Reminder of important clinical lesson

A 15-day-old male infant presenting with jerking movements of the limbs

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Summary

This 15-day-old Caucasian male infant presented to the clinic with his mother for a well-child visit. During the visit, the mother mentioned that she had observed jerking movements of the limbs, arms more than legs. Various tests were performed and observation in the ward helped determine the characteristics of the seizure-like movements. The infant was born by vaginal delivery with vacuum extraction with a history of possible perinatal asphyxia. After seizures were observed in the ward, the patient was referred to neurology where an EEG was performed. It was normal, excluding epilepsy states. A normal newborn screen excluded most metabolic disorders. After history and examination, investigations and observation, benign sleep myoclonus was diagnosed. Benign sleep myoclonus is a benign disorder that consists of jerking movements of the limbs, usually involving the upper limbs more than the lower limbs and usually occurring during sleep. The seizures spontaneously stopped when the infant was approximately 1.5 months old. This was consistent with the final diagnosis of neonatal sleep myoclonus.

BACKGROUND

This case presents a differential diagnosis for seizures in the neonatal period. Seizures in the neonatal period can be related to a variety of causes including conditions with prognosis that can vary significantly depending on early diagnosis.

The incidence of seizures is higher in the newborn period than at any other stage of life. Most neonatal seizures occur within the first few days of life, with an incidence between 1.8 and 3.5 per 1000 live births.

This case helps realise that early diagnosis of neonatal seizures can make a big difference in the final outcome as some of the causes are not as self-limited as benign sleep myoclonus.

Another message is the need to increase seizure awareness among parents and caregivers in infants in the neonatal period as they can easily be missed.

CASE PRESENTATION

A 15-day-old Caucasian male infant presented to a paediatric clinic for a well-child visit. His mother mentioned that she had observed frequent jerking movements of his limbs particularly when he was sleeping and usually the upper limbs more than the lower limbs. She mentioned that she had never seen any other activity such as tongue thrusting, eye rolling or other changes during these episodes that typically lasted from 15 to 30 s. They disappeared spontaneously and holding the child or the limb did not make them stop sooner. The child afterwards appeared his normal self. There was never any associated fever or other signs of illness. The mother had on one occasion observed an episode while the infant was awake. She asked whether this was normal behaviour for an infant, as he otherwise appeared well.

Suspecting seizure activity, she was asked to keep a diary recording the episodes, when they occurred and their characteristics.

Significant medical history included possible perinatal asphyxia due to a nuchal cord around the neck at the time of delivery; a prolonged and difficult delivery requiring vacuum extraction and an umbilical cord arterial evaluation consistent with possible perinatal asphyxia. However, the infant had an uneventful course in the newborn nursery and later was discharged with the mother.

The infant lived at home with his parents and two siblings. No history of seizures or any other genetic or metabolic disorders in the family was noted. Family history was significant for type II diabetes mellitus in the father's family.

On a physical examination, the infant was found to be large for gestational age and appeared well. Cardiovascular, neurological, chest and ENT examination was normal. Growth parameters were 75% for age height, 75% for head circumference and over the 90% for weight. Remainder of the physical examination was normal. Seizure characteristics were recorded in the ward when the infant was admitted to rule out other causes and the patient was monitored. The infant was observed to have rhythmic jerking movements of the upper limbs on three occasions over a 24 h observation period and of the lower limbs on one occasion. He was sleeping in all these episodes while he experienced the movements. No eye rolling, frothing or foaming at the mouth, oxygen desaturation or changes in heart rate, electrocardiographic tracing, breathing or colour was recorded. No emesis was seen. The episodes lasted 15–40 s and afterwards the infant appeared well with no ill effects.

INVESTIGATIONS

Initial investigations that indicated possible perinatal hypoxia included an umbilical cord arterial blood gas analysis showing a pH of 7.21, pCO₂ 60 mm Hg, pO₂ 23 mm Hg, base excess of -3.7 mmol/l, HCO₃ of

Figure 1 Normal EEG pattern in a neonate.

24 mmol/l and blood gas oxygen saturation of 70%. An arterial blood gas from the neonate 20 min after birth showed a pH of 7.23, $p\text{CO}_2$ of 55 mm Hg, $p\text{O}_2$ of 60 mm Hg, HCO_3 of 17 mmol/l and a base excess of -3.4 . A repeat arterial blood gas 2 h after birth was normal with a pH of 7.34, $p\text{CO}_2$ of 41 mm Hg, $p\text{O}_2$ of 80 mm Hg, base excess of -3.2 mmol/l, HCO_3 of 23 mmol/l and blood gas oxygen saturation of 90%. This indicated that the initial period of perinatal asphyxia was resolved.

Newborn screen was normal. Other tests included a repeat newborn screen, complete blood count, basic metabolic panel, liver function tests and lumbar tap for spinal fluid examination, a brain MRI at 2 weeks of age when the jerking movements started and an EEG at 3 weeks of age. These were all normal; the EEG did not show any focal or generalised seizure activity (figure 1).

The fact that the EEG was normal indicated that the seizures were of subcortical origin. This is concurrent with the final diagnosis of neonatal sleep myoclonus, which is considered subcortical in origin as compared to epilepsy states that originate from the cerebral cortex.

DIFFERENTIAL DIAGNOSIS

The most common cause of neonatal seizures is hypoxic–ischaemic encephalopathy (HIE) brain injury due to any of a variety of causes including intrauterine causes such as abruptio placentae, cord compression, preeclampsia or

chorioamnionitis. Postnatal conditions such as persistent pulmonary hypertension of the newborn, cyanotic congenital heart disease, sepsis and meningitis can also result in HIE brain injury. Rarely, primary epilepsy can present with neonatal seizures.¹ Usually, seizures due to HIE develop within 24 h of injury; however, this is not a reliable marker for HIE.²

Seizures due to intracranial haemorrhage may also be associated with hypoxic–ischaemic or traumatic injury as these events are frequently associated with each other. Onset of seizures due to subarachnoid haemorrhage or subdural haemorrhage is usually on the second or third day of life, whereas those due to germinal matrix–intraventricular haemorrhage present after the third day.

Seizures due to neonatal sleep myoclonus usually appear after the first few weeks of life and are not associated with cardiorespiratory compromise. They are usually self-limited, as in this case.

TREATMENT

In consultation with paediatric neurology, no pharmacological treatment was provided to the infant after confirmation of the normal EEG, MRI and laboratory studies. This is in accordance with the latest recommendations for management of benign sleep myoclonus. The infant's mother was asked to keep a record of episodes and their characteristics. The parents were taught infant cardiopulmonary resuscitation in case of apnoea or respiratory difficulty.

Figure 2 Suppressed pattern of EEG, which may indicate depressed central nervous system including infective states, drug-induced coma, brain damage from stroke or hypoxic–ischaemic encephalopathy, and which may alternate with bursts of high-amplitude activity (burst suppression).

Counselling regarding sudden infant death syndrome and avoiding soft toys or clothing around the infant's face was provided. Close clinical follow-up was provided.

OUTCOME AND FOLLOW-UP

Spontaneous cessation of seizures occurred at 1.5 months of age confirming the diagnosis of neonatal sleep myoclonus. At the time of writing, the infant is 8 months old and has

not had a recurrence of the seizure like activity. He is developmentally well and achieving appropriate milestones.

DISCUSSION

The incidence of seizures is higher in the newborn period than at any other stage of life. This is related to a change in the neuronal response to γ -aminobutyric acid. Most neonatal seizures occur within the first few days of life,

Figure 3 Seizure pattern of EEG. Seizures in neonates seen on EEG may have multiple causes including hypoxic–ischaemic encephalopathy, neonatal ischaemic or haemorrhagic strokes, infantile spasms, metabolic or electrolyte abnormalities. EEG can appear to have focal or multifocal spikes or sharp waves.

Figure 4 Burst suppression pattern of EEG. High-voltage periods of slow, sharp and spiking activity alternating with a suppressed background have been termed burst suppression. Indicates an obtunded or comatose state, often with a poor prognosis.

with an incidence between 1.8 and 3.5 per 1000 live births. Premature infants have a higher frequency of seizures and their seizures are less organised.

A clinical seizure results from excessive depolarisation of neurons in the central nervous system. Hypoxaemia, ischaemia and hypoglycaemia can result in significantly decreased energy production and increased release of glutamate, the principal excitatory neurotransmitter in the cerebral cortex. Hypocalcaemia and hypomagnesaemia cause increased depolarisation by increasing sodium influx across the neuronal cell membrane. In addition, the inhibitory pathways are not well developed early in life.

EEG is essential in diagnosis and management of neonatal seizures. As there may not be ictal activity on EEG even during a seizure (*electroclinical dissociation*), serial

EEGs or continuous EEG monitoring (when available) are of benefit. The EEG is analysed for ictal activity (focal or multifocal spikes or sharp waves and focal monorhythmic discharges) and for background activity. Background abnormalities are predictive of the risk of electrographic seizures and poor prognosis (figures 2–4).

Neonatal primary seizure disorders or epileptic syndromes do occur, but at a very low frequency.¹ The major aetiologies of neonatal seizures include HIE, intracranial haemorrhage, metabolic disturbances, electrolyte disturbances (hypoglycaemia, hypocalcaemia and hypomagnesaemia), intracranial infection, developmental defects and drug withdrawal (table 1).

Diagnostic evaluation includes glucose, electrolytes, calcium, magnesium, and phosphorus in order to identify

Table 1 Clinical characteristics, classification and presumed pathophysiology of neonatal seizures^{2 3}

Type of seizure	Typical age of onset	Location	Clinical	Aetiology	Relation to EEG
Subtle (motor automatisms)	Preterm and term	Usually not generalised	Eye deviation, (term) blinking or fixed stare, (preterm) repetitive mouth and tongue movements, apnoea, bicycling or rowing movements, tonic posturing of limbs	Non-epileptic	Most often, these clinical seizures occur without EEG seizure activity. However, EEG seizure discharges may be present correlating with seizure
Tonic (generalised or focal)	Primarily preterm	May be focal or generalised	Tonic extension or flexion of limbs, eye deviation	Generalised tonic: intracranial haemorrhage in preterm, obtunded or lethargic infants ⁴ Focal tonic: epileptic	Focal tonic epileptic in origin, associated with focal EEG seizure activity. Generalised tonic in infants with depressed EEG background indicating forebrain depression ⁵
Clonic	Primarily term	May be focal or multifocal	Clonic limb movements (synchronous or asynchronous, localised or often with no anatomic order to progression); consciousness may be preserved, cannot be suppressed by restraint; <i>Jacksonian march</i> can occur	Often signals focal cerebral injury, epilepsy	Close EEG relation to seizure activity with each contraction having a consistent, time-locked relationship to EEG seizure discharges
Myoclonic	Rare	Focal, multifocal or generalised	Sudden jerks of the extremities, upper more than lower, may be provoked by stimulation. Resolve spontaneously at 2–3 months of life in <i>neonatal sleep myoclonus</i>	Epileptic or non-epileptic; may be cortical or subcortical in origin (most cases of neonatal sleep myoclonus)	Some correspond to EEG seizure discharge, others do not have a corresponding EEG seizure discharge

an immediately correctable metabolic condition. Spinal fluid analysis is performed to identify a potential bacterial infection. CT and magnetic resonance imaging (MRI) can delineate the brain anatomy with high sensitivity and resolution (MRI better than CT). MRI scanning is very effective for determining the presence and extent of HIE injury and of parenchyma brain injury.⁴ Ultrasound is not adequate for the diagnosis of neonatal seizures as it is not effective at detecting subdural or epidural bleeds or identifying parenchymal injury. Lesions of HIE injury can be identified within the first 2–3 days after the asphyxial event. The EEG is used to confirm the presence of seizure activity and to define the background electrical activity, which is valuable in estimating prognosis.

Treatment of neonatal seizures⁶ should focus on the primary aetiology as well as on direct seizure control. Neonates are less likely to incur seizure-related injury. Treatment of underlying metabolic disturbances such as hypomagnesaemia, hypoglycaemia or hypocalcaemia is necessary. Ensuring proper ventilation and perfusion. Treating any underlying infective cause. When pharmacological therapy is considered useful, phenobarbital is often used as the first-line anticonvulsant, followed by phenytoin and lorazepam. Oral phenytoin is poorly absorbed from the gastrointestinal tract of the infant.

Prognosis varies as a function of primary aetiology and gestational age of the infant. The background EEG activity is also correlated with outcome in both term and preterm infants. Infants with normal background activity are less likely to have neurological sequelae than those with moderate to severe abnormalities such as burst-suppression pattern, voltage suppression and electrocerebral silence.

The infant in this case report was followed up for course of events. The mother was asked to keep a record of the seizure-like movements. Neurology referral was done where all the investigations were studied including the EEG. The combination of the normal EEG indicating the subcortical origin of the seizures as well as the nature of the observed episodes and the normal laboratory findings and normal MRI supported the diagnosis of neonatal sleep myoclonus. This diagnosis was confirmed at 1.5 months of age when the seizures spontaneously ceased.

Another article on neonatal sleep myoclonus has been published in PubMed prepared by the Institute of

Neurology, Catholic University of Rome, Policlinico Agostino Gemelli, Rome, Italy.⁷

Learning points

- Neonatal seizures are a fairly common occurrence that has a variety of causes ranging from benign, such as in this case (neonatal sleep myoclonus), to potentially life-threatening, such as intracranial haemorrhage or neonatal meningitis.
- Neonatal seizures often go unrecognised by parents and caregivers as they may not be the generalised tonic clonic type of seizure seen more in older patients.
- Appropriate education of parents and caregivers to recognise and report seizures early in their course is important.
- Early recognition of neonatal seizures is vital as it can lead to earlier treatment if required in conditions such as intracranial haemorrhage, meningitis and metabolic conditions or hypoxic–ischaemic encephalopathy.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Wolfe P**. International classification of the epilepsies. In: Engel J Jr, Pedley TA, eds. *Epilepsy: a comprehensive textbook*. Philadelphia: Lippincott-Raven, 1997:773.
2. **Co JP**, Elia M, Engel J Jr, et al. Proposal of an algorithm for diagnosis and treatment of neonatal seizures in developing countries. *Epilepsia* 2007;**48**:1158.
3. **Perlman JM**. Intrapartum hypoxic-ischemic cerebral injury and subsequent cerebral palsy: medicolegal issues. *Pediatrics* 1997;**99**:851.
4. **Leviton A**, Nelson KB. Problems with definitions and classifications of newborn encephalopathy. *Pediatr Neurol* 1992;**8**:85.
5. **Murray DM**, Ryan CA, Boylan GB, et al. Prediction of seizures in asphyxiated neonates: correlation with continuous video-electroencephalographic monitoring. *Pediatrics* 2006;**118**:41.
6. **Chapman KE**, Mizrahi EM, Clancy RR. Neonatal seizures. In: Wyllie E, ed. *Wyllie's treatment of epilepsy: principles and practice*. 5th edn. Philadelphia: Lippincott, Williams & Wilkins.
7. **Vaccaro ML**, Valenti MA, Carullo A, et al. Benign neonatal sleep myoclonus: case report and follow-up of four members of an affected family. *Clin Electroencephalogr* 2003;**34**:15.

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