Letters

Transient global amnesia and migraine in twin sisters

Sirs: In 1958, Fisher and Adams coined the name “Transient global amnesia” for a syndrome already described simultaneously by Bender and Guyotat and Courjon in 1956. The physiopathology of transient global amnesia is still uncertain and the role of migraine has been discussed by some authors. Three familial observations of transient global amnesia have been previously published. We report the case of twin sisters who experienced attacks of transient global amnesia and migrainous accompaniments.

The first sister suffered from classic migraine, mostly left-sided, since her adolescence. In February 1980, at the age of 64 years, she experienced her first episode of amnesia. At 1 a.m., she felt unreal and was disoriented for place and time. She repeated the same questions over and over. She was unable to recognize her house or physician. Retrograde amnesia seemed to extend for several years. At 2 p.m., 4 hours later, neurological examination was otherwise normal. She progressively recovered but complained of a violent left-sided headache relieved by the administration of dihydroergotamine. Patchy left-sided arteries. She had complained of left-sided headaches since adolescence and her attacks were characterized by severe left-sided hemiplegic migraine.

In the second twin, both right-handed, had the same physical apearance, the same blood groups (O Rh+ C CDEe Cw– kk, MNS Lea+b– Pl+, Kpa– b+ Fya+b+ Jka+b+ Jsa– b+, Laa– b+) HLA determination (A2 A3 B35 BW60 BW6 R CW3 CW4) and the same caryotype variants. They have two other sisters, out of four, who also suffer from classic migraine.

Both the twins suffered transient global amnesia. In view of its low frequency, estimated at least to 2.75/100.000/year by Miller, a coincidence is improbable. To our knowledge only three familial observations have been reported and none included twins. In two reports, the patients were not migrainous and the clustering of cases was attributed to vertebro-basilar atheromatosis or to a particular susceptibility of infero-medial parts of temporal lobes. The third familial case concerned migrainous siblings, one having experienced transient global amnesia, the other a transient partial amnesia. The occurrence of transient global amnesia in our homozygotic twins suggests a genetic component.

Both twin sisters had suffered from mainly left-sided migraine since adolescence. Later, both of them experienced episodes of alexia, dysphasia, right hemianopsia, right-sided sensory disturbance followed by headache. According to Fisher’s criteria, these symptoms, the absence of carotid atheromatata and 13 years of benign evolution in at least one twin make them migrainous manifestations. The two transient global amnesia spells of the first sister were followed by left headaches and left EEG dyshrythmia, making a migrainous manifestation probable. The second twin had no headache after transient global amnesia.

Refereces

Letters


Muscle histology of hypocalcaemic myopathy in hypoparathyroidism

Sir: In addition to having the typical signs of idiopathic hypoparathyroidism, the patient described here showed features of hypocalcaemic myopathy, rarely reported previously in this condition.1–3

A 65 year old woman was admitted to hospital with complaints of intermittent carpopedal spasm (tetany) and easy fatigability. She had noted carpopedal spasm 20 years before. Five years before admission, she had had operations for bilateral carpal fractures. For the past year, she had had three droisy attacks associated with vomiting and cold sweating, each of which lasted for 5 to 10 minutes. There was no past history or family history of neuromuscular disease.

On physical examination, the patient was normal in stature and appearance. She had varicose veins and ecchymoses on both legs. The skin was dry and not infected. Neurological examination revealed hoarseness, mild dysphagia and moderate weakness predominantly affecting the proximal limb muscles. Muscle stretch reflexes were diminished with hypotonia. There was no sensory disturbance or incoordination. The WAIS score revealed a verbal IQ of 86. A typical carpal spasm appeared 30 seconds after a blood pressure cuff was inflated. The Chvostek sign was present on both sides.

On laboratory findings, serum calcium was low (5.0 mg/dl) and phosphorous 7.0 mg/dl. Unfortunately serum myoglobin level was not measured. The parathyroid hormone (PTH) concentration, 0.18 ng/ml, was less than normal. Reaction to exogenous PTH (Ellsworth-Howard test) was positive. Serum vitamin D3 level was 12 ng/ml (normal 14–42). The level of serum creatine kinase (CK) was high (756 IU/l, normal < 110) with mild elevation of lactic dehydrogenase and aldolase activity. The MB isozyme of CK was 5% and MM 95%. Electrocardiography showed a prolonged QTc of 0.58 second. Echocardiographic examination revealed cardiomyopathy and a pericardial effusion of 350 ml. Cranial computed tomography revealed bilateral symmetrical calcification of the basal ganglia and the dentate nuclei. Electroencephalography showed slowing of background activity without localising features. An electromyographic finding was an increase in polyphasic long-duration potentials in the distal part of the leg, but was normal in the proximal muscles. Motor nerve conduction velocities were normal.

The muscle biopsy specimen was taken from the quadriceps femoris muscle. Light-microscopical examination revealed variety in fibre size without fibre necrosis, vacuole or inflammatory cell infiltration. Routine ATPase reaction showed type 2 fibre atrophy (fig a). Electron microscopical examination showed perinuclear accumulation of mitochondria and focal myofibrillar degeneration. A few muscle fibres had concentric laminated bodies. In addition to these examinations, immunohistochemical localisation of myoglobin was studied. The formalin-fixed paraffin-embedded 3-μm-thick sections were stained using anti-myoglobin rabbit serum (1:200, DAKO, Denmark) by PAP method of Sternberger. In our patient, negative immunoreaction for myoglobin was observed in some muscle fibres, many of which correspond to atrophic type 2 fibres; positive reaction, on the contrary, was observed in the remaining fibres (fig b). In control patients with amyotrophic lateral sclerosis, almost all fibres, including atrophic type 2 fibres, were immunostained for myoglobin, though the intensity was varied. Normal muscle fibres of necropsied patients without neuromuscular diseases were also immunostained.

After administration of calcium and 1α-hydroxyvitamin D3, serum level of calcium and CK returned to normal values. Tetany, muscle weakness, easy fatigability, pericardial effusion and other symptoms and signs disappeared within 4 weeks.

The calcium ion plays many important roles in neuromuscular function. In hypocalcaemic patients, increased excitability of neuromuscular junction results in the well-known manifestation of tetany, mainly affecting the distal muscles. However, several authors1–6 have noted other manifestations: proximal muscle weakness, easy fatigability and elevated muscle-associated enzymes, and designated them as hypocalcaemic myopathy related to direct effect of hypocalcaemia on muscle fibres. Frame, however, pointed out that the elevated serum enzyme levels could be related to the tetany. In our patient, tetany appeared only two or three times a day, lasting for a few seconds, and serum CK activity did not fluctuate after the tetany of three minutes’ duration was induced by forearm ischaemia, suggesting that the tetany could be minimally related to the high CK activity.

Fig Quadriceps femoris. (a) Routine ATPase stain, demonstrating type 2 fibre atrophy (cryostat section, × 150). (b) Immunostaining for myoglobin, demonstrating negative staining of atrophic fibres (paraffin section, × 300).
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