandibular advancement device or nocturnal cPAP in order to normalise sleep fragmentation.