Periodic eye opening and swallowing movements associated with post-anoxic burst-suppression EEG pattern

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ABSTRACT – We report a video-electroencephalogram study of a 50-year-old woman who developed a clinical picture consisting of stereotyped periodic eye opening followed by eye closing, with or without swallowing movements, after a prolonged cardiopulmonary arrest. These movements were associated with a burst-suppression pattern on the electroencephalogram. [Published with video sequences]

Key words: periodic myoclonus, post-hypoxic myoclonus, burst-suppression pattern, seesaw phenomenon, swallowing movements

This report describes a video-electroencephalogram (v-EEG) study of a patient who developed a clinical picture consisting of stereotyped periodic eye opening followed by eye closing, with or without swallowing movements, after a prolonged cardiopulmonary arrest. We have found only a few similar cases describing this peculiar clinical phenomenon, which was always associated with a burst-suppression pattern on the EEG (Wolf, 1977; McCarty and Marshall, 1981; Reeves et al. 1997). Although its pathophysiology remains unresolved, some authors have considered this unusual condition as a periodic, post-hypoxic myoclonus (PHM) (Wolf, 1977).

Case study

A 50-year-old woman was admitted to our hospital after resuscitation from acute cardiopulmonary arrest. Two hours before her admission, she had experienced headache and two consecutive generalized tonic-clonic seizures. She had experienced head trauma causing cerebral concussion at the age of 24 years, but without residual neurological deficits. On neurological examination, she was comatose. She remained unresponsive to any external stimuli. The pupils were miotic and slowly reactive. Brainstem reflexes were preserved and plantar responses were indifferent. A computed tomography scan of the brain showed a vascular malformation localized in the left frontotemporal region, with evidence of discrete subarachnoid hemorrhage. At this time, antiepileptic treatment with phenytoin (100 mg/8 h) and sedation with propofol (4 mg/kg/24 h) were instituted and seizures did not recur. Dur-
ing the following hours, an attempt by stopping sedation was carried out; however, the occurrence of repetitive episodes of eye opening, at times accompanied by swallowing movements and proximal jerks in both shoulders, led us to restart propofol therapy. On day 2, a v-EEG, after briefly stopping the propofol infusion, was carried out in the intensive care unit. The EEG revealed the presence of periodic, fairly regularly every 4-5 seconds, generalized burst of slow waves intermixed with prominent spikes, polyspikes and sharp-waves, lasting from 2 to 3 seconds, interrupted by flat periods constituting a periodic burst-suppression pattern (figure 1). Interestingly, each periodic burst was always accompanied by a phenomenon of eye opening (see video sequence). In addition, swallowing movements were also seen but occurring with distinct regularity (every 5, 10 or 15 seconds). Most frequently, each two episodes of eye opening were followed by one episode of the swallowing movements. The myoclonus could not be modified by manual maneuvers. Rhythmicity showed a 1:8 ratio to heart rate. Sporadic eyelid and generalized jerks were observed. In view of these findings, a diagnosis of periodic PHM was proposed. These clinical and electroencephalographic features were abolished with propofol infusion. Forty-eight hours later, the clinical state of the patient deteriorated. At that time, the oculocephalic, corneal and ciliospinal reflexes were all lost and myoclonus ceased. A second EEG revealed electrocerebral inactivity. Finally, she died on day 6 after hospital admission.

Discussion

Acute PHM usually occurs early after an anoxic episode and is characterized by severe, generalized myoclonic jerks in deeply comatose patients (Venkatesan and Frucht, 2006). Nevertheless, focal or multifocal intermittent jerks affecting only facial muscles, particularly the eyes and mouth may also occur (Jumao-as and Brenner, 1990). The jerks may be frequent, but only on rare occasions have a periodic pattern (Wolf, 1977). Although the exact pathophysiology of the acute PHM remains unresolved, it is postulated that the myoclonus arises from a brainstem generator because the cortex is severely damaged in these patients and may not be capable of generating synchronous cerebral activity (Hallet, 2000). The occurrence of periodicity seems to suggest the activation of a pacemaker responsible for this clinical phenomenon.
Periodic PHM is a rare and fascinating condition, which is generally associated with poor outcome. Wolf (1977) described five patients suffering from post-anoxic coma that developed a picture of periodic and stereotyped myoclonus. This author suggested a brainstem origin for this condition because all his patients had a lower pontine/upper oblongata syndrome. Interestingly, the patient HB of Wolf’s paper experienced a seesaw automatism rather similar to that described in the case reported here. This author observed that the slow jerks of the levator palpebrae superiori muscles occurred synchronously with EEG discharges, coinciding with the relaxation of both corrugator glabellae muscles. He interpreted these findings as a reciprocal inhibition phenomenon.

In this case, in addition to the continuous seesaw automatism as described by Wolf (1977), the patient experienced complex movements of swallowing immediately after the eye opening. Most likely, the movements described represent periodic synchronous activation of bulbar muscles. However, while all burst of generalized epileptiform discharges were associated with the seesaw alteration, the swallowing movements appeared only with some. This finding seems to suggest that different pathways were activated by a same generator. Less likely, it may indicate that two different pacemakers were responsible for the clinical manifestations.

Unfortunately, we did not employ more sophisticated methods such as surface electromyogram to examine the clinical manifestations of our patient. As Wolf concluded, we think that a brainstem origin seems most likely for the movements described here. However, the existence of burst of generalized polyspikes appears to suggest a certain participation of the cortex, possibly originating discharges diffusely as a consequence of a reticular activation. It is probably that damaged cortex may not be capable of generating any centrifugal activity, but some centripetal, reticulo-thalamus-cortical connections could remain functionally active for several hours. In addition, its universal association with a burst-suppression electroencephalographic pattern seems to support this hypothesis because this EEG finding has been attributed, in certain cases, to recurrent activation of thalamic networks (Chatrian and Turella, 2003). Interestingly, a tonic opening of the eye often occurs during absence seizures, suggesting equally a possible brainstem involvement (Bogacz et al. 2000). Although the genuine epileptic nature of these clinical manifestations may be debatable, the association of focal or generalized jerks or even generalized tonic-clonic seizures (as in our observation) appears to suggest that the use of antiepileptic drugs is indicated.

The characteristics of the clinical features described here deserve more discussion. Reeves and colleagues (1997) reviewed the variety of movements, in 12 comatose patients, associated with a burst-suppression EEG pattern. Nine patients had had a confirmed or presumed cardiac or respiratory arrest. Complex movements including swallowing or chewing were reported and the authors accepted that implementation of motor programs at the brainstem or subcortical levels could have played a role in the origin of these movements. It is interesting to point out that in our case, eye closing was slower when it was associated with swallowing movements, indicating an interaction between both clinical features.

In conclusion, periodic PHM is a rare entity whose origin remains unclear. As previously pointed out by other authors, its occurrence in the context of anoxia is an agonal clinical sign. Further v-EEG studies are necessary so that we may better understand and delineate this condition.

Legend for video sequence
Video recorded in the intensive care unit. Note the seesaw phenomenon occurring periodically every approximately five seconds. Swallowing movements after the eye opening occurred with distinct time intervals, most frequently every 10 seconds.

References