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Isolated transient myoclonus in the elderly: An under-recognized condition?

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ABSTRACT

Background: Cases of transient myoclonus without other neurological manifestations in the elderly have very rarely been reported.
Objective: To report clinical features of elderly people with isolated transient myoclonus.
Methods: Clinical and laboratory features of 11 consecutive patients with isolated transient myoclonus (six men and five women; mean age, 75 years) were reviewed. Transient myoclonus was defined as an acute onset of tremulous myoclonus with/without asterixis in adults without other neurological symptoms.
Results: Preceding infections were recorded in five patients (pneumonia, two; upper respiratory tract infection, two; and septic arthritis of the shoulder, one). Myoclonus predominantly affected the head and/or neck ($n = 10$) and upper extremities ($n = 11$), compared with the trunk ($n = 2$) and lower extremities ($n = 6$). Asterixis was observed in six patients. Laboratory testing, neuroimaging, and electroencephalograms revealed no specific abnormalities. With or without treatment using benzodiazepines, myoclonus in all patients resolved completely within 1–4 days, although five had recurrence 2–19 months after their first episodes. Among these five patients, the accompanying asterixis patterns (presence or not) in four were different in the first and subsequent episodes.
Conclusions: Isolated transient myoclonus with or without asterixis may be more common than generally believed, and it could be a clinical entity or disease spectrum. Transient myoclonus is a benign condition in the elderly, but can be under-reported or misdiagnosed. Therefore, it is important to recognize that the elderly may have this syndrome.

Myoclonus is defined as sudden, brief, shock-like, involuntary movements caused by muscular contractions or inhibition [1–5]. Muscular contractions produce positive myoclonus, whereas muscular inhibitions (interruptions of tonic muscle activities) produce negative myoclonus or asterixis. In general, positive myoclonus is more common, whereas negative myoclonus frequently occurs in hospital settings, as a result of toxic or metabolic causes [5]. Myoclonus may be related to several etiologies and mechanisms, and a combination of both positive and negative forms may be present in the same disease such as post-hypoxic encephalopathies. In 1992, Hashimoto et al. reported seven elderly Japanese patients who developed acute co-occurrences of generalized positive myoclonus and asterixis that lasted for a few days; they named this syndrome as "transient myoclonic state with asterixis" [6]. Two of seven patients had chronic renal failure, but no metabolic or organic abnormalities were observed in the other five. Since this report was published, transient myoclonus (TM) with or without asterixis has been reported in elderly patients in Japan. However, isolated TM has rarely been reported in other countries. In the English literature, only two case series have been published from Japan, including the original report [6,7]. This shows that TM is not recognized worldwide and most review articles of myoclonus neglect TM, except for a review written by Shibasaki [8]. In this study, we report the clinical features of 11 cases of TM and review previous reported cases.

1. Methods

1.1. Patients

We retrospectively studied the case records of 11 consecutive patients with TM who were admitted to the neurology department of our institution between April 2005 and March 2013. The criteria for inclusion (modified from criteria related to transient myoclonic state with asterixis by Hitomi et al. [7]) in this study were as follows: (1) acute onset of tremulous myoclonus

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with/without asterixis in adult patients without other neurological symptoms, including impaired consciousness and opsoclonus; (2) spontaneous recovery or marked improvement after receiving medication within a few days; and (3) documentation of all signs by a neurologist. Patients with a history of seizure disorders, drug abuse, liver cirrhosis, chronic renal failure (>serum creatinine 2.0 mg/mL) or dialysis syndrome, hyponatremia (<135 mEq/L), hypoglycemia (<70 mg/dL), hyperglycemia (>200 mg/dL), hypoxia or post-hypoxic events, post-traumatic events, heat stroke, electric shock, and decompression injury were excluded from this study because these conditions can induce myoclonic movements [2]. Recurrence was defined as TM relapse, where the neurologist in our department confirmed myoclonus directly during the episode.

1.2. Laboratory testing

Blood cell counts and routine blood chemistry tests were performed during the first neurological examination. All patients underwent brain computed tomography (CT) and/or magnetic resonance imaging (MRI). A conventional 10–20 system electroencephalogram (EEG) was recorded for nine patients.

1.3. Treatment

Each responsible neurologist made final decisions regarding treatment using oral benzodiazepines.

2. Results

2.1. Clinical features

The patients were six men and five women with a mean age of 75 years (range, 54–89 years). The family history of myoclonus was negative in all cases. The clinical features are summarized in Table 1. Ten patients had chronic diseases such as hypertension and received medication such as antihypertensive agents. However, none received anesthetic agents or anticonvulsants. Preceding infections were reported for five patients (pneumonia, two; upper respiratory tract infection, two; septic arthritis of the shoulder, one). Some cases had myoclonus in the recovery phase of an infection, whereas others had myoclonus in the acute phase. However, all cases were referred to our institution because of myoclonus and not because of infection. The myoclonus distribution was as follows: head and/or neck, 10; trunk, two; upper extremities, 11; and lower extremities, six. No patients exhibited laterality of myoclonus. Asterixis was observed in six patients in the first episode.

2.2. Laboratory testing and neuroimaging

The serum creatinine levels were 0.5–1.5 mg and were mildly elevated in two patients (1.4 and 1.5 mg/dL). The serum sodium, potassium, and transaminase levels were normal in all patients. The serum ammonia levels were evaluated in nine patients, which were also normal. The blood sugar level was 77–159 mg/dL. Brain CT/MRI detected no specific changes, although old cerebral infarctions were observed in four patients. Of nine patients who underwent EEG, five had no abnormalities and four had nonspecific mild slow activities or irregularities; none had epileptiform discharges or triphasic waves.

2.3. Clinical course and recurrence

Four patients received benzodiazepines. However, myoclonus completely resolved within 1–4 days in all patients, irrespective of whether they received benzodiazepines. None required maintenance medication for myoclonus. Five patients experienced TM recurrence 2–19 months after their first episodes, and one of them had preceding pneumonia during both episodes. All of the TM recurrences were resolved within a few days. In the five patients with TM recurrence, the pattern of the accompanying asterixis (present or not) differed in four cases between the first and recurrent TM episodes.

3. Discussion

Our study found that 11 elderly patients presented with a benign transient myoclonic state syndrome with/without asterixis, which had a benign prognosis, although it relapsed occasionally. Drug-induced myoclonus was unlikely because the myoclonus recovered completely within a few days without discontinuing drugs that are usually taken for chronic disease. Epileptic myoclonus in elderly patients has been reported as progressive myoclonus epilepsy [5]. Progressive myoclonus epilepsy was ruled out in our patients because of the lack of family history, tonic or clonic seizures, progressive neurological decline, or epileptiform discharges in EEG, even though only nine patients underwent EEG. Moreover, none required maintenance medication with antiepileptic drugs for myoclonus and all had benign courses.

The first case series by Hasimoto et al. applied the term “transient myoclonic state with asterixis” to all cases with positive myoclonus and asterixis, while subsequent case series or case reports of TM with/without asterixis were referred to as “benign transient shuddering-like involuntary movement” [9], “transient involuntary shuddering movement” [10], “benign transient shivering-like involuntary movement” [11] or “transient myoclonic state” [12]. These were believed to be the same conditions because the clinical features were extremely similar to Hashimoto’s original report, except for the presence or absence of asterixis. Transient myoclonic state with asterixis and TM without asterixis may be a common clinical entity or a disease spectrum because our study showed that the presence of accompanying asterixis often differed in the same patients between the first and recurrent episodes. Thus, the presence of asterixis may not be a strong characteristic in the diagnosis of this syndrome. Therefore, “TM” may be a more suitable name than transient myoclonic state with asterixis. The published cases of TM in the elderly are summarized in Table 2. The age of reported TM cases ranged from 51 to 86 years and there were no obvious sex-related differences. Seasonality or outbreaks were not reported. The reported cases and our cases all lacked opsoclonus or ocular flutter. Therefore, they were distinct from opsoclonus–myoclonus syndrome. The clinical features of previous reported TM cases and our case series were as follows: (1) a sudden onset of myoclonus with/without asterixis and myoclonus was common or predominant in the upper extremities, shoulder, or head/neck; (2) in some cases, myoclonus was enhanced by actions or postures [6,7,9,11] but diminished while sleeping [9,11]; (3) patients often had chronic diseases such as hypertension; (4) a history of recent infectious illness was sometimes recorded; (5) no signs of altered consciousness, seizures, or other clinical signs; (6) spontaneously resolved within several days, although benzodiazepines (diazepam or clonazepam) may be effective; and (7) frequent relapses. A case with two recurrences of TM after urological surgery on each occasion was reported [13], while one of the authors of our study (A.H) experienced TM the day after urological surgery in another hospital (unpublished data), which suggests that infection, surgery, or other forms of stress can produce TM.

Myoclonus can be classified in three ways based on its distribution, etiology, and anatomic origin, which are based mainly