SHORT REPORT

“Isolated” postinfectious myoclonus

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Abstract
Two cases are reported who developed myoclonus as the only manifestation of a post-infectious syndrome without evidence of encephalitis or the opsoclonus-myoclonus syndrome. Case 1 had generalised myoclonus following an influenza-like illness, while case 2 had right upper limb segmental myoclonus following uncomplicated chicken pox. Neither had any localising neurological signs or abnormality on investigation. Both recovered completely within six months of the onset. Similar cases are reviewed from the literature and it is suggested that such cases be called “isolated” post-infectious myoclonus.

(Myoclonus can be a clinical feature of many infectious encephalitides and was notable in the acute and chronic phases of epidemic encephalitis lethargica. More recently it has been recognised as a pathognomonic feature of subacute sclerosing panencephalitis and Creutzfeld-Jacob disease. Myoclonus has also been described in acute encephalitis due to a variety of viral and non-viral agents which either directly invade the CNS, or by an immunological process cause an acute disseminated encephalomyelitis (also called post-infectious encephalitis). In either case, the clinical picture is of an acute illness characterised by seizures, alterations in consciousness, and focal neurological signs. Myoclonus, when present, is usually only a minor feature of the overall clinical syndrome. If the patient survives the acute illness the myoclonus usually disappears. Segmental myoclonus following herpes zoster infection is also well known and is usually transient. The opsoclonus-myoclonus or the “dancing eyes and dancing feet” syndrome is a brain stem encephalitis preceded or associated with a variety of known and often unknown infectious agents. This self limiting condition is easily recognised because of the association of the myoclonus with the characteristic eye signs.

Here, we draw attention to patients who develop generalised or focal myoclonus, as an isolated phenomenon, after a mild illness (probably viral) without any features of encephalitis or of the opsoclonus-myoclonus syndrome. These patients have no other abnormality on clinical examination or investigations and the myoclonus is usually short lasting. Although post-infectious encephalitis as a cause of myoclonus has been included in lists of aetiological classifications, the syndrome of isolated myoclonus following infection has not been clearly defined. In the past such patients have been referred to ambiguously and one report included them within a group labelled “myoclonus without seizures or neurological or mental deficit.” We report two cases with myoclonus as the sole manifestation of a post-infectious syndrome, and have reviewed the few similar cases from the literature.

Case reports
Case 1  A 23 year old woman, presented with a 14 week history of an influenza-like illness dominated by generalised body ache, lethargy and fever. Two weeks after the onset she developed episodic generalised jerks of the arms and legs, each lasting for up to 30 seconds, and occurring every two hours or so, without loss of consciousness, or other epileptic phenomena. She was otherwise normal between attacks. Treatment with clonazepam improved the spasms but made her very drowsy. When discontinued 2 weeks later, the spasms became more intense and responsive to noise, sudden movement and startle. She also developed constant twitching of her arms and legs making it difficult for her to use her hands, walk, and speak. She was investigated elsewhere at this stage. An EEG carried out showed runs of polyspike/spikes at about 5/6 per second, associated with the episodes of jerking. CT scan of the head and CSF examination at that time were normal. She was treated with clobazam which benefited her initially, but she developed tolerance to it, as she did to propranolol which was tried next. At this stage, when seen in our department, she had the jerking attacks only once or twice a day. However, she had spontaneous, repetitive wriggling jerks of arms, legs, and head, lasting many minutes and increasing on stress. At the beginning of her examination, she could not walk because of the jerking of her legs. She also had almost continuous rhythmic multifocal myoclonus of both arms, and occasional bursts of titubation of the head. Twenty minutes later the large jerks gradually settled and disappeared. She could then walk normally heel to toe and hop on either leg. The rest of the
CNS examination revealed no abnormality in tone, power, and deep tendon reflexes with the plantar responses being flexor and sensation normal. She was advised to take sodium valproate 500 mgs, to which she showed an excellent response and the myoclonus subsided almost completely in a few months.

Case 2 A 14 year old girl presented with myoclonic jerking of the right arm, two weeks after the onset of uncomplicated chicken pox which resolved in about 10 days. The jerking of the arm progressively got worse and reached its peak after three weeks making the use of her right hand difficult, especially for writing. The jerking worsened when she was anxious or tired. She had no weakness or sensory symptoms in that arm. Five weeks after the onset when seen by us she had begun to notice some improvement. Her past medical history was unremarkable.

On examination higher mental function was normal as were the cranial nerves. There were occasional jerks of the right triceps, brachioradialis, wrist and finger extensors and biceps with sparing of the spinati and pectorals. Some jerks appeared myoclonic while others were longer lasting and resembled tonic spasms. The jerks increased on sustained posture. There was no stimulus or action sensitivity. Occasional dystonic posturing of the right hand was also noted. On one occasion she was seen to have some jerks of the left upper and lower limbs but this was not a repeated observation. There was no wasting, and the tone, power and reflexes were normal. The deep tendon reflexes and plantar responses were normal and there was no sensory deficit. General examination was normal except for a mild thoracic scoliosis with convexity to the left.

Electrophysiological investigation showed a single long duration burst and short trains of 3 bursts between 50 and 100 milliseconds in the right brachioradialis and triceps muscles. Back-averaging the EEG activity preceding the jerks did not show any time locked cerebral events. Cortical somatosensory evoked responses were of normal latency and morphology. Magnetic stimulation of the motor cortex produced normal responses in the right arm. There was no stimulus sensitivity to percussion or electrical stimulation of the median nerves. EEG showed some asymmetry in the activity between the hemispheres with better formed alpha activity on the right. In addition, there were some runs of slow activity on the right. Full blood count, ESR, renal function tests, liver function tests, calcium, copper and ceruloplasmin, thyroid function, ANF and autoantibodies were all normal or negative. A CSF examination was not done because the patient was unwilling to undergo a lumbar puncture. A CT scan of the head was normal. The patient recovered completely within 6 months.

Discussion

Generalised myoclonus developed in case 1 following an influenza-like illness whilst case 2 developed segmental myoclonus following uncomplicated chicken pox. Neither patient had altered consciousness, seizures, or other clinical signs to suggest an acute encephalitis or menigitis. Myoclonus was the sole manifestation of their post-infectious syndrome. We suggest that the term isolated post-infectious myoclonus is appropriate to describe the myoclonic syndromes exhibited by these two patients and in others who satisfy the following criteria: 1) A sudden onset of generalised, multifocal or segmental myoclonus; 2) a history of a recent preceding infectious illness; 3) no features of encephalitis or the opsonolus-myoclonus syndrome; 4) a non-progressive course without seizures, ataxia, or dementia and 5) recovery in a short but variable period of time.

A number of similar cases are described in the literature where myoclonus developed following presumed viral infections of the upper respiratory tract or following an influenza-like illness (table4-15). The interval between the initial illness and the onset of myoclonus was short. None of these cases had any focal neurological signs. The course of the condition was benign and the prognosis good. Both our cases recovered in about six months and most in the literature recovered over a period of weeks. Silfverskiold17 described three young girls with the subacute onset of rhythmic myoclonus of upper and lower limbs who had no preceding illness. Although there was no definite evidence of previous infection, they were presumed to be post-infectious on epidemiological grounds, because all three lived in the same area, and developed a similar benign myoclonic syndrome in the autumn of 1958. All recovered over 6 to 18 months of onset.

There are other cases in the literature of myoclonus attributed to preceding infections.16,19 In a large survey of myoclonus, Lance16 described one patient (case 34) who developed generalised action myoclonus,
ataxia and an abnormal EEG two weeks after a presumed meningoencephalitis illness with complete recovery over the ensuing three weeks. Hopkins and Michael¹ described a 36 year old male who developed rhythmic myoclonus of the pelvis and lower limbs which lasted one week, was associated with absent knee and ankle jerks and electrophysiological evidence of a spinal origin for the myoclonus. There was no history of a preceding illness but a CSF pleocytosis and raised protein level suggested a recent (probable viral) infection. These patients did have evidence of CNS inflammation (encephalitis¹⁶ and myelitis¹⁷) and so were excluded as examples of isolated post-infectious myoclonus according to our criteria.

Why some patients should develop myoclonus without clinical or investigative evidence of structural damage to the CNS following non-specific or uncomplicated infectious illnesses is unclear. The pathophysiological origin of the myoclonus in our two cases is uncertain. Case 1, with generalised myoclonus, on one occasion was found to have runs of polyspike/spike potentials at 5–6 Hz on EEG corresponding to her jerks pointing towards a probable cortical origin of the myoclonus. In case 2, back-averaging of the EEG did not reveal any cortical correlates nor did she have enlarged somatosensory evoked potentials, suggesting that the myoclonus was probably not of cortical origin. No EEG correlates corresponding to the myoclonic jerks were reported in Silverskiold's cases with rhythmic upper and lower limb myoclonus.¹⁷ As might be expected therefore it appears that post-infectious myoclonus is a heterogenous entity and that different sites of the CNS can be affected. The underlying pathology is also uncertain.

We thank Dr R F Gledhill for referring case 2.


Early notions of hydrocephalus

The truth of claims that Hippocrates recognised hydrocephalus and drained the ventricles through the fontanelle is uncertain. Two patients are described in the second edition of Vesalius' De Fabrica (1555): in a two year old girl he had obtained “9lbs of water from the ventricles; I marvelled at nothing more than the amount of water had for so long collected in the ventricles of the brain without greater symptoms." In 1761 Contugno described the cerebrospinal fluid (CSF). "The remarkable Stockholm mystic, Emmanuel Swedenborg (1688–1772), left manuscripts unpublished until 1882 and 1887 showing that he recognised not only the four ventricles, but also the secretory role of the choroid plexuses and the circulation of CSF through the 4th ventricle in the medulla, "through a cleft in its ceiling, between the pia and dura mater, and thence to the spinal cord". The eighteenth century anatomists Morgagni, and the physician Robert Whytt demonstrated the relevant CSF pathways and made some sense of the pathology of hydrocephalus.¹

In the nineteenth century Magendie (1828), Luschka (1855) and above all, Key and Retzius described the CSF flow and formation. In his "Essay on hydrocephalus acutus" (1806), John Cheyne recorded 23 cases; all but six succumbed and the aetiology in most was probably tuberculous meningitis.¹

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