CASE REPORT

Stimulus Sensitive Constant Micro Macro Square-Wave Jerks in a Comatose Patient

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ABSTRACT

Saccades are generally thought of as being cortically generated and not seen in comatose patients. We describe a patient, comatose secondary to a large intracerebral hemorrhage, who developed constant rhythmic small amplitude square-wave jerks with an intersaccadic interval of 130 ms. Despite the patient's comatose state the eye movements would stop transiently in response to auditory or tactile stimulation and then quickly regain their previous amplitude. The case suggests that the presence of repetitive saccades in the form of square-wave jerks does not indicate consciousness.

Keywords: Coma, eye movements, square-wave jerks

INTRODUCTION

It is generally considered a clinical axiom that, when examining a patient with a decreased level of consciousness, the presence of spontaneous saccades indicates some level of cortical function, if not consciousness.¹ Although rapid movements of different types have been described in comatose patients, we have been unable to find a report of rhythmically repetitive square-wave jerks in a comatose patient.

CASE REPORT

SH is a 52-year-old male who was found unconscious on the floor of his apartment after being last seen at work the day of admission. On arrival to the emergency room he was unresponsive, requiring intubation, and mechanical ventilation. Neurosurgical exam documented nonreactive pupils measuring 2.5 mm OD and 4 mm OS, a left cranial nerve III paresis, and decerebrate posturing to noxious stimuli consistent with a transtentorial herniation syndrome. A CT revealed a large left thalamic haemorrhage with intraventricular extension and hydrocephalus (Figure 1). Additionally, he was

found to be in acute renal failure with a positive urine drug screen for cocaine.

Bilateral ventriculostomy tubes were placed, and initial intracranial pressure (ICP) measurements were 60 mmHg. He was started on mannitol 25 gm every six hours for intracranial hypertension and Dilantin 300 mg daily for seizure prophylaxis. After the ventriculostomies his condition stabilized. The patient's oculomotor paresis and anisocoria resolved, although his pupils remained nonreactive. He responded only to noxious stimuli.

Two days later his pupils were nonreactive and miotic at 2.5 mm on the right and 2.0 mm on the left. Oculo-cephalic and corneal reflexes were judged to be absent, but gag and spontaneous respirations were present. ICP measurements were consistently 10–20 mmHg. Repeat CT revealed that the blood volume had decreased in the anterior horns of the lateral ventricles but was unchanged in the IIIrd and IVth ventricles. The hydrocephalus was also unchanged.

On the fourth day of admission he remained unresponsive, but continuous horizontal eye-movements were noted prompting neuro-ophthalmologic consultation. Neuro-ophthalmologic examination confirmed nonreactive miotic pupils, but vestibuloocular

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FIGURE 1. CT: representative slices of the initial CT scan showing a large thalamic haemorrhage with intraventricular extension.

reflex (VOR) was minimally present bilaterally in response to horizontal head movements. The eye movements were felt to be continuous square-wave jerks. Formal eye movement recordings confirmed square-wave jerks occurring constantly in a rhythmic pattern at 3.1 Hz. The average intersaccadic interval (the time from the end of one saccade to the beginning of the next) was 130 ms with a standard deviation of 17 ms. The amplitude range was 1.5–3° (Figure 2a). Analysis showed that the saccades fit a normal main sequence with the exception of abducting saccades in the right eye, which were slowed indicating a mild right sixth nerve palsy. The eye movements were also demonstrated to stop transiently after either auditory or tactile stimuli (Figure 2b and c). Bilaterally sluggish corneal reflexes were demonstrated. The next morning the patient was thought clinically to have a small amplitude, constant left beating nystagmus. That afternoon no spontaneous movements were noted but an eye movement recording documented an intermittent right beating nystagmus with an amplitude less than 1°. An EEG showed generalized severe slowing with no evidence of epileptic activity. There were no pupillary reactions but a horizontal VOR could still be elicited. He remained otherwise clinically unchanged over his subsequent hospital stay. On hospital day 9, he expired after supportive care was discontinued at the request of his family.

DISCUSSION

During the examination of a patient with a decrease level of alertness, the presence of saccades is typically considered a sign that the patient, if not conscious, has at least some cortical function.¹ Although this is

clearly true for voluntary saccades known to be generated by the cortex, it is also most often also true for saccades which are part of nystagmus induced by brainstem reflexes. This is supported by a study done by Leigh *et al* that looked at eye movements induced by head rotations in comatose patients and those in a vegetative state. Quick phases were not seen in coma, but were present in vegetative subjects.² Another study using caloric testing found no nystagmus in 81 comatose patients.³

Nonetheless, several different types of eye movements that include saccades have been described in comatose patients. These include the quick phases of nystagmus, various bobbing syndromes, saccadic ping pong gaze, as well as the saccades of opsoclonus.^{4–7} To our knowledge, no comatose patients have been reported with square-wave jerks.

Spontaneous saccadic eye movements, no matter what the state of consciousness, can be divided into two basic categories. By definition those with saccades separated by an intersaccadic interval are called square-wave jerks. These are distinguished from double saccadic pulses, ocular flutter, and opsoclonus, all of which have back to back saccades with no intersaccadic interval.

Historically various types of square-wave jerks have been described. “Standard” square-wave jerks (SSWJ) are intermittent and nonrhythmic. They are characterized by a normal saccade with an amplitude typically less than 3°. After 200–250 ms, there is an equal but opposite saccade returning the eyes to the original position. These are a common and nonlocalizing abnormality. They can be seen in normal subjects as well as generalized disorders as varied as schizophrenia, progressive supranuclear palsy, and Alzheimer’s disease. They are also well described in focal lesions involving both supra and

FIGURE 2. Eye movement recording: all segments from a several minute eye movement recording showing continuous alternating saccades in the form of square-wave jerks. Rightward movements are displayed as upward movements while movements to the left are displayed as downward movements. (a) The clear periodicity. The square-wave jerks show dynamic overshoots and evidence of a leaky integrator more on left gaze than right. The other two figures show both a position and velocity trace of the right eye and demonstrate the transient cessation of repetitive saccadic movements with sensory stimulation. (b) A 0.5 s cessation after an auditory stimulus and (c) a slightly longer cessation after a painful stimulus.

infratentorial structures.^{8–11} SSWJ typically are very irregular even when nearly constant.

Two other eye movement abnormalities of similar wave form, macro square-wave jerks and macrosaccadic oscillations, have been described, both of which are most commonly seen in patients with cerebellar disease. Macro square-wave jerks are clearly a type of square-wave jerk. Macrosaccadic oscillations look like a continuous series of SSWJ that oscillate about fixation.^{12,13} Macrosaccadic oscillations start with a small saccade off of fixation followed by a series of saccades oscillating about fixation with an increasing then decreasing amplitude. The typical intersaccadic interval is 200–250 ms. Amplitudes can be as large as 40° about fixation. Macro square-wave jerks can be distinguished from SSWJ by their larger amplitude (typically greater than 10°), as well as their much shorter intersaccadic intervals, ranging from 50 to 150 ms.

Although not incorporated into the definition, it is striking that in many of the reported cases, macro square-wave jerks occur not only as isolated square waves but as a longer string of repetitive rhythmic alternating saccades. A cursory look at smaller

sections of these repetitive macro square-wave jerks would appear quite similar to macrosaccadic oscillations. An overview of the recording would show that the rhythmic macro square-wave jerks go to and from fixation rather than oscillating about fixation as in macrosaccadic oscillations. The frequency of square-wave jerks during bursts of macro square-wave jerks and macrosaccadic oscillations are both consistently 2–3 Hz, similar to our patient.^{12–15}

As the above movements are defined by their relationship to fixation, in our comatose patient it seems impossible to definitively identify the movements in our patient as either macro square-wave jerks or macrosaccadic oscillations. The intersaccadic latency of 130 ms is typical of macro square-wave jerks and quite different from those in macrosaccadic oscillations. The eye movements in our patient were identical to the reported rhythmic series of macro square-wave jerks with the exception of the amplitude of the movements, suggesting that a suitable name might be micro macro square-wave jerks. It is not clear from the literature whether or not the most important characteristic of macro square-wave jerks

that allows diagnosing cerebellar dysfunction is the amplitude or the short intersaccadic latency. It has been proposed that the aetiology of pathologic saccadic intrusions is secondary to the lack of inhibition of bursts cells.¹⁶ The role of the superior colliculus in controlling saccades has been well documented in animal studies. In particular, these studies suggest a prominent effect of the superior colliculus in controlling the latency of saccades. In animals with muscimol injections into the superior colliculus, saccades to a target have an increased number of saccades with shorter latencies (express saccades).^{17–19} It is also striking that macro square-wave jerks have been noted in a monkey with superior collicular lesions as well as following muscimol injections into the mesencephalic reticular formation, which contains cells and connections to the superior colliculus.^{20,21} The short latency of the square-wave jerks in our patient suggest a similar pathophysiology. Unfortunately, even though the superior colliculi seem to be compressed by the haemorrhage as shown in Figure 1, the pathology in our patient was diffuse. Thus our case cannot be considered definitive in supporting this localization.

An additional striking feature in our patient was the observation that the pattern of rhythmic to and fro saccades could be temporarily interrupted by severe nail bed pressure as well as loud auditory stimuli. This made us question the assumption that the patient was comatose. However, other than the saccadic movements there was no other indication clinically or by EEG that the patient was awake. Despite the clinical quadriplegia, there was no paralysis of horizontal eye movements and no retention of voluntary vertical eye movements ruling out the possibility of being locked-in from an unsuspected pontine lesion that did not show up on CT. The volume of blood on the CT scan would also be most commonly associated with coma rather than a locked in state. The pathophysiology underlying this finding is unclear. Another comatose patient with repetitive eye movements has also been reported to have the movements suppressed by painful stimuli. The eye movements in that patient were in the form of short cycle periodic alternating (ping pong) gaze.²² That same report refers to a previously reported, awake patient with similar eye movements suppressed by painful stimuli.

Clinically it remains important to consider that a patient who appears comatose may be either locked in or minimally conscious rather than truly comatose. Spontaneous saccades in the form of isolated saccades or simple square jerks are suggestive of this. This case emphasizes that one must be alert to the possibility that saccades as part of other known abnormal eye movement patterns can be seen in patients who are not awake.

DECLARATION OF INTEREST

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of this article.

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