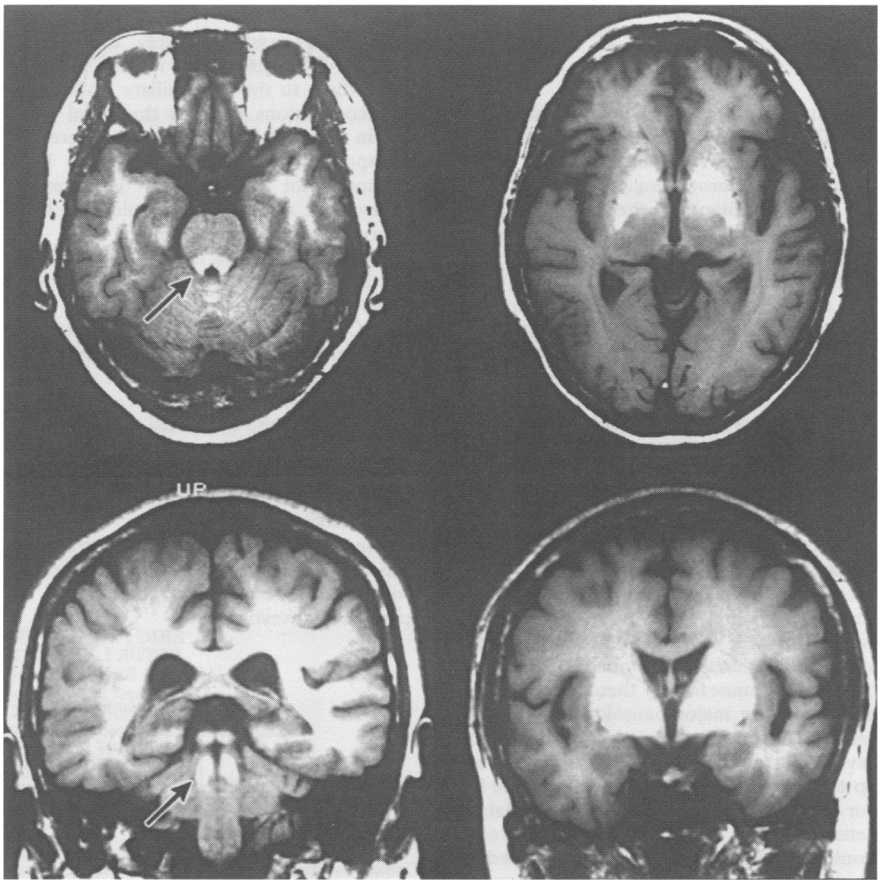
transfusion at the age of 32. Sclerosing treatment had been performed twice for oesophageal varices. She had had no episodes suggestive of cerebral haemorrhage or hepatic encephalopathy.

Neurological examination was normal. She had no mental impairment or extrapyramidal signs. T1 weighted MRI of the brain showed symmetric high signal intensity in the globus pallidus, subthalamus, cerebral peduncle, substantia nigra, and the periaqueductal region of the mesencephalic and upper pontine tegmentum (figure). T2 weighted MRI and brain CT were normal. Abnormal data in routine laboratory tests included erythrocyte sedimentation rate 44 mm/hour, red blood cell count 3 240 000/mm^3, white blood cell count 2 200/mm^3, and platelets 64 000/mm^3, packed cell volume 27%, haemoglobin 8·8 g/dl, total bilirubin 1·7 mg/dl, and raised values in other liver function tests. A test for antihepatitis C virus antibody was positive. Metals in serum were (normal range in parenthesis) iron 45 (56–117) μg/dl, unsaturated iron binding capacity 416 (150–336) μg/dl, copper 148 (103–159) μg/dl, zinc 48 (61–121) μg/dl, and manganese 0·55 (0·1–0·27) μg/dl. Whole blood contained 8·33 (0·4–2·0) μg manganese/dl. The patient's orbital pain disappeared spontaneously. She was given ferrous sulphate (320 mg per day), and was recommended to avoid manganese rich foods.

Hyperintensity in T1 weighted MRI in the basal ganglia and in the midbrain tegmentum was the characteristic MRI finding in our patient. An increased lipid, melanin, and methaemoglobin concentration, or calcified tissues, and ectopic Schwann cells in neurofibromatosis may cause similar MRI changes.[4 5] In our patient, the possibilities of raised methaemoglobin, calcified lesions, or ectopic Schwann cells were ruled out by normal CTs, no stigmata of neurofibromatosis, and no history of stroke. Symmetric hyperintensity of basal ganglia in T1 weighted MRI has been reported in patients with chronic liver diseases and possible manganese intoxication.[2 3 6 7] Pujol *et al*[2] reported that on T1 weighted MRI, 33 of 45 patients with advanced liver dysfunction had symmetric high signal intensity in the globus pallidus. In seven of 11 patients who had liver transplants, MRI abnormalities decreased, and disappeared in four patients, suggesting a direct linkage between MRI findings and liver dysfunction. Manganese shortens spin lattice relaxation times via its paramagnetism and increases signal intensity on T1 weighted MRI.[3] The high manganese concentration in whole blood in our patient suggested that the changes on MRI may reflect manganese accumulation, as postulated in chronic hepatopathy.[7] T1 hyperintense lesions in the basal ganglia have not been reported in patients with Hallervorden-Spatz or Wilson's diseases, which result in iron or copper deposition in the brain, especially in the basal ganglia.[8–10] Therefore, we postulate that the hyperintensity in T1 weighted images may represent an alteration resulting from an accumulation of paramagnetic substances such as manganese, but not such non-specific changes as cell loss, necrosis, or glial proliferation. In iron deficiency anaemia the enteric absorption of iron and manganese is increased.[11] Thus in our patient iron deficiency may have enhanced manganese absorption, whereas liver dysfunction may have resulted in the reduction of manganese excretion.



Axial and coronal T1 weighted MRI (TR 500 ms, TE 15 ms) showing high signal intensity in the basal ganglia and periaqueductal region of the upper brain stem (arrow).

Several experimental and clinicopathological studies on manganese intoxication emphasised vulnerability of the basal ganglia, especially the globus pallidus.[3 11] The present patient showed T1 high signal intensity in the tegmentum of the upper brain stem as well as changes in the basal ganglia. The pattern of distribution was almost identical to that seen in our previous patient with manganese intoxication during total parenteral nutrition.[6] These two findings suggest that the basal ganglia and the periaqueductal structures in the tegmentum of the upper brainstem may have common metabolic characteristics with respect to certain trace metals.

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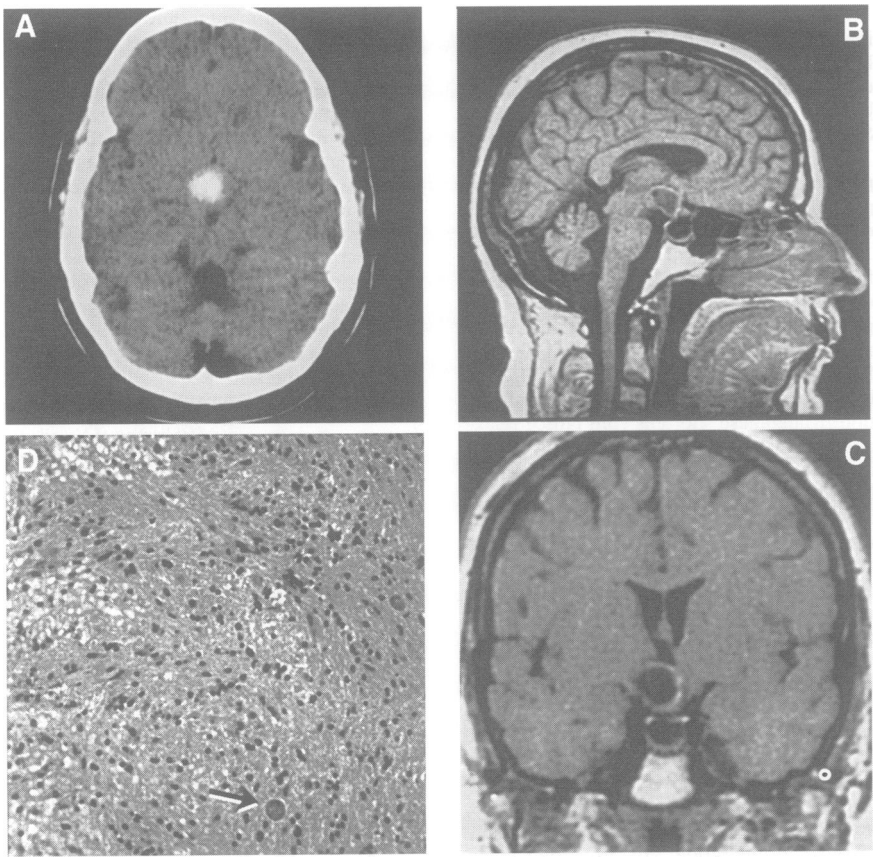
Transient amnestic syndrome after spontaneous haemorrhage into a hypothalamic pilocytic astrocytoma

The aetiology of the syndrome of transient global amnesia is unknown.[1 2] A relation with migraine has been proposed and in a case-controlled series, no association was found with cerebrovascular disease, but 7% of patients developed epilepsy within one year of their first attack.[1] Controversy exists as to the relation between tumours or structural brain lesions and transient global amnesia, and in the reported cases the



(A) CT of the head without contrast obtained 48 hours after clinical recognition of the patient's acute problem. This shows an area of increased attenuation within the hypothalamus. (B) sagittal and (C) coronal T1 weighted MRI without contrast. These images show an expansile mass centred in the right hypothalamus which extends across the midline and obliterates the floor of the third ventricle. The mass has a moderately hypointense central portion, and a hyperintense rim. Incidental note is made of a slightly enlarged "partially empty sella" with the pituitary gland having a concave margin. (D) haematoxylin and eosin stain (× 60) of the biopsy specimen showing a biphasic cellular pattern, granule bodies (arrow), and pilloid appearance consistent with a pilocystic astrocytoma.

importance of epilepsy in the pathogenesis has been understated.[3 4] We report a case of spontaneous haemorrhage into a hypothalamic pilocytic astrocytoma, presenting with an episode with clinical features suggestive of transient global amnesia.

A 58 year old previously healthy white woman was found by her husband early one morning acting strangely. She recognised her husband, and was not impaired physically or in speech. He described her as looking "puzzled," but she was not delayed in her responses, and had normal comprehension to simple commands. She seemed unable to recall the answers given to her less than a minute previously, and repetitively asked questions such as "what day is it," "where are the kids," and "where are we going?" There was no abnormal posturing or laughing. By late afternoon she was slowly improving and by the next morning her memory deficit had resolved, but she was completely amnestic for the preceding day. She presented for review to her family physician the next day, and her examination was reported to be normal. A cranial CT showed a 2·5 cm area of increased attenuation in the hypothalamus, without contrast enhancement (figure (A)). Cranial MRI showed an expansile mass within the medial aspect of the right hypothalamus, displacing the third ventricle from right to left, and the optic chiasm inferiorly (figure (B and C)). This mass had mildly decreased signal intensity on T1 weighted images and profoundly decreased signal intensity on T2 weighted images consistent with acute haemorrhage. Carotid angiography was normal. An open biopsy of the lesion was performed via a subfrontal approach at another institution. After the operation she had short term memory impairment, and she was referred to our institution for further evaluation and treatment recommendations. Review of the histopathology showed a pilocytic astrocytoma (figure (D)). The postoperative memory deficit has shown only slight improvement at 12 months follow up. Subsequent cranial MRI and CT have been consistent with resolving haemorrhage. There have been no further transient neurological episodes suggestive of complex partial or other seizures.

Caplan has defined the following strict criteria for the diagnosis of transient global amnesia[3]; (*a*) the attack onset should have been witnessed; (*b*) dysfunction during the attack should have been limited to repetitive queries and amnesia; (*c*) there should have been no other major neurological signs or symptoms; and (*d*) the memory loss should have been transient, usually lasting hours or up to one day. The onset of the episode in our patient cannot, however, be reliably determined, as the patient either awoke from sleep with the amnesia or developed it soon after awakening. This is a not uncommon finding in patients with presumed transient global amnesia. In the series by Miller *et al*[2] 20 of the 347 episodes of transient global amnesia were first noted on awakening.

In our patient, the first neurological manifestation was a transient global amnesia-like episode, with complete resolution within the time course normally seen with transient global amnesia. Previous reports of transient global amnesia in association with tumours have been comprehensively reviewed by Caplan,[3 4] with reports of primary or secondary tumours involving the temporal lobe, thalamic region, and in one report a very large tumour diffusely involving the anterior nucleus of the thalamus and fornix. The mechanism of transient amnestic episodes in these patients is uncertain. An epileptic mechanism seems likely in many cases, with involvement of temporal lobe structures, and some of these reported cases had clear epileptic features with the amnestic event, later developed epilepsy, had focal neurological signs at presentation, or developed focal neurological signs before the memory loss resolved. "Transient tumour attacks" may occur in patients with intracranial tumours, and investigations for epilepsy or a vascular cause are often negative.[5] One of the cases described by Ross had features suggestive of transient global amnesia, but had a right superior homonymous hemianopia due to a left temporal lobe neoplasm.[5] The aetiology of the cerebral disturbance causing the amnesia remains uncertain. In our case, the temporal lobes were not involved, and there were no other features at presentation or follow up to suggest a diagnosis of epilepsy. There was clearly an acute event with haemorrhage into the tumour, and we believe that the resultant sudden mass effect and oedema were responsible for the sudden transient global amnesia through pressure on pathways and structures involved in memory. The proximity of the pilocytic astrocytoma to the mammillary bodies and their connections suggests that local pressure in this region from oedema related to the spontaneous haemorrhage may have accounted for the memory impairment. Involvement of the fornix could also have interrupted cholinergic inputs from the medial septum to the hippocampus, which may be potentially important for learning processes.

By Caplan's more stringent criteria,[3] our case would not be considered to fulfil all the clinical diagnostic criteria for the diagnosis of transient global amnesia. It does, however, resemble the syndrome sufficiently to cause diagnostic confusion. Transient amnesia can have many causes, including epilepsy,[1] cerebrovascular disease,[3] and those associated with tumour,[3 4] whatever the mechanism. Before attributing transient amnesia to transient global amnesia, other more serious causes must be excluded by appropriate investigation.

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Musical hallucinations associated with post-thyroidectomy hypoparathyroidism and symmetric basal ganglia calcifications

Bilateral symmetric basal ganglia calcifications most often result from parathyroid disorders and subsequent imbalances in calcium and phosphorus metabolism. Associated neurological and psychopathological symptoms are, however, controversial.[1]

"Musical hallucinations" are usually defined as the hearing of tunes and melodies without relevant external stimuli. They are reported to be very rare compared with verbal hallucinations.[2]

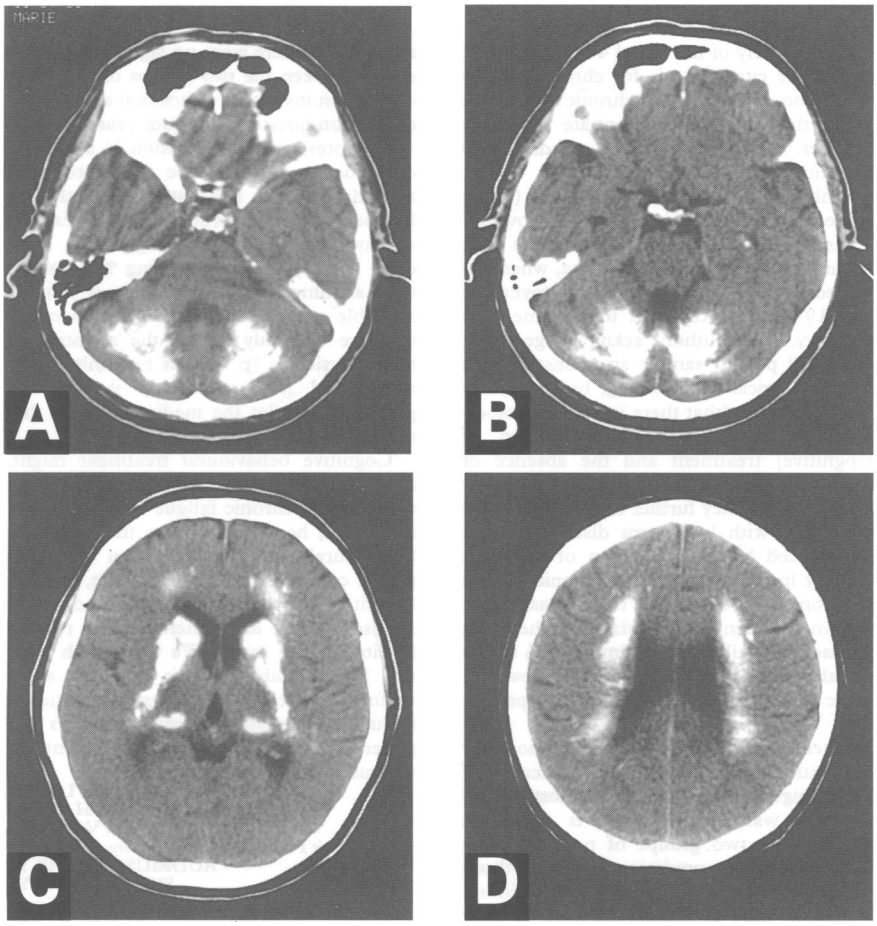
A 63 year old, right handed housewife with a 40 year history of post-thyroidectomy hypoparathyroidism presented with a chronic progressive syndrome, with musical hallucinations, mild intellectual impairment, and cerebellar ataxia.

She reported having suddenly started hearing music about five months before admission. The tunes, which were characterised by a stomping rhythm ("that terrible modern pop music"), did not vary much ("like a short tune being played all over again"). The hallucinations occurred continuously during the daytime, when awaking at night, or when talking. Insight into the character of hallucinations was achieved after initial deception.

Ear examination showed a mild pancochlear perceptive hearing loss due to presbyacusis, more prominent on the left side. Neurolinguistic and neuropsychological examinations failed to show signs of aphasia, agnosia, visuospatial disorders, or hemispatial neglect. The patient had an IQ of 86 on the Hamburg Wechsler intelligence test for adults with low scores on verbal scales. Verbal and visual short term memory were impaired slightly, as measured by the Benton visual retention test and Amthauer (IST 70) test indicating acquired cognitive deficits of the organic type.

Electrophysiological examinations (EEG, topographical EEG, and acoustically and visually evoked potentials (AEP; VEP) showed no evidence of epileptic activity or consistent focal activity. P300, AEP, and VEP latencies were in the normal ranges.

Cranial CT showed dense areas of calcification predominantly involving the basal ganglia (for example, pulvinar thalami, striatum), cerebellar dentate nuclei, and periventricular white matter (figure).



Cranial CT (unenhanced) with bilateral symmetric and confluent calcifications of dentate nucleus of the cerebellum, (A, B) pulvinar of thalamus and striatum (caudate nucleus and putamen), (C) and hemispheric white matter (C, D).

At admission ECG showed massive prolongation of the Q-T interval (0·48 ms; frequency adapted upper limit: 0·36 ms); ECG was normal on the day of discharge. Complete blood count and all routine laboratory tests were in the normal ranges, except serum concentrations of calcium on admission and discharge of 1·45 and 2·48 (normal range 2·0–2·75) mM and inorganic phosphorus concentrations of 6·03 and 1·45 (normal range: 0·8–1·5) mM. Twenty four hour urinary calcium excretion rates on the day of admission and discharge were 0·77 and 6·17 (normal range: 3·25–8·25) mM and inorganic phosphorus excretion rates were 7·3 and 21·7 (normal range: 10–32) mM. Intact parathormone was below the detection limit (<1 pg/ml; normal range: 10–60 pg/ml).

The patient initially received 1500 mg Ca^{2+} and 4 mg dihydrotachysterol orally per day. Three additional intravenous applications of Ca^{2+} were given to avoid manifest tetany. The hallucinations started to fade on day 12 of treatment and dihydrotachysterol was adjusted to 2 mg per day. Three weeks after admission the patient was free of hallucinations. Cerebellar symptoms, and signs of neuromuscular instability (Chvostek's and Trousseau's signs) improved significantly. Tinnitus and mild deafness remained unchanged three months later, the patient had no complaints, and her neurological state was normal on the medication described.

The disappearance of the musical hallucinations coincided with the normalisation of serum electrolyte balance. Although electrolyte disturbances after hypoparathyroidism have long been known to cause neuropsychiatric symptoms,[3] musical hallucinations have not been reported.

Two basic theories regarding the pathogenesis of complex auditory hallucinations have been proposed.

The "perceptual release" theory is based on the pathogenetic role of the sensory impairment. Our patient had had a mild progressive hearing loss associated with tinnitus. Despite the disappearance of auditory hallucinations, however, there was no improvement in hearing and tinnitus. This is not compatible with either the persistent nature of otogenic auditory hallucinations due to progressive hearing loss, or with transient hallucinations of acute otopathic origin, as no acute lesion could be found. Complex auditory phenomena may also occur with lesions of the tegmentum of the pons and lower midbrain; however, CT and AEP failed to disclose a brainstem lesion in our patient.

According to the other pathogenetic concept, local metabolic dysfunction might lead to a dissociation of circuits in the association cortex. In our patient, cranial CT showed that the principal areas of extensive bilateral calcifications included the striatum, the pulvinar thalami, and the cerebellar dentate nuclei (figure). Calcifications of the dentate nuclei are usually thought to result in cerebellar ataxia. Medial (internal) pallidal efferents are supposed to inhibit thalamocortical neurons, and an increased thalamocortical drive to lateral orbitofrontal and anterior cingulate cortex due to lesions near the pulvinar thalami might lead to auditory hallucinations.[4] There is still considerable controversy regarding a link