SELF ASSESSMENT QUESTIONS

An unusual presentation of a common disorder

Gillian Hall, Brian Mc Namara, Julian Ray, Simon Boniface

Answers on p 376.

Addenbrookes Hospital, Cambridge, UK: Department of Neurology
G Hall

Department of Clinical Neurophysiology
B Mc Namara
J Ray
S Boniface

Correspondence to:
Dr Brian Mc Namara, Department of Clinical Neurophysiology Box 124, Addenbrookes Hospital, Cambridge CB2 2QQ, UK
(e-mail: b.mcnamara@medschl.cam.ac.uk)

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A 59 year old right handed man presented with a one week history of twitching of the right hand side of his face. This would spread to involve his right arm and hand. There was no background medical history of note and he was on no medication. On examination the right hand side of his face was continually twitching. Neurological examination did not reveal any further abnormalities. The results of investigations are shown in box 1.

Box 1: Investigations
- Computed tomography of the brain: normal
- Electroencephalography: focal slow wave activity over the left parietal region
- Urea 17.9 mmol/l
- Sodium 131 mmol/l
- Creatinine 97 mmol/l
- Potassium 4.7 mmol/l
- Serum osmolarity 313 mOsmol/l
- Glucose 31.4 mmol/l

Questions
(1) What is this uncommon epilepsy syndrome called?
(2) What is the connection between the abnormal biochemistry and the neurological presentation?
(3) In what proportion of cases of this syndrome will computed tomography or magnetic resonance imaging of the brain be abnormal?

Young male with recurrent flaccid quadriparesis and complete recovery

A S Kashyap, S Kashyap

Answers on p 376.

Armed Forces Medical College, Pune 411 040, India: Department of Medicine
A S Kashyap

Department of Hospital Administration
S Kashyap

Correspondence to:
Dr A S Kashyap

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A 25 year old male shopkeeper was brought to the emergency room with sudden onset flaccid quadriparesis of five hours' duration. The proximal muscle groups were weaker than the distal. He gave a history of similar episodes for the last four years. The episodes usually followed a period of rest after moderate to severe exertion. Each attack lasted three to 10 hours, and was followed by spontaneous complete recovery. The frequency varied from once per day to three times per week. There was no history of loss of consciousness, diplopia, drop attacks, dysarthria, dysphagia, alcohol, or drug intake. There was no history suggestive of hyperthyroidism. His parents and two sisters denied any history of similar neurological illness or hyperthyroidism. Clinically he had flaccid motor paralysis with absent deep tendon jerks. There was mild wasting of the thigh and arm muscles. Cranial nerves, fundi, higher mental functions, and sensations were normal. There was no goitre, tachycardia, tremor, or weight loss. Abnormal investigations at admission are listed in box 1.

Blood counts, creatinine, serum sodium, chloride, bicarbonate, calcium, glucose, cortisol, and supine aldosterone concentrations were normal. Nerve conduction velocities were normal.

Box 1: Investigation results (normal range)
- Potassium: 2.1 mmol/l (3.5–5.5)
- Free thyroxine 43 pmol/l (9–24), triiodothyronine 3.5 nmol/l (1.2–3.1), and thyroid stimulating hormone <0.01 mU/l (0.2–5 mU/l)
- Electrocardiography: normal sinus rhythm, flattened ST segment, presence of U waves, and prolonged QU interval
- Serum creatine kinase: 1190 U/l (10–190)
- Electromyogram: myopathic pattern
- Muscle biopsy: vacuolar myopathy

Questions
(1) What is the diagnosis?
(2) What is the basic pathophysiology of this disorder?
(3) How will you prevent further attacks?

Answers on p 377.

Swelling in the caesarean section scar

M Hussien, J W R Peyton

South Tyrone Hospital, Carland Road, Dungannon, County Tyrone, Northern Ireland BT71 4AU, UK
M Hussien
J W R Peyton

Correspondence to:
Mr Hussien (e-mail: maged@maged.fsnet.co.uk)

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A 38 year old women, para 3+1, presented with a two month history of painful swelling in her caesarean section scar. She had two caesarean sections seven and 10 years earlier. She had a long history of irregular menstrual periods.

On examination there was a tender, firm 2 × 2 cm swelling in the right side of the phannensteil incision scar. There was no cough impulse.

Questions
(1) What is the differential diagnosis?
(2) What investigations should you consider?
(3) How would you treat this patient?

An unusual case of chorea gravidarum

Answers on p 378.

A Qasim

St Mary's Hospital Portsmouth PO3 6AD, UK

Correspondence to:
Dr Qasim (e-mail: dr@asifqasim.freeserve.co.uk)

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A 24 year old right handed Pakistani woman who was eight weeks' pregnant presented with a two day history of involuntary movements of her left arm, hand, and foot. There was no significant past medical history or family history, and she was taking no regular medication. On examination there were choreiform movements of the left upper limb and left foot. Her speech was normal and there was no other neurological abnormality. She was apyrexial, and examination of the cardiovascular, respiratory, and abdominal systems was unremarkable. Fundoscopy and slit lamp examination of the eyes was normal. Full blood count, urea and electrolytes, liver function tests, and thyroid function tests were normal. She had a polyclonal rise in IgG, with no evidence of autoantibodies. C reactive protein was <6 mg/l and the antistreptolysin O titre 320. Electrocardiography and echocardiography showed no abnormalities. Obstetric ultrasound was performed which showed a viable fetus with estimated gestational age of 10 weeks. Magnetic resonance imaging (MRI) of the brain was normal. She was treated with haloperidol 1 mg three times daily and her chorea improved markedly over the next three days.

On day 8 she suffered a primary focal seizure with violent movements of the right arm leading to a secondary generalised seizure. Emergency computed tomography of the brain with and without contrast showed no abnormality. Cerebrospinal fluid obtained by lumbar puncture was sterile with normal cell counts. An electroencephalogram was asymmetrical with generalised excess of left sided theta and delta waves unaffected by photic stimulation. In the 24 hours after the seizure she developed expressive dysphasia, right sided hemiplegia, and worsening left sided chorea. MRI of the brain was performed on day 10 (fig 1). She was anticoagulated with intravenous unfractionated heparin, and within six hours there was an increase in right sided power to grade 4 out of 5 and improvement of her dysphasia and chorea. After bleeding from her vagina on day 27 miscarriage was diagnosed on obstetric ultrasound. She was discharged after 35 inpatient days: she had mildly reduced right sided power (grade 4 out of 5) and mild dysphasia. Life long treatment with warfarin is planned and she has been advised against future pregnancies.

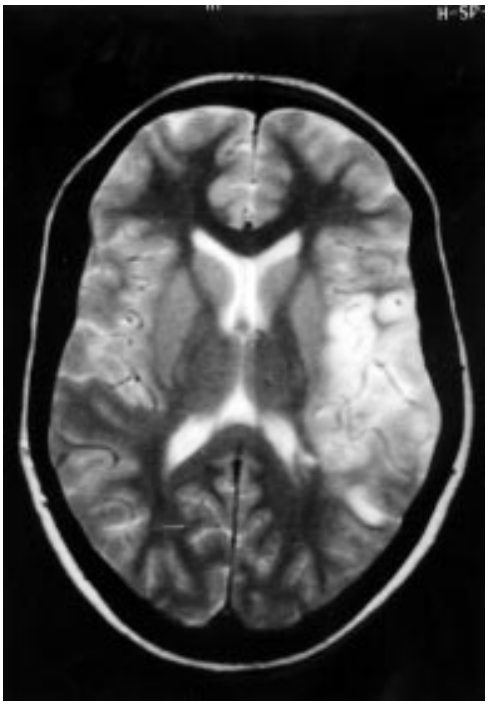

Figure 1 Axial image from MRI on day 10 after admission.

Questions
(1) Give a differential diagnosis of chorea in pregnancy
(2) Describe the abnormalities in the MRI image (fig 1)
(3) What underlying causes should be sought in this patient?

Nausea and vomiting, a cause for concern?

Aftab Ala, Natasha Arnold, Cho Cho Khin, Niall van Someren

Answers on p 379.

A 72 year old man with manic depression, who had recently started lithium treatment, presented to casualty with a two day history of severe nausea and vomiting. His serum urea and electrolytes were normal (sodium 139 mmol/l, potassium 4.1 mmol/l, creatinine 94 mmol/l, and urea 5 mmol/l) and he was discharged from the accident and emergency department with a presumptive diagnosis of gastroenteritis.

He represented 12 hours later with persistent vomiting and oliguric prerenal failure initially thought to be secondary to lithium toxicity. There was only minimal tenderness in the epigastrium but no signs of peritonism or abdominal masses; his pulse was 90 beats/min and blood pressure 105/50 mm Hg without a visible jugular venous pressure. His blood urea was now 28 mmol/l, sodium 141 mmol/l, potassium 4.3 mmol/l, and creatinine 310 mmol/l, with normal full blood count, blood gases, creatinine kinase, amylase, urine microscopy, a subtherapeutic lithium concentration 0.35 mmol/l (normal range 0.4–0.8 mmol/l), and an electrocardiograph showed sinus tachycardia only. His central venous pressure was low (−2 mm Hg) and he was resuscitated with fluids. Chest radiography was performed (figs 1 and 2) and a nasogastric tube was inserted but with some difficulty, draining only 100 ml of gastric juice. Twelve hours later his symptoms still had not improved. His renal function was unchanged and his vomitus had now become blood stained.

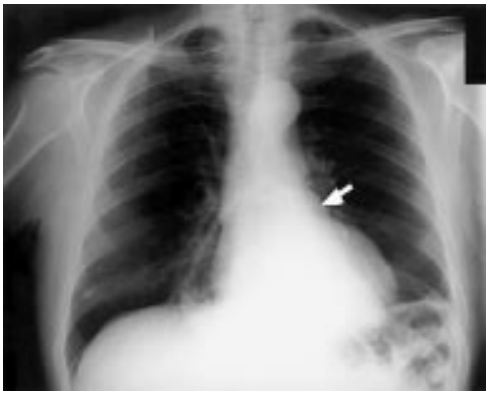

Figure 1 Erect posteroanterior chest radiograph. The fluid level (arrowed) represents a paraoesophageal hernia with a large gastric volvulus.

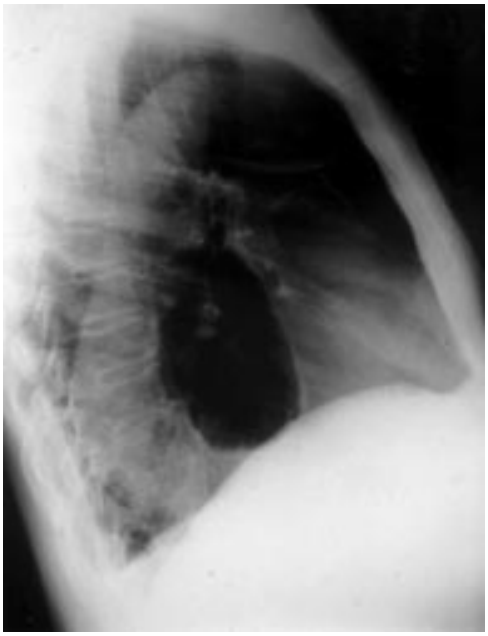

Figure 2 Lateral chest radiograph.

Questions

(1) What do the chest radiographs show?
(2) What single diagnostic investigation is required?

Department of Gastroenterology, Chase Farm Hospital, Enfield, Middlesex, UK
A Ala
N Arnold
C C Khin
N van Someren

Correspondence to:
Dr A Ala, Centre for Hepatology, Department of Medicine, Royal Free Hospital, Pond Street, London NW3 2QG, UK
(e-mail: aftab.ala@easynet.co.uk)

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SELF ASSESSMENT ANSWERS

An unusual presentation of a common disorder

Q1: What is this uncommon epilepsy syndrome called?
Epilepsia partialis continua (EPC).

Q2: What is the connection between the abnormal biochemistry and the neurological presentation?
Hyperglycaemia is a well recognised metabolic cause of EPC.[1]

Q3: In what proportion of cases of this syndrome will computed tomography or magnetic resonance imaging of the brain be abnormal?
50%–60%.

Discussion
EPC is defined as clonic muscular twitching repeated at fairly short intervals in one part of the body for a period of days or weeks. It is a form of focal status epilepticus manifesting as continuous focal motor seizures.[2] It is associated with a wide range of electroencephalographic abnormalities including focal spikes and focal slow waves[2]. However in some cases the electroencephalogram may be normal.[3] Causes of EPC include structural abnormalities such as central nervous system tumours, trauma, cerebral infarction, intracerebral haemorrhage, cerebral abscess, neuronal migration disorders, and vascular malformation.[2] However in up to 50% of cases conventional brain imaging may be normal.[3] Causes where brain imaging may be normal include hyponatraemia, hyperglycaemia, hepatic encephalopathy, and encephalitis.[2]

The association between hyperglycaemia and EPC is well recognised. In some cases patients have an underlying structural abnormality such as cerebral infarction. In others hyperglycaemia can cause EPC in the absence of any neurological disease. In these patients treatment of the seizures and reversal of the metabolic disorder prevents any long term neurological sequelae.[1] In the majority of patients with EPC associated with hyperglycaemia EPC occurs before impairment of consciousness and coma; it is important therefore to recognise this association. All patients presenting with EPC should have immediate determination of blood glucose concentrations. This could lead to the detection of previously undiagnosed diabetes mellitus and prompt reversal of the neurological condition. Our patient was treated with insulin, intravenous fluids, and a carbamezepine 400 mg daily. His seizures quickly resolved and he has not had any recurrence. Neurological examination at a follow up neurology outpatient clinic three months later did not reveal any abnormalities.

Final diagnosis
Epilepsia partialis continua, secondary to hyperglycaemia.

1 Singh BM, Strobos RJ. Epilepsia partialis continua associated with hyperglycaemia: clinical and biochemical profile of 21 patients. *Ann Neurol* 1980;**8**:155–60.
2 Schomer DL. Focal status epilepticus and epilepsia partialis continua in adults and children.. *Epilepsia* 1993;**34**(suppl 1):S29–36.
3 Cockerell OC, Rothwell J, Thompson PD, *et al*. Clinical and physiological features of epilepsia partialis continua (cases ascertained in the UK). *Brain* 1996;**119**:393–407.

Young male with recurrent flaccid quadriparesis and complete recovery

Q1: What is the diagnosis?
The diagnosis is thyrotoxic hypokalaemic periodic paralysis (THPP). This rare condition is seen in young Latin American and East Asian men with thyrotoxicosis. The clinical features of thyrotoxicosis are usually not apparent. The human leucocyte antigen types A2 Bw22 and Aw19 B17 increase the relative risk. The age of onset is usually before 30 years, and familial occurrence is unusual. The frequency of attacks varies from daily to yearly, and each attack lasts two to 12 hours. The attacks are precipitated by a high carbohydrate diet, sodium intake, emotional stress, and strenuous exertion followed by rest/sleep. It never occurs during vigorous physical activity. The higher mental functions and sensations are not affected. It is characterised by episodic, sudden onset, flaccid quadriparesis, with absent or hypoactive reflexes. The proximal limb weakness is more than the distal, and lower limbs are more prominently involved. The ocular, bulbar, and respiratory muscles are rarely involved. It responds to potassium administration. The frequency and severity of THPP episodes is reduced by β-adrenergic blockers, antithyroid drugs, and by avoidance of strenuous physical activity and a high carbohydrate diet. Rendering the patient euthyroid abolishes this condition. In the absence of effective treatment chronic progressive interattack weakness may develop. The muscle biopsy specimen often shows vacuolar myopathy.[1]

Q2: What is the basic pathophysiology of this disorder?
The basic pathophysiology in THPP is the development of hypokalaemia due to an intracellular shift of potassium. The total body potassium is normal. The cause of this shift is uncertain. Molecular defects in skeletal muscle calcium channels, leading to a decrease in activity of the calcium pump have been identified.[1] Calcium transport disturbances may adversely change muscle excitation-contraction coupling and account for acute paresis in THPP. The contractile apparatus is normal. Effects of insulin on potassium uptake in muscle suggest that an abnormality of muscle membrane may be involved. Increased