

Ophthalmology review demonstrated no non-refractive visual problems.

MRI 5 months after operation showed that the haemorrhagic pineal lesion demonstrated previously had almost completely resolved. Only a 9 mm × 7 mm focus of predominantly low signal intensity on T1 and T2 weighted images remained in the pineal. There was no pathological enhancement, nor any oedema. No hydrocephalus was identified. There was no evidence of a pineal or posterior third ventricular mass. There was evidence of altered blood product in the pineal gland with a small amount of haemosiderin staining of the adjacent right thalamus (Fig. 2). Given the 5-months interval since the acute presentation, and the normal appearance of the pineal, a haemorrhagic tumour was effectively excluded. MR angiography demonstrated no evidence of an arteriovenous malformation.

Clinical review at 8 months showed the patient remained asymptomatic with a normal neurological examination. Further MRI at 18 months showed no change from the 5 months scan and she remained well.

DISCUSSION

Apoplexy refers to ill-defined symptoms subsequent to acute haemorrhage into a normal structure or lesion and is an uncommon intracranial event. Clinical significant pineal apoplexy is a rare event, with only 12 cases found encompassing a heterogeneous range of pathology.^{1–11} Causes of pineal apoplexy include haemorrhage into a pineal cyst, or tumour, or haemorrhage from an adjacent vascular malformation.¹ There have been no reports of haemorrhage into a normal gland but this seems plausible in the context of anticoagulation therapy. In cases such as ours with no enhancement, the most likely underlying lesion may be pineal cyst. Non-neoplastic cysts of the pineal gland are common incidental findings in as many as 40% of routine autopsies, and asymptomatic pineal cysts are present on 1.5–4.3% of MRIs.¹² Two reviews have revealed only 75 reports of symptomatic pineal cysts.^{1,2} Pineal apoplexy is the most uncommon presentation of a symptomatic pineal cyst. Modes of presentation include the syndromes of: paroxysmal headache with gaze paresis; chronic headache, gaze paresis, papilloedema, and hydrocephalus; and pineal apoplexy with acute hydrocephalus.¹²

The first reported case of pineal apoplexy was in 1976.³ The current report represents the second reported case of pineal apoplexy in a patient undergoing anticoagulation therapy. Several authors have advocated the need for surgical removal of apoplectic pineal lesions with acute hydrocephalus as either a primary treatment or following operative CSF diversion.^{1,2}

Kobayashi et al.¹ advocate surgical extirpation for the treatment of pineal apoplexy, on the basis of a single case of cavernous angioma. Repeated bleeding is thought to be a risk for patients with pineal apoplexy but there is no firm data to indicate the risk of this occurrence. Mukherjee et al. have said that placement of a CSF shunt should not be the sole management of hydrocephalus from a pineal mass lesion as this approach may result in progressive and permanent gaze difficulty.² The risk of such deficits has not been quantified and all patients reported to date have had pineal resections. The other advantage of resection is the provision of a histological diagnosis.

This is the first report of successful management of pineal apoplexy by CSF diversion without resection of the lesion. Subsequent MRIs to 18 months have shown no significant residual pathological lesion in the pineal region, although because digital subtraction angiography has not been performed, a small dural AVF has not been completely excluded. Given this patients age,

history of pulmonary thromboembolic disease requiring anticoagulation, and uncertainty about the natural history of small dural AVFs, it was decided that angiography was not clinically indicated.

CONCLUSION

Consideration should be given to serial imaging following resolution of pineal hemorrhage. Pineal surgery could be reserved for definitive abnormalities on follow-up imaging. This approach may reduce operative morbidity for patients with pineal apoplexy.

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Transient global amnesia in a patient with acute unilateral caudate nucleus ischemia

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Summary The pathogenesis and localization of neuronal dysfunction in transient global amnesia (TGA) is still disputed more than 40 years after the first description of this clinical entity. Previous studies have indicated that structural abnormality is rare in TGA. We report a case of TGA in a patient with acute ischemia in the body of right caudate nucleus. This provides evidence in support of an ischemic hypothesis as the possible etiology of TGA. The role of caudate nucleus in human memory is also reviewed.

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INTRODUCTION

Transient global amnesia (TGA) is a well-recognized clinical entity now and its incidence has been estimated as about 3–30 per 100,000/year affecting people above 50 yrs of age.¹ However, the pathogenesis and localization of neuronal dysfunction in TGA is still disputed. Several hypotheses including ischemia, epilepsy, migraine and psychological disturbances have been proposed.¹ Previous studies have indicated that structural abnormality is rare in TGA and it would result from transient disturbances in brain regions traditionally deputed to memory such as temporal lobes and the thalami.^{2,3} We report a case of TGA in a patient with an acute ischemia in the body of right caudate nucleus.

CASE REPORT

A 56-year-old right handed bus driver was admitted to the acute medical assessment unit of our hospital. His wife reported that while getting ready for his morning shift he suddenly appeared perplexed and started asking her and his brother-in-law repeatedly where he was. He was neither able to remember the events of the same morning or being visited by his daughter and her family the day before. His wife mentioned that he complained of feeling unwell the preceding evening and skipped his dinner to go to bed early but the patient had no recollection of this. No other neurological symptoms were reported. There was no history of previous similar episodes and there was no history of hypertension, diabetes, transient ischemic attacks, stroke, epilepsy, migraine, psychiatric illness, head trauma and alcohol abuse. He was not on any medication and he did not smoke. He was alert, cooperative but appeared restless during the course of examination. His speech was normal without any paraphasic errors and word finding difficulties. His comprehension was intact, although he was unable to recollect having had conversations a few moments later. He was completely disoriented in time but was well oriented in place. He was unable to give an account of how he came to be in the hospital. He had no recollection of the earlier events of the same day as well as of the preceding day. Immediate memory as assessed by repetition of the sentence and forward digit span was normal. He was able to spell five-letter words backwards. Recollection of the visual and verbal material after several minutes was very poor and he demonstrated no familiarity with the material presented. Memories of significant personal events, remote events and own identity were intact. He acknowledged his memory difficulties and expressed concern, but there was no evidence of confabulation. He was able to name objects correctly describing their uses and his reading, writing, calculating and visuoconstructive (copying of geometric figures) abilities were normal. The remainder of the neurological examination and general physical examination was normal. The memory deficit gradually improved over the next 18 h. A repeat assessment 24 h after the onset of symptoms demonstrated no memory disturbances other than total amnesia concerning the day of the attack.

NEURORADIOLOGICAL INVESTIGATION

MRI was performed on a Gyroscan Intera 1.5T (Philips Medical Systems) with echo-planar (EP) capability approximately 7 h after the onset of symptoms. A single shot EP diffusion-weighted (DW) sequence (EP spin echo TR 4454 ms/TE 86 ms, field of view 23 cm, a 77×256 matrix with NSA of 1, slice thickness 5 mm with 1 mm gap, 5 b values = 0 to 1000 s/mm^2 and scan time 26.7 s) demonstrated hyper intense signal changes in the body of right caudate nucleus (Fig. 1) consistent with acute ischemia. On apparent diffusion coefficient map (not shown), a corresponding reduction was noted. Fluid-attenuated inversion recovery (FLAIR) sequence (TR 6000 ms/TE 1000 ms/TI 1900 ms, field of view 25

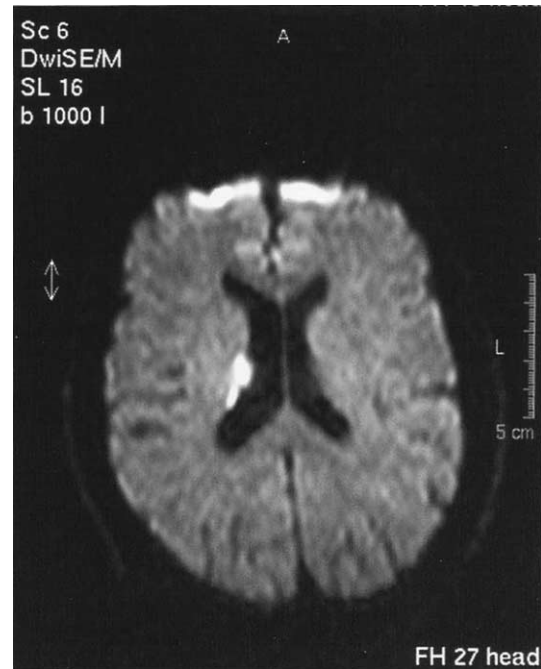


Fig. 1 DW image b 1000.

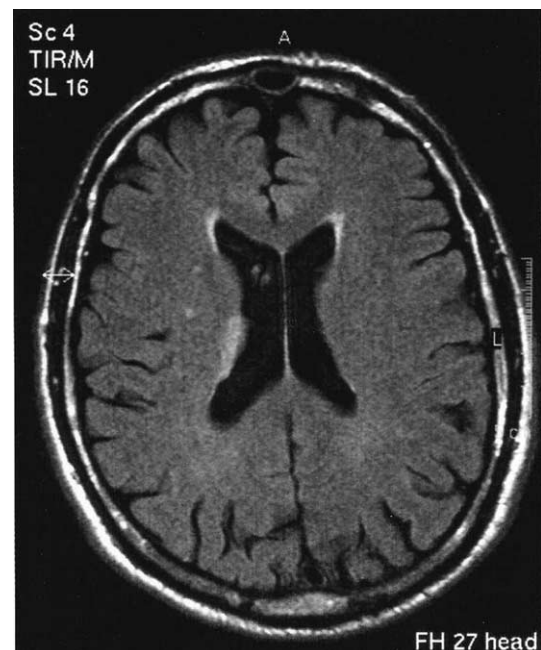


Fig. 2 FLAIR image.

cm, a 280×512 matrix with NSA of 2, slice thickness 5 mm with 1 mm gap, scan time 4.12 min) also demonstrated T2 hyperintense signal in the same area (Fig. 2) as well as few tiny scattered areas of T2 hyperintensity in the frontal white matter.

DISCUSSION

The present case fulfills the clinical criteria for TGA proposed by Hodges and Warlow.⁴ It also meets the gold standard of combined neuropsychological assessment and neuroradiological investigation involving modern techniques during the acute stages of TGA fulfilling all the criteria of Hodges and Warlow. Recent studies have demonstrated the value of DW MRI in achieving the primary objective in the evaluation of an acute cerebral ischemic event: early identification of stroke, its anatomy and its etiology.⁵ Early ischemic brain injury detected by DW MRI has a high correlation with postmortem infarction, acute and chronic clinical sensitivity and clinical outcome.⁶ DW MRI is also more accurate than FLAIR for the detection of stroke for all subtypes in all anatomic distributions and it can depict ischemic tissue injury within minutes after onset of symptoms, therefore it is invaluable in evaluating transient amnesic states.⁷ In our patient, the acute tissue ischemia was diagnosed based on the DW hyperintensity on *b* 1000 image and corresponding reduction in the apparent diffusion coefficient map. It also had corresponding changes seen on the FLAIR sequence. The tiny scattered areas of hyperintense signal in the frontal white matter on FLAIR imaging appear to be long standing as no corresponding abnormality was noted on the DW imaging. Our positive finding in this case contrasts with the findings of study by Gass et al. who have reported lack of evidence of acute ischemic tissue changes in TGA.⁸ Controlled studies have demonstrated that patients with well-defined diagnosis of TGA have the same prevalence of vascular risk factors as the general population and a better prognosis than TIA patients.^{9,10} In fact, structural abnormality has been rarely reported in TGA.¹¹ Transient abnormalities of tissue have been detected using single-photon emission CT and positron emission tomography^{12,13} and as several of these patients have a precipitating event it has been suggested that spreading depression of Leão might be responsible for TGA. This phenomenon consists of gradual spread of a wave of depolarization, followed by neuronal depression across the cortex occurring in response to a wide variety of insults.¹⁴ Alternative hypotheses including epilepsy, migraine and recently suggested functional mechanism also do not definitely explain the mechanism of TGA.¹ At best the pathogenesis of TGA remains controversial but the present case provides evidence in favor of an ischemic hypothesis and highlights the fact that in some patients TGA may not follow a benign course. The diagnosis of stroke in a patient with TGA has important clinical bearing, as it should lead to a thorough search for the cardiovascular risk factors and institution of secondary stroke prevention strategies.

The incidence of caudate stroke has been found to be about 1% in one recent series.¹⁵ Small vessel disease and a cardiac source of embolism are the main causes of caudate ischemic stroke, large vessel disease has been found in only one-sixth of the patients. The commonest involved arterial territories in caudate ischemic stroke are territories of the lateral lenticulostriate arteries from the middle cerebral artery and the anterior lenticulostriate arteries from the anterior cerebral artery. Prominent clinical features of caudate stroke are behavioral and cognitive including abulia, contralateral motor and visuospatial neglect, confusion, dysarthria and aphasia.¹⁶ One third of the patients with left caudate lesions have verbal amnesia while patients with right caudate lesions show visual amnesia, suggesting a role of the caudate nucleus in the integration of visual

and verbal memories. Verbal comprehension and verbal memory deficits are caused by dysfunction of corticocaudate connections.¹⁷ Bilateral caudate lesions may yield global amnesia, while unilateral lesions may cause impairment of the frontal lobe functions and decreased free recall of episodic and semantic items.¹⁸ Previous studies have suggested that there could be multiple memory systems in the brain.^{19,20} It has been suggested that certain aspects of memory function may have been duplicated in the different structures during evolution and basal ganglion structures make their own contribution to overall memory function.²¹ In our patient there was selective disorder of episodic memory (both anterograde [verbal and non-verbal] and retrograde) without any impairment of general cognitive function (other than disorientation in time) or of other components of memory such as short-term memory and semantic memory. In the study conducted by Kumral et al. on caudate vascular lesions, one patient had anterograde (both verbal and visual) amnesia with abulia caused by unilateral caudate infarct¹⁵ and another report has described verbal amnesia lasting for 6 days with auditory hallucination and dysgraphia caused by infarct in the left caudate nucleus.²² The anatomical basis of these cognitive and behavioral changes lies in the fact that caudate nucleus is the principal crossing area of basal ganglia-thalamocortical loops. The caudate nucleus connects associative cortex, including frontal, parietal, and temporal lobes with deeper anatomic structures by cortico-pallido-nigra-thalamocortical loops. These loops are discrete and multiple but partially overlapping and integrated through their passage in pallidum and substantia nigra to the circumscribed nuclei of the thalamus, and from there they are projected back to their original lobar areas.²³ In our patient there was a definite temporal association of TGA episode with the acute ischemia in the right caudate nucleus, implying a possible casual relationship between the two.

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Syringomyelia-like manifestation of subacute combined degeneration

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Summary A 32-year-old male presented with progressive weakness and numbness of both upper limbs of one-month duration. The patient had weakness and wasting of small muscles of both hands with weak grip. Sensory system revealed graded sensory loss to pain, temperature and touch in C5 to T1 distribution and vibration and joint position sense from C5 to C8 in the both upper limbs. There was areflexia in the upper limbs while there was no motor or sensory deficit in the lower limbs. The cortical potential on stimulation of posterior tibial nerve was prolonged on both sides. On MR imaging of the cervical spine there was iso to low intense lesion which was hyperintense on T2-weighted imaging along the dorsal aspect of the cord extending from C2 to C6 level. The axial images showed involvement of the posterior column. The serum vitamin B₁₂ level was found to be low. The patient responded to parenteral cyanocobalamine therapy and the radiological lesion subsequently resolved.

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INTRODUCTION

The neurological manifestations of vitamin B₁₂ deficiency are the result of its effects on the peripheral nerves, spinal cord, optic nerves and brain. Subacute combined degeneration, a myelopathy due to vitamin B₁₂ deficiency, manifesting with sensory disturbances, weakness and spasticity, and primarily involving the dorsal and lateral spinal columns was first described by Russel et al.¹ Subacute combined degeneration is curable, but early detection is necessary for full clinical recovery.²

Although not specific, the MR findings have distinguishing features for subacute combined degeneration.^{3–5} MR imaging may be a useful addition to the clinical assessment in monitoring the efficacy of treatment.⁶ Myelopathy is the common manifestation of subacute combined degeneration, however, syringomyelia-like manifestation is not reported in the literature till date, to the best of our knowledge.

We present a case of subacute combined degeneration who had syringomyelia-like presentation and showed resolution of both clinical as well as radiological abnormalities with B₁₂ therapy.

CASE

A 32-year-old male, non-diabetic and non-hypertensive with previous good health presented to us with progressive weakness and numbness of both upper limbs of one-month duration. The numbness started from finger tips and progressed to involve the entire limb. He denied of ever having experienced sensations of pin pricks or needles. About two weeks of the onset of the initial symptom, he developed weakness of both hands which was slightly asymmetrical, right side more than left. The weakness impaired his activities of daily living like difficulty in buttoning and unbuttoning the clothes, holding the soap cake while bathing and breaking the chapattis. He had no sensory or motor symptom in the lower limbs. On rare occasions, he experienced a current like sensation going down his spine on neck flexion. He denied of any unsteadiness on standing with eyes closed or of difficulty while walking in the dark. He had no bladder or bowel dysfunction. His general physical examination was non-contributory. On detailed neurological examination he had weakness and wasting of small muscles of both hands with weak grip. The power in the wrist flexors was 3/5 (on MRC scale); in wrist extensors 4/5 while at elbow and shoulder it was 5/5. There was no evidence of any weakness in lower limbs with power being 5/5 at all the joints. The biceps, triceps and supinator jerks were absent in both the upper limbs. While the deep tendon jerks of the lower limbs were well elicitable. Plantars were down going on both sides. Sensory system revealed graded sensory loss to pain, temperature and touch in C5 to T1 distribution and vibration and joint position sense from C5 to C8 in both upper limbs. The loss was up to 50% in the hands and the distal aspect of forearm while in the rest of the upper limb it was up to 40%. The sensations in the trunk and lower limbs were normal. He had no cognitive or cranial nerve deficit. Bilateral fundii were normal. There was no cerebellar sign. His hemoglobin was 11 g/dl (13.5–15.5 g/dl), hematocrit was 43% (41–53%), mean corpuscular volume 92 fl (83–96 fl) and the peripheral smear was normal. The biochemical profile for blood sugar, blood urea, serum creatine, lipid, liver function tests and serum electrolytes was normal. CSF study for protein, sugar and cellular response was normal. Oligoclonal bands were not seen in the CSF. Serum vitamin B₁₂ level was 68 pg/ml (normal range 180–900 pg/ml). ELISA for HIV 1 and 2 was non-reactive. Gastric biopsy did not reveal the presence of any abnormal pattern, in particular gastric atrophy. Motor and sensory nerve conduction studies of the conventionally studied nerves of both the