Thyroid Hashimoto’s encephalopathy.

Hashimoto’s encephalopathy. This was originally postulated to be a distinct disease entity by Brain et al in 1966 and there have subsequently been case reports substantiating the hypothesis that it represents a unique condition. The characteristic features are a subacute onset of confusion with altered mental status, and events that respond to steroids and which occur in the context of high anti-microsomal antibody titres. To date all the patients reported have been either euthyroid or hypothyroid at the time of presentation. We present a patient with Hashimoto’s encephalopathy with pronounced thyrotoxicosis, 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 conjunction with a left hemianesthesia. On examination at admission she was flushed, severely hypothyroid with a hyperdynamic hypovolemic, and dural circulatory. Thyroid gland was slightly enlarged but there was no associated bruit. Cranial nerve examination disclosed left visual inattention as the only abnormality. Limb examination showed a moderately severe left hemiparesis with left sensory inattention, generalised hyperreflexia, and bilaterally extensor plantar response. She had wasting of the shoulder girdle muscles and adhesive capsulitis of the shoulder joints bilaterally. In the days immediately after admission she continued to be confused and had florid visual hallucinations, while independently having runs of paroxysmal atrial fibrillation. As a result of the original negative findings (see later) desmethylphenothine (12 mg/day) and acyclovir were started with the presumptive diagnosis of an encephalitis or vasculitis. On this regime she made a dramatic improvement, which was further enhanced by the treatment of her thyrotoxicosis on receipt of her thyroid function tests. The introduction of carbimazole and propranolol was then followed by a reduction and acyclovir. Attempted steroid weaning over subsequent days provoked a recrudescence of her focal symptoms on two occasions, with weakness of her right arm. Eventually the patient was stabilised on prednisolone (40 mg/day) and discharged on a slowly reducing course with no relapses three months after discharge.

Investigations performed during her inpatient stay showed that full blood count, erythrocyte sedimentation rate, urea, electrolytes, blood cultures, blood myoglobin function tests, and serum immunoglobulins were normal. Protein electrophoresis showed an acute phase response with a C reactive protein of 52 mg/l. Her autoantibody screen and VDRL/TPHA serology were negative, but her thyroid function tests showed her to be thyrotoxic with TSH less than 0.03 U/l, free T4 >80 pmol/l, and free T3 41 pmol/l. Her thyroid microsomal antibodies were positive at a titre of 1:6400. Her CSF analysis was normal with negative oligoclonal bands and repeated CSF cytology showed no abnormality. Her chest radiograph was normal but her ECG showed a sinus tachycardia with episodes of paroxysmal atrial fibrillation. Her EEG showed occasional brief bursts of frontal slow activity which spread posteriorly and brain CT and MRI with contrast and MRI with gadolinium were normal. In addition a transchoranic and transsephalochal ECG along with MRI of her heart were all normal.

These results show that she had a pronounced thyrotoxicosis with antinuclear antibodies. There was no evidence for any fixed structural lesion within the CNS accounting for her clinical presentation as evidenced by her normal brain CT and MRI.

Autoimmune thyroid disease can be considered as a range of clinical disorders reflecting the variety of autoantibodies present. Hashimoto’s disease is characterised by the presence of thyroid antimicrosomal antibodies and has rarely been associated with an encephalopathic process of unknown aetiology. All previously described patients have either been euthyroid or hypothyroid and this is the first description of an encephalopathy in combination with thyrotoxicosis. As the mechanism of encephalopathy is uncertain the term thyroid related encephalopathy is preferable. Although atrial fibrillation was present in our patient, the normal heart and head imaging argued against an embolic cause for her condition. Furthermore, her remarkable steroid responsiveness suggests an autoimmune cause for her fluctuating multifocal encephalopathy.

Various mechanisms have been postulated to account for this unusual condition. One possibility is demyelination, which can virtually 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 cerebral autoregulation.

This patient completes the repertoire of thyroid states seen in thyroid related 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 aetiology.

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Cerebral salt wasting syndrome

Excessive natriuresis, resulting in hyponaemia and polyuria, is an often recognised complication after subarachnoid haemorrhage. Initially this was attributed to inappropriate antidiuretic hormone (ADH) secretion resulting in water retention, but...
The cerebral salt wasting syndrome is accompanied by hypovolaemia. We report a patient with cerebral salt wasting after aneurysmal subarachnoid haemorrhage who showed remarkable changes in a few unrelated large families. A 46 year old woman was admitted with blood in the suprasellar cisterns and the left Sylvian fissure. Two days later she developed in the intracranial pressure, during surgery, salt and fluid loss were fully compensated by 0.9% NaCl infusion. On day 9 she was found unconscious with respiratory failure and bradycardia and CT disclosed a recurrent subarachnoid haemorrhage with severe headache and vomiting. Physical examination was unremarkable. Brain CT showed a subarachnoid haemorrhage with blood in the suprasellar cisterns and the left Sylvian fissure. She 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 atrial natriuretic protein in CSF was 640 pmol/l and in serum 678 pmol/l, and the colloidal osmotic pressure was between 18.7 and 24.0 mm Hg. Serum ADH concentrations were normal. Treatment with fluoroicortisone had no effect on renal sodium loss. Despite the high 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 measurements of plasma sodium were also normal. Hyponatraemia, resulting in a decrease in natriuresis. Because an increase of atrial natriuretic protein, resulting in a decrease in natriuresis.

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

"Cerebral autosomal dominant arteriopathy with subcortical infarcts and leuкоencephalopathy" (CADASIL) is a newly defined syndrome characterised, in the absence of hypertension, by recurrent subcortical ischaemic strokes and by peculiar 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, periodic acid Schiff (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.

Electron microscopy showed changes in a few epineurial vessels, consisting of electron dense, acellular granular deposits along the outer aspects of the vessel walls (fig 2A). Most of these granules were on the abluminal surface of pericytes and less often on endothelial cells. Most dense deposits were focally and predominantly in the small myelin fibres. Denser deposits were frequently located in thickened basal laminae and were often pushing back the cell membrane of an adjacent pericyte (fig 2B and C). Most dense deposits were round or oval but some were flat or disc shaped and oriented parallel to the cell surface. The number of dense deposits ranged from none to five or six around a single vessel. Some were found in very small arterioles but most were in large capillaries or meta-arterioles (size 14–15 μm) consisting of endothelial cells surrounding by a basement membrane without the presence of smooth muscle cells. In some vessels, the basal lamina surrounding the endothelial cells was clearly redundant and tortuous (not shown). Many pinoctytic vesicles were found along and underneath the surface of cell membranes. Their density was not altered at the site of close apposition to the cell membrane with the electron dense granular deposits.

The presence of granular electron dense...