Self-assessment questions

A 60-year-old man with expressive dysphasia

N A McAndrew, T J Charles

A 60-year-old man (figure 1) was referred with an incidental abnormality in the right lower zone of his chest X-ray. He was otherwise well. Forty years previously a segment of his left lower lobe had been removed for a benign lesion. Two siblings had died from intracranial haemorrhage. High-resolution computed tomography (CT) scan of the thorax (figure 2) was performed. Before any treatment was given for the lesion visible in figure 2 he was admitted to hospital with right upper motor neurone facial weakness and expressive dysphasia of 2 days duration. CT scan of the head was carried out (figure 3).

Questions

1. What disorder does this patient have and which feature of this does figure 2 demonstrate?
2. What complication has he now developed and how is this related to the lesion in figure 2?
3. What are the options for treatment of the pulmonary lesion?
Answers

QUESTION 1
This patient has hereditary haemorrhagic telangiectasia (HHT) and the high-resolution CT images show a pulmonary arteriovenous malformation (AVM) with its feeding vessels in the lateral part of the right lower lobe.

QUESTION 2
Cerebral abscess. The pulmonary AVM lacks capillaries and as such represents a direct connection between the pulmonary artery and vein providing a right-to-left shunt allowing paradoxical embolisation. Septic emboli or possibly a transient bacteraemia infecting existing cerebral infarcts are thought to result in the formation of a cerebral abscess.

QUESTION 3
Simple observation, surgical removal, and embolisation. Our patient had his cerebral abscess drained with excellent results and subsequently had his pulmonary AVM embolised.

Discussion

HHT is characterised by mucocutaneous telangiectases, recurrent epistaxis and a family history of the disorder. Vascular abnormalities also occur in the lungs, gastrointestinal tract and central nervous system. It is inherited in an autosomal dominant fashion with variable penetrance. Whilst it has been divided into types I and II on the basis of established linkages to the long arms of chromosomes 9 and 12, respectively, this is almost certainly an oversimplification.

Mutations on chromosome 9 (HHT I) affect endoglin, a protein found on vascular endothelial cells. Endoglin binds transforming growth factor beta, which affects endothelial cell migration, proliferation and adhesion and the composition and organisation of the extracellular matrix. The activin receptor-like kinase (ALK1) affected by mutations on chromosome 12 (HHT II) is also found on vascular endothelial cells although its role is unclear. The mechanism by which these mutations cause the angiodysplastic lesions of HHT has yet to be established.

Approximately 15% of people with HHT will have a pulmonary AVM and, conversely, 60–90% of people with a pulmonary AVM will have HHT. It appears that HHT I carries an increased risk of both pulmonary and cerebral AVMs when compared with HHT II. Two-thirds of AVMs occur in the lower lobes, predominantly basally and often multiple. The risk of hypoxia, polycythaemia and haemorrhage with a pulmonary AVM is perhaps better appreciated than is the risk of cerebral ischaemic events, the incidence of the latter being up to 40% in patients with a pulmonary AVM, with a risk of cerebral abscess of 10–15%. Indeed, cerebral ischaemic events are often the first manifestation of both the pulmonary AVM and HHT.

Plain chest radiography and high-resolution CT will detect the majority of pulmonary AVMs. Other methods of diagnosis include angiography, radionuclide shunt studies, and contrast echocardiography. A fall in oxygen saturations between supine and erect postures (due to redistribution of blood through the predominantly basal pulmonary AVM) may also be seen.

The risk of neurological events suggests that pulmonary AVMs should not be treated conservatively, ie, not simply observed. Embolisation, by coil or balloon, has distinct advantages over surgery with lower morbidity (∼15%), lower mortality (<1%) and shorter hospital stay. Embolisation can also be performed on multiple lesions without the loss of substantial lung parenchyma. Closure of larger malformations may lead to enlargement of smaller lesions with time and long-term follow-up is required. Screening families with HHT for pulmonary AVM has been advocated but the best method for doing this is debated. It has been suggested that cerebral abscess and pulmonary AVM be used as markers for each other, but perhaps pulmonary AVM should also be considered when the young person with a stroke is assessed.

Final diagnosis

Cerebral abscess in a patient with HHT and a pulmonary AVM.

Keywords: hereditary haemorrhagic telangiectasia; pulmonary arteriovenous malformation; cerebral abscess; embolisation

Learning point

Pulmonary AVMs have a high rate of complications and are best treated with embolisation

A case of sympathetically maintained pain

A E K Ibrahim, D Pounder, P M Perry

A 47-year-old woman sustained a gunshot wound to the left thigh. The entry wound was on the anteromedial aspect and the exit wound was on the posterolateral aspect of the thigh. She presented a month later with burning, predominantly superficial pain and cold hyperalgesia affecting part of the left leg and foot in a stocking distribution. She had full pulses bilaterally, numbness to both pin prick and light touch on the sole and lateral aspect of the left foot, wasting of the calf muscles, weakness of dorsiflexion, and cold leg.

Temperature profiles were requested (figures 1 and 2). Magnetic resonance imaging (MRI) scan of the left thigh showed evidence of the previous gunshot wound with the sciatic nerve identified in continuity but deviated at the site of the bullet track and surrounded by scar tissue. The patient had some relief of her symptoms following a left lumbar sympathetic block using a local anaesthetic, but continued to take oral analgesics.

Questions

1 What abnormality do the temperature profiles show, and what is the condition called?
2 What is the definition, and what are the diagnostic criteria for this syndrome?
3 What are the former names for these syndromes, and how do they relate to the term ‘sympathetically maintained pain’?
Answers

QUESTION 1

The temperature profile (figure 1) shows a drop in temperature affecting the left leg, while (figure 2) shows the improvement in the temperature profile following treatment. The underlying condition is Complex Regional Pain Syndrome, type II (CRPS type II).

QUESTION 2

CRPS type II is defined as burning pain, allodynia (pain from innocuous mechanical or thermal stimuli), and hyperpathia (exaggerated subjective response to painful stimuli with continuing sensation of pain after stimulus has ceased) usually in the hand or foot after partial injury of a nerve or one of its major branches. The diagnostic criteria are:

- the presence of continuing pain, allodynia, or hyperalgesia (lowered pain threshold and enhanced pain perception) after a nerve injury not necessarily limited to the distribution of the injured nerve
- evidence, at some time, of oedema, changes in the skin blood flow, or abnormal sudomotor activity in the region of the pain
- this diagnosis is excluded by the existence of conditions that would otherwise account for the degree pain and dysfunction.

All three criteria must be satisfied.1

QUESTION 3

The International Association for the Study of Pain, in its second edition on the taxonomy of pain, has replaced the term reflex sympathetic dystrophy with a new term, CRPS type I, while the term causalgia has been replaced by the term CRPS type II.

Sympathetically maintained pain is a type of pain that is maintained by sympathetic efferent innervation or by circulating catecholamines and is a feature of CRPS syndromes and several other types of painful conditions, but it may exist as an entity, not associated with any other condition.2

Discussion

The complications of CRPS are serious and include disuse atrophy of the involved limb, complete disruption of normal daily activity by severe pain, risk of suicide, and drug abuse.3 While a few patients experience spontaneous remission without proper treatment, the majority become progressively worse. Therefore, it is of great importance that the diagnosis of CRPS be made promptly, so that treatment can be started without delay.4 5

The conventional method of treatment for CRPS is sympathetic nerve blockade. This is performed at the level of L2 for the lower limbs.6 Other treatment modalities (listed in no particular order) include percutaneous epidural spinal cord stimulation,7 intravenous regional guanethidine,8 oral nifedipine,9 transcutaneous nerve stimulation,10 and prostaglandin E1 ointment.11

Final diagnosis

Complex regional pain syndrome type II.

Keywords: complex regional pain syndrome; pain; causalgia

Skin fragility and abnormal liver function tests

S Chitturi, M Dakkak, A P Campbell

A 53-year-old man, a worker in a plastic factory, presented with a 12-month history of a non-itchy skin rash on the dorsum of both hands, associated with increased skin fragility and tendency to recurrent scar formation. The biochemical profile revealed an isolated elevation of serum alanine aminotransferase 55 IU/l (normal 5–36 IU/l). Clinical examination did not reveal any stigmata of chronic liver disease. He denied any history of jaundice, blood transfusions, intravenous drug use or family history of skin or liver disease. His alcohol intake was in the range of three bottles of wine a week. Hepatitis B surface antigen and hepatitis C antibodies (ELISA) were not detected. Serum copper, caeruloplasmin and alpha-1-antitrypsin levels were normal. No serum auto-antibodies were detected. Subsequently, a liver biopsy was performed (figure).

Questions

1. What does the liver biopsy show?
2. What is the underlying skin disorder?
3. How is the condition treated?

Figure  Liver biopsy (Perls stain, orig × 400)

Self-assessment questions

**Answers**

**QUESTION 1**
The figure is a high-power view of a portal tract with the adjacent hepatic parenchyma. The striking feature is the presence of dark-staining granules, representing haemosiderin deposition, both in the parenchymal and the stromal cells. There is no evidence of cirrhosis. The overall appearance is consistent with haemochromatosis.

**QUESTION 2**
The underlying skin disorder is sporadic porphyria cutanea tarda (PCT).

**QUESTION 3**
Phlebotomy is the treatment of choice. Abstinence from alcohol is an important adjunct measure. An alternative but less effective form of treatment is low-dose chloroquine.

**Further investigations**
The features of iron overload were confirmed in our patient. Serum ferritin was 1878 µg/l (normal 30–400), serum iron 41 µmol/l (8–34) and serum transferrin 1.9 g/l (2–3.2). Porphyria investigations revealed the following. In urine, markedly elevated uroporphyrin levels at 3244 nmol/24 h (0–50), and normal coproporphyrin levels at 414 nmol/24 h (0–430). In faeces, a normal protoporphyrin level at 80 nmol/g dry weight (0–200), and marginally raised coproporphyrins at 80 nmol/g dry weight (0–76). Thin-layer chromatography of urine and faecal extracts revealed similar changes. In addition, the presence of 5,6,7-carboxylic porphyrins (urine, faeces) and isocoproporphyrins (faeces) was detected. A test for erythropoietic protoporphyria was negative.

**Discussion**
The porphyrias are metabolic disorders arising from enzymatic defects in the haem biosynthetic pathway. Their clinical presentation depends on the predominant form of porphyrin precursor accumulation. They may present as isolated cutaneous lesions, neuropsychiatric disorders, or both.

PCT is probably the commonest type of porphyria worldwide. However, there are striking geographical variations. There are three types: the common sporadic (type I) or the familial types (types II and III). The underlying metabolic defect is a reduction in the activity of the enzyme uroporphyrinogen decarboxylase. In the familial types, the reduction in enzyme activity is confined to the liver. Clinical and experimental studies suggest that this enzyme is reversibly inactivated by an iron-dependent process. The cutaneous manifestations of PCT include photosensitivity, recurrent blister formation, hypertrichosis, hyperpigmentation and scarring. This may result in a psudosclerodematous appearance. Features suggestive of acute intermittent porphyria such as severe abdominal pain and drug sensitivity (apart from alcohol and oestrogens) are notably absent. Hepatomegaly, raised serum bilirubin and aminotransferase levels may occur, especially in alcoholics. A striking association with hepatitis C has been noted in some but not all studies. The presence of hepatitis C may explain some of the pathological changes encountered in the liver, but the role of the hepatitis C virus in the pathogenesis of sporadic PCT is uncertain.

Mild-to-moderate hepatic iron overload is often present in patients with PCT. In less than 10% of patients, this may be in the range usually associated with hereditary haemochromatosis. The relationship between sporadic PCT and hereditary haemochromatosis has been re-examined in the light of the discovery of the probable haemochromatosis gene (HLA-H). Indeed, a significant association has been noted between one of the causative mutations (Cys282Tyr) in the HLA-H gene and sporadic PCT. This mutation is responsible for much of the iron overload in populations of European descent. It has been suggested that, in countries where this mutation is common, patients with PCT should be screened for its presence. Homozygotes thus identified should be treated and their families should be screened for haemochromatosis.

Phlebotomy is the treatment of choice in patients with PCT. This achieves a reduction in the hepatic iron stores. The iron ‘load’ in PCT is not usually marked (in the range of 3–5 g). Phlebotomy is performed once or twice weekly initially. Lasting clinical remission is often achieved after only five or six sessions of phlebotomy. Serum ferritin should be monitored to prevent the development of iron deficiency. Low-dose chloroquine (125 mg) is an alternative form of treatment for patients who cannot tolerate phlebotomy. Avoidance of precipitating factors like alcohol and oestrogens is important.

**Final diagnosis**

Sporadic porphyria cutanea tarda and haemochromatosis.

**Keywords:** porphyria cutanea tarda; haemochromatosis; skin fragility

Syncope, the head ruling the heart

Simon J Hickman, Barrie D White

A previously fit 53-year-old man was referred for investigation of episodes of syncope and pre-syncope. There was no history of convulsions and after one such episode his pulse rate was noted to be 44 beats/min. Examination was unremarkable. An electrocardiogram and echocardiogram were normal. Repeated 24-hour Holter monitoring did not show an arrhythmia during a collapse although his resting pulse rate fell to 39 beats/min.

Over the next few weeks he deteriorated, complaining of unsteadiness, poor short-term memory and frequent headaches. The attacks continued on a daily basis and were associated with urinary incontinence. His gait became broad-based and unsteady but otherwise neurological examination remained normal.

He was admitted after a further attack followed by confusion. On admission he had a Glasgow Coma Score of 14/15, was apyrexic, had a pulse rate of 84 beats/min and a blood pressure of 120/70 mmHg. Papilloedema was noted but there were still no focal neurological signs. Over the next 24 hours he deteriorated to a Glasgow Coma Score of 8. His pulse rate decreased to 40 beats/min and his blood pressure increased to 150/100 mmHg. A computed tomography (CT) scan (figure 1) and subsequent gadolinium-enhanced magnetic resonance imaging (MRI) scan (figure 2) were performed.

Questions

1. Where is the lesion and what effect is it having?
2. What are the ways in which lesions in this area might present?
3. What is the differential diagnosis?
4. Describe the cardiovascular response to his decreased conscious level.
Answers

QUESTION 1
The lesion is blocking the third ventricle and causing acute obstructive hydrocephalus (early periventricular lucencies can be seen on the CT scan, ie, cerebral oedema due to transudation of fluid).

QUESTION 2
Intraventricular tumours causing intermittent or permanent raised intracranial pressure have been reported to present with ataxia, memory problems, headache, vomiting, visual field defects and mental disturbance.1 Intermittent cerebrospinal fluid (CSF) obstruction due to these tumours is frequently silent and may present with sudden death.2

QUESTION 3
The differential diagnosis of a third ventricular mass includes craniopharyngiomas,1 colloid cysts,7 choroid plexus papillomas, ependymomas, meningiomas, gliomas,3 and, very rarely, posterior communicating artery aneurysms.4

QUESTION 4
The decreased conscious level is due to critically raised intracranial pressure. The presentation with bradycardia and hypertension was described by Harvey Cushing in 1902 and is called the Cushing response. In his experiments Cushing infused saline into the subdural spaces of dogs via a cannula inserted through a trephine opening in the skull and dura. When the intracranial pressure exceeded the arterial blood pressure the arterial blood pressure increased in association with bradycardia and slowing of respiration. The response was abolished by cocaineization of the medulla. The hypertensive response was prevented by section of the spinal cord and hence the descending sympathetic pathways. Vagotomy prevented bradycardia. He felt therefore that there was a regulatory mechanism in the vasomotor centre which maintained medullary perfusion.1 In clinical practice, the Cushing response is rarely seen except as an agonal event.

Learning points
- third ventricular tumours can cause intermittent and permanent CSF obstruction
- presentation can be with syncope, ataxia, memory disturbance, visual field defects, raised intracranial pressure and sudden death
- the triad of raised intracranial pressure is classically headache, vomiting and papilloedema
- critically raised intracranial pressure can present with the Cushing response of bradycardia, hypertension and slowing of respiration
- although third ventricular tumours are rare, complete cure is possible with surgical resection, hence rapid diagnosis is very important

Progress
After the CT scan neurosurgical referral was made. He underwent emergency external ventricular drainage which returned his Glasgow Coma Score to 15. MRI scanning to delineate the anatomy further was followed by craniotomy and macroscopic resection of the tumour with decompression of the third ventricle. Histological examination confirmed the tumour to be a craniopharyngioma.

In the immediate post-operative period the patient had a moderate memory deficit and pan-hypopituitarism necessitating hormone replacement. On follow-up 6 months later he was well, mobilising without ataxia and had improving memory. A post-operative scan confirmed that the tumour had been completely excised.

Final diagnosis
Third ventricular craniopharyngioma causing obstructive hydrocephalus.

Keywords: syncope; cerebral ventricle neoplasms; craniopharyngioma; Cushing response

Tachycardia following low-tension electrocution

S Orme, K S Channer

A 33-year-old woman sustained an electric shock to the dorsum of her right hand from a domestic power supply. The only symptom she reported was a transient numbness in her right arm. There was no loss of consciousness. On arrival at the accident and emergency department she was well perfused and alert. Examination of her skin revealed no superficial burns. She had a regular tachycardia at a rate of 150 beats/min, and her blood pressure was 120/70 mmHg. The remainder of the examination was normal. Her admission electrocardiogram (ECG) is shown in figure 1. The patient was given 3 mg and then 6 mg of adenosine intravenously without effect; the outcome of 12 mg of adenosine is shown in figure 2.

Further investigations included serial creatinine kinase (myocardial bound fraction) and troponin I levels to exclude myocardial damage. These were within normal limits. The lung fields were clear on chest X-ray. An echocardiogram showed normal ventricular and atrial dimensions and function and no valvular abnormality.

Questions

1. Describe the features shown in the ECG (figure 1).
2. What does the rhythm strip in figure 2 show, and what information does this give us about the mechanism of the tachycardia?
Answers

QUESTION 1
The ECG shows a narrow complex tachycardia with retrograde P wave activity. The P wave axis indicates that is a junctional tachycardia.

QUESTION 2
The rhythm strip shows normal P wave activity followed by a few sinus beats before the junctional rhythm returns. As the tachycardia was temporarily abolished by adenosine, it must involve the AV node.

Outcome

The following day, as the patient remained in the junctional tachycardia, she was given 25 mg of atenolol which led to a restoration of sinus rhythm and she was discharged home.

Discussion

Electrocution can be subdivided into low-tension shocks and high-tension shocks. It is well recognised that electrical injury may cause immediate cardiac arrhythmias. High-tension shocks (>1000 Volts) from lightening strikes and overhead power cables cause severe skin burns and may cause cardiac arrest usually due to asystole.1 High-voltage injuries lead to a high incidence of cardiac abnormalities, up to 30% of those with cardiac arrhythmias on presentation have long-term reduction in cardiac function measured by echocardiography.2 Changes in cardiac conduction including complete heart block, and prolongation of the QT interval suggest that electric shocks can significantly modify intracardiac conduction.3

Low-tension shocks which occur from domestic appliances (<300 Volts at 50Hz) can cause ventricular fibrillation,1 but the incidence of cardiac arrhythmias following low-tension electrocution is not known. One retrospective study demonstrated that those patients with a normal ECG on presentation did not subsequently develop a serious arrhythmia.1 However, there is an appreciable risk of late ventricular fibrillation in injuries involving the passage of current through the thorax. In these cases, myocardial biopsies have shown patchy myocyte necrosis and fibrosis.1

The likelihood of tissue injury and cardiac arrhythmias following electrocution is dependent on the size of the current flow. For example, an alternating current of approximately 70 mA is required to induce ventricular fibrillation. Current flow is predominantly influenced by the voltage of the shock and resistance of the tissues involved. These principles are expressed by Ohms law:

\[ \text{Current} = \frac{\text{Voltage}}{\text{Resistance}} \]

Bone has the highest resistance of the internal structures of the body, followed by fat. As electrical wounds enter through the skin, the resistance of the skin at the time of the injury contributes significantly to the total body resistance. Therefore, an electrical shock on wet skin has potential to cause more serious damage to tissues.5

Patients should be admitted to hospital and monitored in all cases of high-tension electrocution because of the high incidence of myocardial damage. In low-tension injuries, admission and monitoring of cardiac rhythm is necessary if the admission ECG is abnormal or if there is any suggestion that the passage of electrical current was through the thorax. This may be inferred from any superficial burns at the entry and exit site or from the clinical history. In our patient, the initial ECG precipitated admission for further monitoring.

Final diagnosis

Junctional tachycardia following low-tension electrocution.

Keywords: electrocution; tachycardia

Summary points

- electrocution can be divided into high-tension (>1000 V) or low-tension injury (<350 V)
- there is a high incidence of cardiac damage with high-tension injuries
- low-tension injuries may cause ventricular fibrillation
- patients with high-tension injuries or low-tension injuries with an abnormal ECG or passage of current through the thorax should be admitted to hospital for cardiac monitoring

References

Collapse and hypothermia in an elderly woman in early summer

R O Morris, A Jain, K A Sands, J Snape

A 74-year-old woman was admitted during a warm spell, having been found at home collapsed. She was conscious but confused (Abbreviated Mental Test score (AMTS) 4/10). Her sister related her medical history which included an underactive thyroid diagnosed 8 years before, an operation to close an atrial septal defect as a young woman, a small stroke and several admissions over the last 3–4 years with difficulty coping and confusion. In fact, she had been told by one doctor that her sister had dementia. The patient was taking digoxin 62.5 µg, co-amilo fruse 2 tablets, and thyroxine 50 µg, all once daily. On examination she was pale and drowsy and her rectal temperature was 33.6°C. Body hair was absent. The pulse rate was 60 beats/min, irregularly irregular, and her blood pressure was 120/60 mmHg. Although the heart was clinically enlarged, auscultation was normal. There were fine crackles audible at the lung bases. Apart from the cognitive impairment, central nervous system examination was normal.

Initial investigations showed haemoglobin 10.4 g/dl (mean cell volume 88 fl), white cell count $3.9 \times 10^9/l$, platelets $88 \times 10^9/l$, urea, electrolytes and thyroid function tests were normal. Plasma glucose was 1.4 mmol/l. An electrocardiogram revealed slow atrial fibrillation with a prolonged QT internal and right axis deviation. A chest X-ray confirmed cardiomegaly. The patient was given 50 ml of intravenous 50% dextrose with some improvement in her confusion and she was slowly re-warmed. She was then able to eat and drink. The following day her serum sodium was 123 mmol/l and blood glucose 2.0 mmol/l. Her old notes at this time revealed that during the admissions with confusion and not coping over recent years, her sodium had often, and her glucose had occasionally, been low. The notes also confirmed that she had had primary hypothyroidism diagnosed 8 years before. Computed tomography (CT) was performed and, following injection of contrast medium, a coronal image through the pituitary fossa obtained (figure).

Questions

1. What further investigations would confirm the diagnosis and what is the significance of the CT findings?
2. Why did she present with hypothermia?
3. What is the most probable mechanism causing the hypoglycaemia?
4. What is the cause of the hyponatraemia?
**Answers**

**QUESTION 1**
The further investigations which were carried out are shown in the table.

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The diagnosis is hypopituitarism in association with an empty sella. The patient also has primary hypothyroidism. The empty sella syndrome (ESS) may be primary (idiopathic) or secondary. Idiopathic ESS is thought to arise as a consequence of congenital incompetence of the diaphragma allowing herniation of the arachnoid mater and cerebrospinal fluid (CSF) into the fossa, ultimately compressing the pituitary gland. This is most commonly seen in middle-aged obese women and may be associated with raised intracranial pressure but endocrine dysfunction is rarely observed. In secondary ESS the meninges and CSF herniate downwards to fill the void created by pituitary involution as a result of other pathology, eg, surgery, irradiation, infarction, haemorrhage or tumour necrosis. Although the occurrence of ESS in this patient may be an incidental finding, it is possible that she developed secondary ESS as a consequence of thyroxine-induced shrinkage of thyrotroph cell hyperplasia following treatment for her primary hyperthyroidism. This effect has been previously reported.

**QUESTION 2**
The patient may have been hypothermic because thyroid and adrenal hormones are involved in non-shivering thermogenesis and deficiency of one or both may predispose to hypothermia. Hypoglycaemia is thought to predispose to hypothermia independently of insulin, either as a direct effect or secondary to a glycopenic effect on the thermoregulatory centre. Pituitary disorders, through anatomical proximity to the thermoregulatory centre, may cause disturbances in thermoregulation. The patient's age may also have predisposed to hypothermia.

**QUESTION 3**
Deficiencies in the anti-insulin hormones: adrenocorticotropin and growth hormone are responsible for the hypoglycaemia seen in hypopituitarism. It was felt that the patient's periods of confusion were related to episodes of hypoglycaemia.

**QUESTION 4**
Cortisol deficiency results in increased secretion of vasopressin and dilutional hyponatraemia develops as a consequence. Renal water excretion is also impaired independent of this antidiuretic effect because of decreased tubular fluid delivery to the diluting site. This is in contrast to the salt-wasting state seen in Addison's disease.

**Outcome**
The patient responded well to treatment with hydrocortisone. In particular, her cognitive function improved to an AMTS of 9/10 and has been maintained at that since.

**Final diagnosis**
Hypopituitarism in association with empty sella syndrome.

**Keywords:** hypopituitarism; empty sella syndrome; hypothermia; hypoglycaemia

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A 60-year-old man with expressive dysphasia

N A McAndrew and T J Charles

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