was fully alert. Serum and urinary osmolarity became normal, urinary specific gravity was 1005-1025. She recovered from ovarian hyperstimulation syndrome and laparotomy during the next month.

Brain MRI and CT performed during the next five years were normal, as were repeated neurological and psychiatric examinations. The patient’s IQ was 126-130.

Severe seborrheic with ascites and hydrothorax due to ovarian hyperstimulation syndrome and haemoperitoneum due to tubal pregnancy, with hypovolaemia, anaemia, and hypotension serum concentra-

tions masked an SIADH that was heralded by seizures, followed by a prolonged lethergic state. Collateral evidence of SIADH was obtained by normal creatinine clearance1 with urine hyperconcentration. The symp-

December 2014

...thrombocytopenia followed by new episodes of encephalopathy with brain oedema. Hypoxic-

An 88-year-old woman with a history of hypertension, diabetes mellitus, and dyslipidaemia, was admitted to the hospital with fever, nausea, vomiting, and altered mental status. She had a history of transient ischemic attacks and a stroke 2 years prior. On admission, her vital signs were: temperature 38.5°C, blood pressure 150/90 mmHg, heart rate 100 beats/min, respiratory rate 20 breaths/min, and oxygen saturation 98% on room air. Her physical examination revealed a non-pulsatile neck and diffuse neck pain. Her neurological examination was remarkable for left-sided weakness and unsteady gait. Her initial laboratory findings showed a white blood cell count of 12,500/µL, hemoglobin 11.0 g/dL, hematocrit 33.0%, platelets 150,000/µL, sodium 139 mEq/L, potassium 4.5 mEq/L, chloride 102 mEq/L, bicarbonate 22 mEq/L, blood urea nitrogen 15 mg/dL, serum creatinine 1.2 mg/dL, aspartate aminotransferase 19 U/L, alanine aminotransferase 16 U/L, total bilirubin 0.9 mg/dL, direct bilirubin 0.2 mg/dL, creatinine kinase 79 U/L, troponin T 0.03 ng/mL, and C-reactive protein 2.3 mg/L. The patient was started on intravenous fluids, antibiotics, and dexamethasone 4 mg/day. She was transferred to the ICU for further management.

During the next month, her mental status improved and she was discharged home with a diagnosis of probable subarachnoid hemorrhage. Follow-up imaging studies, including brain MRI and CT, were unremarkable. She made a good recovery and was discharged home with a normal neurological examination.

Letters to the Editor

Thyroid Hashimoto’s encephalopathy

Thyroid disease is associated with several neurological disorders, of which one of the rarest and least well understood is

Hashimoto’s encephalopathy. This was origi-
nally postulated to be a distinct disease entity by Brain et al in 19662 and there have subsequently been case reports substanti-
ating the hypothesis that it represents a unique condition.3,4 The characteristic features are a subacute onset of confusion with altered consciousness, seizures, and movements or events that respond to steroids and which occur in the context of high anti-microbial antibody titers.5 To date all the patients reported have been either euthyroid or hypothyroid at the time of presentation. We present the case of a patient with encephalopathy with pronounced thyrotoxy-

osis, that was successfully managed with steroids, carbimazole, and propranolol.

A 49-year-old woman presented with a six month history of weight loss and a three month history of proximal arm pain and hand tremor. Two weeks before admission she developed a progressive left sided weakness involving the arm and leg in conjunc-

tion with a left hemianesthesia. On examination at admission she was flushed, feverish, and tachycardic with a hyperdynamic circulation. Her thyroid gland was slightly enlarged but there was no associated bruit. Cranial nerve examination disclosed left visual inattention as the only abnormal-

ity. Limb examination showed a moderately severe hypokinesia on left side. There was hyperreflexia, and bilat-

eral extensor plantar responses. She had wasting of the shoulder girdle muscles and adhesive capsulitis of the shoulder joints bilaterally. In the days immediately after admission there was further muscle wasting and had florid visual hallucinations, while independently having runs of paroxysmal atrial fibrillation. As a result of the original negative findings (see later) dexamethasone (12 mg/day) and acyclovir were started with the presumptive diagnosis of an encephalitis or vasculitis. On this regime she made a dra-

matic improvement, which was further enhanced by the treatment of her thyrotoxy-

osis on receipt of her thyroid function tests. The introduction of carbimazole and propranolol was then followed by a reduction in the dexamethasone and a cessation of the acyclovir. Attempted steroid weaning over subsequent days provoked a recrudescence of her fluctuating multifocal encephalopathy. One possibility is demyelination, which can virtu-

ally be discounted on the basis of our results as both MRI and CSF were normal. More likely explanations are either a multifocal abnormality of cerebral perfusion or a patchy defect of the autoimmunity.

This patient completes the repertoire of thyroid states seen in related thyroid encephalopathies and emphasises the need to assess thyroid function and autoantibody status in patients presenting with encephalopathy and stroke-like events in the absence of structural or infectious aetiologies.

Letters to the Editor

Thyroid Hashimoto’s encephalopathy

Thyroid disease is associated with several neurological disorders, of which one of the rarest and least well understood is
Letters to the Editor
wasting after aneurysmal subarachnoid hemorrhage who showed remarkable blood in the suprasellar cisterns and the left Sylvian fissure. Two days later she developed extreme polyuria which was compatible with a subarachnoid hemorrhage in the left Sylvian fissure. The patient regained consciousness and she gradually recovered from a mild aphasia and right facial weakness. However, from day 12 onwards she again developed a progressive polyuria of up to 21 200 ml per day (on day 22) and a 24 hour renal sodium loss of 2630 mmol. The sodium concentration in plasma was between 128 and 142 mmol/l, and the colloidal osmotic pressure was between 18.7 and 24.0 mm Hg. Serum ADH concentrations were normal. Treatment with furosemide had no effect on renal sodium loss. Despite the marked polyuria, plasma atrial natriuretic protein concentrations were within the normal range (up to 11.1 pmol/l), normal 3–23 pmol/l); atrial natriuretic protein in CSF was not assessed. Daily transcranial Doppler sonography was indicative of cerebral vasospasm and therefore angiography was performed on day 12 and again on day 24. The left middle cerebral artery was disclosed, which was successfully clipped on day 24. Whereas the diuresis 24 hours before and after the neurosurgical procedure was 600–700 ml/hour, the mean intraoperative diuresis was unchanged in paraffin sections. The arteriopathy of CADASIL is apparent from the age of 35. At the age of 40 she had MRI changes similar to the index patient, and one had had recurrent episodes of aphasia, headache, and hemianopia. Six members of this family, three affected and three healthy, have been genotyped with eight chromosome 19 markers spanning the CADASIL locus. No differences were found with any of those markers. Maximum lod scores were obtained with markers D19S226, D19S253, and D19S199, strongly suggesting that this family is linked to the CADASIL locus.

A patient with cerebral autosomal dominant arteriopathy with subcortical infarcts and leucoencephalopathy (CADASIL) confirmed by sural nerve biopsy

“Cerebral autosomal dominant arteriopathy with subcortical infarcts and leucoencephalopathy” (CADASIL) is a newly defined syndrome characterised, in the absence of hypertension, by recurrent subcortical ischaemic strokes and by peculiar non-amyloid, non-arteriosclerotic angio­pathy of cerebral vessels. On MRI circum­scribed subcortical ischaemic lesions and diffuse areas of leukoaraiosis are seen both in symptomatic and asymptomatic family members. Recently, genetic linkage analysis in two unimpaired families has assigned the disease locus to chromosome 19q12 with the most likely location of the disease gene between D19S221 and D19S222.

A few postmortem studies have been reported, showing predominant involvement of the cerebral white matter with diffuse myelin loss, multiple small deep infarcts, and occasional haemorrhages. As first reported by Baudrimont et al., the small subcortical and leptomeningeal arteries and arterioles display fibrous thickening and an eosinophilic, perivascular PAS positive, granular material in the muscle layer. Electron microscopy shows swollen myocytes in the media surrounded by collagen, elastin, and a compact electron dense material.

The arteriopathy of CADASIL is apparently not restricted to brain vessels as identical vascular lesions have been found in small myocardial arteries and sural nerve.

We present a 55 year old woman with a history of recurrent pulmonary embolism from the age of 55. At the age of 40 she experienced a feeling of heaviness in her left arm for about two days. Fifteen years later the patient described episodes of a burning sensation on her tongue and tinging as well as weakness of the left side of her face and her left arm. Six months later she complained of numbness and weakness of her left arm and leg, from which she recovered slowly. No risk factors such as arterial hypertension, diabetes, or migraine were reported. Neurological examination showed a slight left sided ataxia, hemiparesis, and hypophonia. MR imaging showed reduced cognitive performance and flexibility, a deficit in learning and memory, and abnormal visual constructional abilities which were compatible with a subcortical dementia. Brain MRI showed extensive hypodense changes in the left temporal and occipital lobes, as well as mild hyperintense changes in the left basal ganglia. The patient described episodes of aphasia, headache, and hemianopia. Six members of this family, three affected and three healthy, have been genotyped with eight chromosome 19 markers spanning the CADASIL locus. No differences were found with any of those markers. Maximum lod scores were obtained with markers D19S226, D19S253, and D19S199, strongly suggesting that this family is linked to the CADASIL locus.