Co-trimoxazole has been used successfully as a sole agent in resolving dementia and intestinal involvement in Whipple’s disease, and in conjunction with ampicillin and chloramphenicol in reversing another case of CNS disease. Co-trimoxazole penetrates well the blood-brain barrier, confirmed as in this case by CSF antibiotic assay. The role of steroids with antibiotics is more controversial, but prednisolone may have been responsible for the rapid improvement in right knee effusion, and in mobility, and the swift resolution of left wrist weakness. Prednisolone has been suggested previously as responsible for recovery from meningeal involvement in Whipple’s disease. As disordered immunity may play a role in Whipple’s disease, the use of steroids is perhaps indicated if neurological damage is threatened. Full recovery from nervous system disease however may be limited by glosis and neuronal loss. Finally, peripheral nervous system involvement should be considered as part of the spectrum of Whipple’s disease.

We thank Dr G Wakefield, Consultant Neurologist at Royal United Hospital, Bath, for permission to submit this report; also Dr P Burton, Consultant Pathologist and Dr D Reeves, Consultant Microbiologist at Southmead Hospital; and Dr T Moss, Consultant Neuropathologist at Frenchay Hospital, Bristol.

Address for correspondence: Dr T Ferguson, MD, FRCP(E), Consultant Neurologist, Southmead Hospital, Southmead Rd, Westbury-on-Trym, Bristol, BS10 5NB, UK.

References


154

Asymptomatic cardiac arrhythmias in periodic paralysis

Sir: After the original report by Klein et al. in 1963, several new cases of periodic paralysis associated with cardiac arrhythmias have been described (2–7) (see table). In all of the reported cases, alterations of cardiac rhythm were principally characterised by isolated and paired ventricular ectopic beats and a directional ventricular tachycardia. This serious and rare arrhythmia may be considered a possible sign of periodic paralysis when found in a young patient. These cases of periodic paralysis with associated arrhythmias have been regarded as exclusively ventricular in origin with poor prognosis. We present what we believe is the first report of a case of periodic paralysis associated with a benign cardiac arrhythmia of atrial origin.

A 16 year old white male with no family history of neuromuscular disease suffered acute episodes of paralysis involving upper and lower limbs, lasting 2–4 days twice a year, since the age of 9 years. During hospitalisation in our Department because of one of these crises at the age of 13, laboratory data, including myoglobin, CK, aldosterones and thyroid function tests were all normal. Serum and urinary potassium and other electrolytes were measured daily and were consistently normal. A biopsy of the left quadriceps muscle showed no structural alteration and no glycogen accumulation with PAS. Trichromic stain showed a slight increase in interstitial connective tissue and the electron microscopy showed a few fibres...
with centralised nuclei. The ECG performed during the crisis showed a sinus rhythm with sporadic atrial ectopic beats with aberrant conduction. The dynamic ECG (Holter) after recovery of muscle strength showed a high incidence of atrial ectopic beats (mean = 105/h), many of which were conducted with varying degrees of aberrance. During the stress test, atrial ectopic beats were frequent up to 75 W, disappeared until the end of the test and were again frequent in the recovery phase. Bidimensional echocardiogram and radio-isotope cineventriculography with Tc99m revealed a slight enlargement of the right ventricle (diameter 30 mm) with normal contractions. A load test with 6 grams of potassium chloride by mouth provoked no reduction in strength or modification in the dynamic ECG pattern. A load test with 100 grams of glucose by mouth and 10 IU insulin brought on paralysis of the lower limbs after 2 h; 2 grams of KCl were administered by mouth with full recovery of motor function within 2 h. A prolonged ECG performed during the test showed 570 (mean = 114/h) atrial ectopic beats of which the majority were aberrant. The patient was then discharged, being given acetazolamide 250 mg twice a week and a low carbohydrate diet. During follow up (3 years) the patient experienced only one episode of paralysis of the legs which ceased 2 h after administration of KCl. The Holter ECG (3 recordings at intervals of 12 months for a total of 68 h) showed a high incidence of atrial ectopic beats (mean = 95/h), most of which were conducted with various degrees of aberrance, and numerous episodes of bigeminy with many paired beats.

The peculiarity of this case lies in the arrhythmias observed in the numerous ECG and Holter recordings. Our patient had a constantly high incidence of atrial arrhythmias often characterised by aberrant conduction. The atrial arrhythmias did not seem to be influenced by serum potassium concentrations, or by the presence or absence of neurological signs.

Periodic paralysis associated with cardiac ventricular dysrhythmias may be modified by hypokalaemia or potassium load (see table). However, although arrhythmias accompanying an attack of muscle weakness have been known to occur, in all these cases arrhythmias were clearly independent of attacks of paralysis and plasma potassium levels. Thus the most widely accepted hypothesis to explain these rare cases of periodic paralysis associated with cardiac ventricular arrhythmias is that there is a structural or functional alteration in the cardiac muscle at cell membrane level, as there is in the skeletal muscle. $^4$ $^8$ $^9$ Excitable muscle cells have proved to be constantly hypo-polarised, and even a slight depolarisation at cardiac level could be sufficient to generate arrhythmias. $^8$ $^11$ However, if we accept this hypothesis, it is not easy to explain the rare occurrence of cardiac arrhythmias associated with periodic paralysis.

That our case is a familial cardiac supraventricular rhythm disturbance associated with periodic paralysis cannot be tested, since the parents and relatives showed no attacks of paralysis and constantly normal ECG.

Since supraventricular arrhythmias are not uncommon in healthy young people, though not with such a high number of ectopic beats as found in our case, $^12$ we cannot exclude an incidental association. A more intensive cardiac survey of patients with periodic paralysis should be conducted to estimate the true incidence of asymptomatic arrhythmias in this population.

References

Eight days following hyperbaric treatment, he received one right unilateral ECT. He received 0-2 mg glycine muscle. His breathing was normal. Protective recording from the muscle was not interrupted with any complications. Within several hours, the patient began exhibiting prominent verbal sexual innuendos, undressing and exposing himself, and lunging at the breasts of female staff and groins of male staff. These outbursts of sexually aggressive behaviour lasted 10 to 15 minutes and were interspersed with otherwise lethargic and apathetic behaviour. The patient ate excessively, constantly complained of hunger, showed excessive handling of objects, and was easily distracted. He displayed prominent amnesia, both retrograde and anterograde, and denied any memory of his inappropriate behaviour. Disorientation to time and severe attention deficit were present. Neurological examination revealed ataxia, moderate rigidity, and brisk muscle stretch reflexes. Electroencephalography demonstrated low-to-moderate voltage 4 to 6 Hz semirhythmic theta and paroxysmal high voltage rhythmic 5 Hz theta activity. These diffuse paroxysmal theta bursts were associated clinically with blank staring followed by agitation and poor compliance with commands. The patient was placed on phenytoin. Haloperidol and lorazepam were required to control the outbursts of aggressive hypersexual behaviour that continued to alternate with long periods of somnolence. The inappropriate behaviour resolved over a course of 10 days and neurololigic and benzodiazepines were gradually withdrawn.

Repeat EEG 10 days after the initial recording revealed minimal-to-moderate generalised 6 to 7 Hz theta without paroxysmal irregularities. Cranial CT scan was normal. Mental status examination progressively improved in all spheres. At discharge 4 weeks following admission, neuropsychological testing revealed only mild-to-moderate deficits in naming and recent memory. There was no evidence of inappropriate behaviour or neurologic deficits. As the encephalopathy cleared, the patient’s primary affective disorder again became more apparent.

In 1937, Kluver and Bucy described an unusual syndrome appearing in rhesus monkeys following unilateral temporal lobectomy. These monkeys displayed (1) “psychic blindness,” (2) hyperorality, (3) an increased tendency to touch and examine objects (“hypermetamorphosis”), (4) loss of normal anger, fear, and pleasure responses, and (5) increased and altered sexual behaviour. The human Kluver-Bucy syndrome (KBS) closely parallels the syndrome seen in rhesus monkeys, involving (1) visual agnosia (especially prosopagnosia), (2) hyperphagia, (3) hypermetamorphosis, (4) placidity, apathy, and blunted affect, and (5) alterations in sexual behaviour.

addition, the human KBS is accompanied by more complex behavioural manifestations, such as aphasia, amnesia, dementia, and/or seizures. Partial or complete human KBS has been described in association with a variety of encephalopathic processes invariably associated with bilateral temporal lobe involvement. This paper is the first description of human KBS occurring in association with delayed carbon monoxide poisoning.

Neuropsychiatric sequelae of carbon monoxide poisoning occur in 15–40% of surviving patients. These sequelae frequently develop after a period of apparent recovery from the carbon monoxide exposure. The clinical picture of delayed neuropsychiatric deterioration following carbon monoxide poisoning has been described in numerous case reports.

These patients are usually deeply comatose following carbon monoxide inhalation but regain consciousness within 24 to 48 hours. Subsequent improvement is rapid, and they are often discharged from the hospital within a week. Choi found that a period of apparent recovery between the carbon monoxide insult and the onset of neuropsychiatric sequelae occurred in 11.8% of patients requiring hospital admission. The latest period varied from 2 to 40 days with a mean of 22.4 days. Recovery from the delayed sequelae occurred in 75% of patients within 1 year.

Similar descriptions of delayed post-cardiac arrest, hypotension, and anoxic encephalopathy have been reported following anaesthetic complications, cardiac arrest, hypotension, and anoxic encephalopathy. Several authors have reported that the delayed onset of neurological deterioration occurred immediately following an increase in patient activity, after discharge from the hospital, after emotional stress, and in one case, following ECT. While a period of bed rest following carbon monoxide poisoning has been recommended as a means of preventing delayed post-anoxic deterioration, it has yet to be determined that this therapeutic measure is effective.

The usual neuropsychological substrate of delayed neuropsychiatric deterioration following carbon monoxide poisoning is believed to be selective injury to the cerebral white matter. Cases with a delayed onset...