CASE REPORTS

Juvenile myoclonic epilepsy in an elderly patient

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Abstract

Juvenile myoclonic epilepsy constitutes 5–10% of idiopathic generalised epileptic syndromes. The clinical triad of absence seizures, myoclonic jerks and generalised tonic clonic seizures (GTCS) rarely begin after the second decade of life. We present the case of a 74-year-old lady presenting for the first time with myoclonic jerks and absences. The electro encephalogram revealed generalised polyspike and wave epileptiform activity with photosensitivity. On close questioning, she appeared to have had a single episode of GTCS at the age of 10. She remained asymptomatic for 64 years before the diagnosis was made. Although rare, idiopathic myoclonic epilepsy could remain asymptomatic for decades and can present in the elderly. Proper classification of this epileptic syndrome, even in the elderly, is essential in view of the response to appropriate antiepileptic therapy.

Keywords: juvenile myoclonic epilepsy, elderly

Introduction

Juvenile myoclonic epilepsy (JME) is characterised by a clinical triad of myoclonic jerks, absence seizures and generalised tonic clonic seizures (GTCS) [1–3]. The National General Practice Study of Epilepsy reported a 13% incidence of newly diagnosed seizures in patients over the age of 70 years [4]. The majority of epileptic seizures presenting after the age of 60 are presumed to be due to cerebrovascular disease or intracranial neoplasms. Idiopathic generalised epilepsy of late onset, if this condition actually exists, is likely to be the consequence of a genetic predisposition triggered by acquired epileptogenic factors [5].

A recent study on idiopathic generalised epilepsy with onset after the age of 20 years reported the age of onset of myoclonic epilepsy as 24 (±4.5) years with a presenting age of 20–39 years [6]. Juvenile myoclonic epilepsy has been anecdotally described to occur in unexpected age groups [7, 8].

We report the case of a 74-year-old lady who presented for the first time with a history of myoclonic jerks and absences, with electro encephalogram (EEG) features consistent with idiopathic generalised epilepsy.

Case report

A 74-year-old lady was referred to the neurology clinic with an 8-week history of episodic myoclonic jerking. Although there was no definite loss of consciousness, she felt that she ‘lost it’ during these episodes. Her daughter described episodes of her being unresponsive ‘appearing to think or concentrate’ for ∼1–2 minutes. There were no other neurological symptoms or signs. The medical history included high blood pressure, non-insulin-dependent diabetes mellitus, atrial fibrillation, hypothyroidism and breast cancer (in 1993). In particular, no previous history of similar events was recalled during the initial consultation. There was no significant family history. The magnetic resonance imaging (MRI) brain scan showed cerebrovascular ischaemic changes with normal bilateral carotid dopplers.

Standard interictal EEG showed generalised and bilaterally synchronous bursts of high-amplitude (up to 220 μV) spike and wave activity at 3.5–5 Hz. During intermittent photic stimulation with the eyes open, the EEG displayed several bursts of polyspike/spike and wave activity (Figure 1).

On further questioning after the EEG, the patient reported having had a single GTCS at the age of 10, during the Second World War. On retrospect, the patient volunteered a history of several falls in school as a teenager, which were attributed to ‘tripping’. These were never investigated, and she remained symptom-free for the next 64 years.

In view of the generalised epileptiform activity in the EEG, with a history of myoclonic jerks, absences (‘unresponsive’ episodes) and a probable history of GTCS aged 10, a diagnosis of ‘juvenile’ myoclonic epilepsy was made, which was indolent for over 60 years. She was commenced
on sodium valproate, which led to total seizure control at a dose of 500 mg twice daily.

**Discussion**

Juvenile myoclonic epilepsy has a classical triad of myoclonic jerks, absence seizures and GTCS on awakening, showing a characteristic age-related onset [1, 3]. If absence seizure occurs, they begin between the age of 5 and 16 years, with myoclonic jerks following 1–9 (mean 4) years later, usually around the age of 15. They rarely begin after the second decade of life, and GTCS usually, but not always, appear a few months later than the myoclonic jerks [1, 3].

There are very few case reports of myoclonic epilepsy diagnosed after the age of 65 [7–9]. In all these cases except one [9], the patients have had a diagnosis of epilepsy, albeit wrongly classified. The diagnosis was ‘rediscovered’ when they were reviewed for poor seizure control. The patient described by Marini et al. [9] did not seem to have had any episodes of absences, myclonus or tonic-clonic convulsions throughout her life. Our patient, in spite of having had a single episode of GTCS at the age of 10 and possible absences as a teenager, was asymptomatic for the following 64 years and hence not investigated or treated. Although the age, vascular risk factors and MRI findings made symptomatic partial epilepsy secondary to cerebrovascular disease possible, the presence of generalised polyspike and wave pattern with photosensitivity in EEG was in keeping with idiopathic generalised epilepsy. In this clinical context, this would support the diagnosis of JME.

Sodium valproate is considered the most effective anticonvulsant in controlling JME. Clonazepam (0.5–2 mg) may be useful for myoclonic jerks, but can make the GTCS worse [10]. Although lamotrigine has been reported to occasionally worsen the myoclonic jerks [11], it is being increasingly recognised as a useful alternative as monotherapy or in combination with valproate [12, 13]. It is possible that levetiracetam would be an effective add-on therapy in refractory patients [14]. Most patients would require lifelong therapy.

We believe that this is the longest seizure-free interval in an untreated case of JME, presenting for the first time at the age of 74.

**Conclusions**

Our case shows that JME should be suspected at any age and demonstrates the importance of obtaining a precise history and performing an EEG with photic stimulation in older adults with myoclonic seizures, GTCS or absence-like events. Although such cases are probably very rare, a correct diagnosis is essential, since carbamazepine and phenytoin, commonly used for focal epilepsy in this age group, can worsen the seizures.
Key points

- Proper evaluation of history is essential in the syndromic classification of epilepsy at any age for instituting the appropriate treatment.
- Juvenile myoclonic epilepsy can present even in the elderly.
- Carbamazepine and phenytoin may worsen myoclonic seizures.

Conflicts of interest

None declared.

References


Acute Epstein–Barr virus infection in two elderly individuals

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Abstract

Most individuals acquire Epstein–Barr virus (EBV) infection in young age. Because of uncommon presentation and misdiagnosis, clinical manifestations are less well described in older age. We present two cases of elderly patients with predominant symptoms attributed to cold agglutinin haemolytic anaemia due to acute EBV infection without fever, lymphadenopathy, pharyngitis or splenomegaly. We conclude that misleading clinical manifestations are frequent in older individuals and may lead to inappropriate diagnostic invasive procedures.

Keywords: acute EBV infection, elderly individuals, cold agglutinin haemolytic anaemia, elderly