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M E D I C A L Mah-jong-induced seizures: case reports and review of twenty-three patients

'Mah-jong epilepsy' is a rare reflex epilepsy syndrome, manifesting as recurrent epileptic seizures triggered by either playing or just watching mah-jong. We present three patients with this condition and review all the reported cases. Mah-jong-induced seizures can be considered a subtype of cognition-induced epilepsy. Nonetheless, these patients have distinctive clinical and electrophysiological features: late age of onset, different seizure patterns, single seizure-trigger, lack of spontaneous seizures, and electroencephalographic findings not supportive of idiopathic generalised epilepsy. The pathophysiological mechanism underlying mah-jong-induced seizures may be different from the other cognition-associated reflex epileptic phenomena.

Introduction

In patients with epilepsy, a whole range of homeostatic variations, such as body temperature, sleep deprivation, and hormonal changes during menstrual cycles can influence susceptibility to seizures. Besides these well-recognised provocations, some specific stimuli can trigger seizures in certain patients. Examples of this reflex phenomenon include photic-induced seizures,¹ musicogenic epilepsy,² hot water epilepsy,³ reading epilepsy,⁴ and seizures induced by thinking and spatial tasks.⁵ Mah-jong is one of the most popular leisure activities in many Chinese communities. In our locality, the majority of people play mah-jong for fun rather than for gambling. In this paper, three cases of reflex epilepsy associated with mah-jong are described.

Case reports

Case 1

Patient 1 presented to us in March 1999 at the age of 79 years with a generalised tonicclonic seizure (GTCS). He had been playing mah-jong for 8 hours prior to its onset, and the seizure occurred while he was holding a ready hand. On admission, he had postictal drowsiness, which gradually subsided over a few hours. Both the neurological examination and computed tomography (CT) of the brain were unremarkable. An interictal electroencephalogram (EEG) was also normal with no demonstrable photosensitivity.

This man had experienced two similar events before. Three years ago, he had his first GTCS, which happened while he was shuffling the tiles. The second attack occurred 1 year later, also during a mah-jong game. None of the events were associated with sleep deprivation or excessive betting. He had been playing mah-jong for many years for pleasure but only played infrequently. Not every game was associated with seizures. He had no other neurological disorders, including spontaneous seizures, and no family history of epilepsy.

Key words Cerebral cortex; Electroencephalography; Epilepsy, generalized; Epilepsy, reflex; Gambling

In view of his recurrent seizures, our patient was given sodium valproate. He also refrained from mah-jong playing (after his third attack) and has remained seizure-free since then. Sodium valproate was discontinued after 5 years.

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Case 2

Patient 2 had been a regular mah-jong player from 20 years of age. In October 1996, at the age of 42, he developed his first seizure while playing mah-jong, and was admitted. The attack was a GTCS and was preceded by a vague prodrome of tiredness. He had no family history of epilepsy. A physical examination did not reveal any neurological deficits. His brain CT and EEG were normal. Intermittent photic stimulation elicited no electrical changes. Shortly after discharge, he had a second GTCS, which was also precipitated by playing mah-jong. Seizure prophylaxis with phenytoin was commenced. From then on he abstained from playing mah-jong and had no seizures. He chose to stop taking phenytoin after 1 year of treatment.

Three years later, he resumed playing mah-jong regularly and remained asymptomatic for another 3 years before his condition relapsed: he experienced two more GTCS, both provoked by playing mahjong. Despite resumption of phenytoin, he had two further mah-jong–induced attacks. All his seizures were generalised at the onset. He noticed no definite association between their occurrence and particular stages of a round or game, significant sleep deprivation, or excessive excitement. Eventually, he decided to quit mah-jong for good, and has been seizure-free for the last 18 months.

Case 3

Patient 3 was an occasional mah-jong player. He had good past health and no family history of epilepsy. In July 2006, he was admitted after developing GTCS during a mah-jong game. The attack occurred after 2 hours of playing. Our patient had abrupt loss of consciousness and was unable to recall the exact details of the seizure. The event was witnessed by three people, whose descriptions did not suggest a focal seizure.

Two years earlier, when aged 39 years, he had a similar event that was also triggered by mahjong playing. Other than these two attacks, he had never experienced any seizures. A neurological examination was unremarkable and the EEG only showed a mild excess of theta discharges over the frontal leads bilaterally with no photoparoxysmal response. Magnetic resonance imaging (MRI) of the brain demonstrated no abnormalities. He was seen regularly at the Epilepsy Clinic and was advised to avoid mah-jong. So far, there has been no seizure recurrence.

Discussion

Reflex seizures are defined as seizure attacks provoked habitually by specific afferent stimuli or activities.^{6,7} Reflex seizures can be precipitated by simple exogenous stimuli (eg visual, auditory, tactile) or more complex events involving higher cortical functions (eg praxia, cognitive tasks, emotions). Reflex epilepsy or pure reflex epilepsy refers to cases in whom all seizures are exclusively triggered events (ie those without spontaneous seizures). Conversely, reflex seizures can be part of a complex trait underlying other epilepsy syndromes; 5 to 6% of epileptic patients exhibit a certain degree of reflex phenomena,⁷ with photosensitivity being the commonest.

麻將遊戲誘發癲癇發作23例報告與回顧

「麻將癲癇」是一種罕見的反射性癲癇綜合徵,指參加或觀戰麻將遊 戲引起陣攣性癲癇發作。本報告描述三個病例,同時回顧所有病例紀 錄。由麻將遊戲誘發的癲癇病也是一種認知性癲癇的亞類。患者都有 明顯的臨床和電生理特徵,包括發病年齡遲、有不同的發作形式、具 單一誘發因素、沒有患過慢性癲癇,以及腦電圖結果未有顯示病人所 患的是特發性全面癲癇。由麻將遊戲誘發的癲癇病發病機制可能和其 他與認知相關的反射性癲癇不同。

Reflex seizures associated with mah-jong, brought on by either playing or watching the game, constitute a rare condition not recognised until recently. To date, only 20 cases of 'mah-jong epilepsy' (MJE) have been reported in the English literature.⁸⁻¹⁰ Their clinical features and investigation findings, together with those of our three patients, are summarised in Table 1.

Among the 23 patients with MJE, mean age of seizure onset was 54 years (range, 34-76 years), and a prominent gender asymmetry was observed (M:F, ≥10:1). Most patients had GTCS. Attack frequencies ranged from once every 3 years to more than 30 per year, depending on how often mah-jong was played and individual susceptibility to developing seizures. Seizure latency, defined as the time lapse before seizure occurrence, ranged from less than 1 hour to 11 hours after starting the game. Besides activities associated with mah-jong, three cases also had seizures triggered by Chinese chess or card games. Eighty-seven per cent of patients had never experienced spontaneous seizures and, therefore, can be classified as pure reflex epilepsy. Their EEGs were either normal or showed non-specific changes and did not demonstrate the widespread epileptiform discharges typically seen in idiopathic generalised epilepsies (IGE). Computed tomography or MRI only picked up incidental abnormalities.

Could MJE be a coincidence? Playing mah-jong is associated with considerable stress, especially when monetary bets are involved, and prolonged playing can result in sleep deprivation. These physiological factors can certainly lower the seizure threshold in susceptible subjects. Nevertheless, the majority of MJE patients had no seizures other than those associated with mah-jong, and seizure latency was as short as 1 hour. Together with the unique pattern of presentation, mah-jong–induced seizure is most likely a genuine phenomenon (the epileptogenic effects of mental stress and sleep deprivation should not be ignored, though).

Mah-jong is a cognitively demanding game. It involves substantial higher mental processing and outputs: memory, concentration, calculations,

Source	Age of onset (years)	Sex	Seizure patterns	Seizure triggers
Present study	76	М	GTCS	Playing mah-jong
(Hong Kong)	42	М	GTCS	Playing mah-jong
	39	М	GTCS	Playing mah-jong
Kwan and Su ⁸ (Taipei)	41	М	GTCS	Playing mah-jong
	74	М	GTCS	Playing mah-jong
	73	М	GTCS	Playing mah-jong
	71	М	GTCS	Playing mah-jong
	53	М	Partial seizure with 2nd generalisation	Playing mah-jong, Chinese chess
	72	М	GTCS	Playing mah-jong
	70	М	GTCS	Playing mah-jong
	63	М	GTCS	Playing mah-jong
	55	М	Partial seizure with 2nd generalisation	Playing mah-jong
	55	F	GTCS	Playing mah-jong
	59	М	GTCS	Playing mah-jong
	71	М	GTCS	Playing mah-jong
Wan et al ⁹ (Kaohsiung)	40	М	Partial seizure with 2nd generalisation	Playing and watching mah-jong, computer mah-jong game
	36	М	GTCS	Playing mah-jong, Chinese chess
	50	М	GTCS	Playing and watching mah-jong
	37	М	GTCS	Playing mah-jong
	34	F	GTCS	Playing mah-jong
Chuang et al ¹⁰	51	М	Partial seizure with 2nd generalisation	Playing mah-jong, card games
(Kaohsiung)	44	М	GTCS	Playing mah-jong
	34	М	Partial seizure with 2nd generalisation	Playing mah-jong

TABLE I. Summary of clinical features and investigation findings in our patients and other reported cases of mah-jong-induced seizures*

CT denotes computed tomography, EEG electroencephalogram, GTCS generalised tonic-clonic seizure, Lt left, MRI magnetic resonance imaging, and Rt right

None of the patients had photosensitivity on testing

reasoning, strategies, sequential thinking and planning, consideration of alternative solutions, and a lot of decision-making. From this perspective, mahjong-induced seizures are best classified as a subtype or manifestation of cognition-induced epilepsy.¹¹ Other terms that have been used for this group of reflex epileptic syndromes include 'reflex epilepsy induced by thinking and spatial tasks',5 'epilepsia arithmetices',12 and 'decision-making epilepsy'.13 In this group of reflex epilepsies, the provoking event usually consists of three components: thinking, a visuospatial task, and a manual task, which interact in a complex manner.⁶ Simple sensory inputs, such as patterns on the mah-jong tiles or the sounds of them hitting each other, may or may not contribute to the epileptogenic process. Typical seizure triggers in decision-making epilepsy include chess and card games (first reported in Shanghai by Ch'en et al¹⁴), other strategic board games, such as Mastermind and checkers, performing complex calculations, drawing or copying geometrical figures, and manoeuvring the

Rubik's cube (it is more appropriate to classify 'videogame epilepsy' under a separate category because most cases involve photic- or pattern-sensitivity in addition to the cognitive component).

Goossens et al⁵ delineated decision-making epilepsy as a homogeneous syndrome related to IGE and aberrant parietal cortical activation. The neurophysiological mechanism for epileptogenesis in this condition is unknown. It is postulated that in subjects with hyperexcitable neuronal circuits, such as IGE, regional activation through complex mental activities could recruit areas of hyperexcitable cortex and, upon reaching a 'critical mass', spark off a generalised epileptic event.⁶

Furthermore, the possibility that seizure triggers in decision-making epilepsy are culturally determined should be considered. The relevance of a particular syndrome might simply reflect the relative popularity of certain games or intellectual activities within a population and susceptible subjects would

Spontaneous seizures	Interictal EEG [†]	Neuroimaging findings
No	Normal	Normal brain CT
No	Normal	Normal brain CT
No	Occasional bifrontal theta waves	Normal brain MRI
No	Intermittent slowing in bilateral fronto-temporal regions, more on Lt	6 mm lesion at Rt medial temporal region on MRI, disappeared after 7 months
Yes	Spikes in Lt medial and anterior temporal regions	Lacunar infarction at Lt thalamus on MRI
Yes	Intermittent slowing in Lt anterior and mid-temporal regions	Normal brain CT
No	Intermittent slowing in Lt fronto-temporal region	Normal brain CT
Yes	Spikes in Lt anterior temporal region	Lt parietal hyperdense lesion on CT
No	Spikes in Rt medial temporal region	Normal brain CT
No	Normal	Not available
No	Normal	Normal brain MRI
No	Normal	Normal brain CT
No	Normal	Normal brain CT
No	Normal	Cerebral atrophy on MRI
No	Normal	Lacunar infarctions at Rt basal ganglion and Lt caudate nucleus on CT
No	Rare Rt frontal sharp waves	Old haemorrhage at Rt basal ganglia and pituitary adenoma on MRI
No	Normal	Normal brain MRI
No	Normal	Midbrain degeneration on MRI
No	Normal	Normal brain MRI
No	Normal	Normal brain MRI
No	Slightly slowed background	Mild brain atrophy on MRI
No	Normal	Normal brain MRI
No	Few Rt frontal theta activity	Normal brain MRI

TABLE 2. 'Mah-jong epilepsy' and decision-making epilepsy: a comparison of clinical features and patient characteristics

	Decision-making epilepsy ⁵	'Mah-jong epilepsy'
Mean age at seizure onset (range) [years]	15 (11-39)	54 (34-76)
M:F	19:6	21:2
Seizure patterns		
Generalised tonic-clonic seizures	96%	78%
Myoclonic jerks	76%	0
Absence seizures	60%	0
Partial-onset seizures	0	22%
More than one seizure-trigger	76%	13%
Presence of spontaneous seizures	91%	13%
Generalised epileptiform discharges (spike, spike-wave, polyspike-wave) on EEG ⁻	68%	0
Focal abnormalities on EEG	28%	35%
Photoparoxysmal response on EEG	32%	0
Seizure remission achieved with medication(s)	Commented to be 'generally good'	53%

* EEG denotes electroencephalogram

manifest the syndrome provided an effective, rather than specific, stimulus is encountered. This hypothesis, which is yet to be proven, might explain the uneven distribution of game-related epilepsies or 'special' seizure triggers in different populations: chess and card games in westerners, soroban in Japanese, punchi in Sri Lankans, mah-jong in Chinese, etc.

Nevertheless, despite similarities in the seizure triggers, patients with mah-jong–induced seizures have clinical features clearly distinctive from those with classical decision-making epilepsy (Table 2), suggesting that MJE is a unique syndrome. The implications of these observations, including

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the pathogenic mechanisms for MJE versus other cognition-induced epilepsies, and the underlying physiological alterations that predispose these patients to triggered events, are yet to be defined.

In the three Taiwanese series,⁸⁻¹⁰ seizure prophylaxis with carbamazepine, sodium valproate, or phenytoin was attempted in 17 patients. Treatment was effective in nine (53%) patients only. Therefore, MJE appears to be refractory to medical therapy, and avoidance of precipitants may be the best means of achieving seizure control. This is also concordant with our experience. The prognosis for this group of patients should be favourable, provided they refrain from playing and watching mah-jong.

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