CENTRAL PONTINE MYELINOLYSIS WITH COMPUTERISED TOMOGRAPHY CONFIRMATION: A CASE REPORT

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SYNOPSIS

A 34 years old patient who developed central pontine myelinolysis after 6 months of severe drinking is described. Although the diagnosis of central pontine myelinolysis is usually made at post mortem, it may be suspected clinically on the basis of alcohol abuse, electrolyte disturbance, and progressive neurological deficits. This patient had CT scan showing symmetrical areas of low attenuation in the brain stem which helped confirm the diagnosis. Prompt correction of electrolyte imbalance resulted in good recovery.

INTRODUCTION

In 1959, Adams, Victor and Mancall (1) published their now well recognised syndrome of 'central pontine myelinolysis' (CPM). The disease as described in their patients, occurred on a background of alcoholism and malnutrition, and the characteristic histopathological finding was a symmetrical central zone of myelin destruction in the pons with sparing of axons and nerve cells. Quadriplegia and pseudo bulbar palsies were the main clinical findings. It is now appreciated that CPM may be associated with many disease states like chronic alcoholism (2) malnutrition (3) hyponatraemia (4-8), toxin (4), chronic debilitation (9-11), cerebral ischaemia (12), cerebral oedema (13), dehydration (14), Inappropriate secretion of anti diuretic hormone (7,8), acute haemorrhagic pancreatitis (14), liver cirrhosis (15), amyloidosis (13), leukaemia (16) and many others.

Although the diagnosis of CPM is almost exclusively made post-mortem, it may be suspected clinically on the basis of the following criteria made by Messert et al (17): 1) electrolyte distrubances manifested mainly by hyponatraemia; (2) progressive neurological deficits resulting in a 'locked-in- syndrome (3) usually, but not necessarily, alcohol abuse; (4) frequent iatrogenic precipitation of the syndrome by inappropriate rehydration of patients at risk. With the advent of computerised tomography (CT), premortem diagnosis has become a reality (18). We also report a case with typical CT findings. Unlike previous reports, our patient recovered and had serial CT scans done.

CASE REPORT

This 34 year old Chinese man was admitted on 14th October 1981 to a Medical Unit for complaints of loss of appetite, loss of weight and lethargy for six months; weakness of lower limbs for one month and diarrhoea for four days. Six months ago, he was retrenched from his job as a salesman and he became so depressed that he took to heavy drinking of alcohol. On admission, he was ambulant but described as being weak, lethargic and pale. There was no neck stiffness and no other neurological deficit detected. His cardio respiratory systems were normal. BP 120/70 mmHg, pulse rate 78 per min. Initial laboratory data were significant in that the serum sodium was 95 mmol/litre with serum osmolality of 276 mOsm/litre and urine osmolality of 424 mOsm/litre. Other investigations on admission were as follows: serum potassium 3.0 mmol/litre, chloride 91 mmol/litre, urea 6 mg%, calcium 7.8 mg%, phosphate 4.5 mg%, Total protein 5.9gm%, globulin 2.8gm%, bilirubin 2.6 mg%, alkaline phosphatase 400 iu/litre, SGPT 46 iu/litre, SGOT 160 iu/litre, CXR — bilateral opacities over both upper lobes with cavities suggestive of pulmonary tuberculosis, sputum for AFB smear and culture were negative. The patient was given an infusion of hypertonic 3% sodium chloride solution with potassium supplement and mist Kaolin et opii. He was noted to be noisy and'out of control' on second night of hospitalisation, and irrational and disorientated on third hospitalisation day. On the fourth hospital day, he developed acute respiratory distress syndrome with Pa02 50.8 mmHg and PaCO2 29.7 mmHg and was intubated and kept on artificial respiration for the next ten days. On the eight hospital day, the nursing staff in the intensive care unit noted that he was not moving all four limbs. Clinical examination showed flaccid quadriplegia with increased jaw jerk, bicep and tricep jerks but absent knee and ankle jerks. Both plantars were down going. He was conscious but uncommunicative with labile emotion. Eye movements were normal but there was absent gag reflex and he was unable to protrude his tongue. Further investigations done were as follows: EEG showed frequent theta activity and theta-delta activity over both temporal regions without lateralisation; CT scan (figure 1) showed a hypodense area in the pons and mid-brain with no mass effect. There was no enhancement after injection of contrast medium. He was diagnosed as having central pontine myelinolysis and peripheral neuropathy as a result of alcohol abuse with pulmonary tuberculosis.

He was given a course of thiamine intramuscularly and three per cent hypertonic saline together with antituberculous drugs. The electrolyte imbalance was rapidly corrected, and he made a slow but steady recovery from the 'locked-in' state. A repeat CT scan done six weeks later showed similar findings (figure 2). He was able to walk home unaided on 7 March 1982 after a course of rehabilitation.

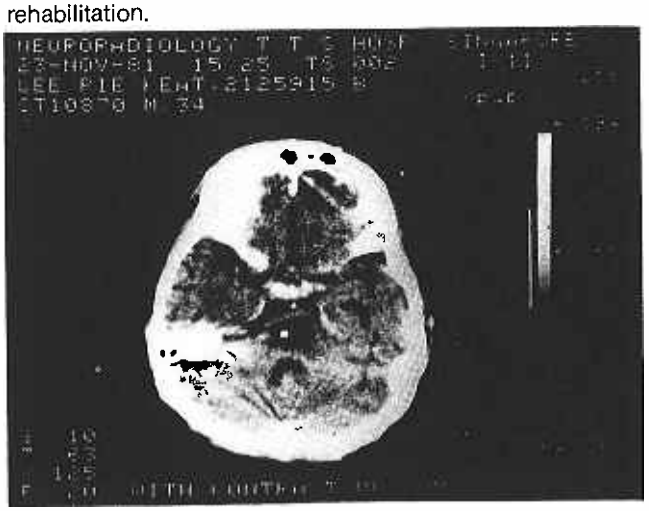

Figure 1 — First CT Scan shows a large hypodense area in the middle of the pons. There is no enhancement after infusion of contrast medium.

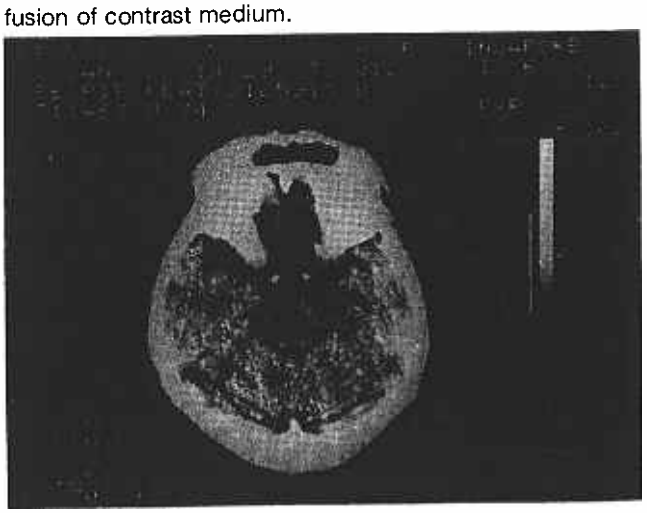

Figure 2 — Second CT Scan six weeks later shows a persistent area of radiolucency in the same area as in Figure 1.

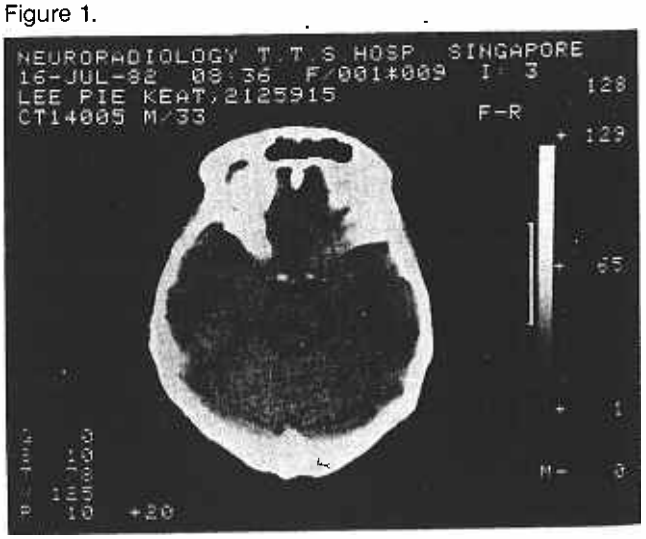

Figure 3 — Third CT Scan eight months later shows some diminution in size of the radiolucent area in the pons.

When he was seen again on 2nd July 1982, cranial nerves were intact and all the deep tendon reflexes were present though slightly hyperreflexic. There were only minimal weakness of upper motor neurone pattern over both lower limbs. However, there was not much changes in the repeat CT Scan (Figure 3). He was reviewed again one year later and his condition has remained stable.

DISCUSSION

Our patient has the typical clinical picture of CPM. His sickness developed over a period of one to two weeks after hospitalisation and the maximum neurological deficit persisted for 2-4/52 before recovery. Improvement was gradual over a period of many months and this was most probably due to clearing of oedema in the brain stem. However continuing improvement after six months suggests re-myelination as serial CT scan shows some diminution in the size of hypodense area.

It has been postulated that chronic alcoholism leads to impairment of liver function and hyponatraemia in the body. When the patient was admitted to hospital, there was a sudden cessation of alcohol intake; and it has been shown that 2-3 hrs after the last alcohol intake the return of anti-diuretic hormone would cause the decrease in urine flow resulting in water retention. And, if volume replacement was given at this time, it would precipitate hyponatraemia leading to brain oedema. Oedema of the pons would strangulate the myelin sheath of the transverse fibers in the ventral pons. In mild cases, this produces transient reversible long tract signs. However in more severe cases, this will lead to demyelination and death.

A review of the literature showed that seventy per cent of the reported cases had documented hyponatraemia (5, 19). Despite this observation, the role of sodium derangement in the genesis of CPM is far from clear. To begin with, hyponatraemia is not a universal finding. In fact, some showed hypernatraemia (5, 19). However, correction of electrolyte imbalance is still of paramount importance in managing the patient.

The patient described in this report satisfied the criteria of diagnosis postulated by Messert et al (17). Although there was no histopathological confirmation, we considered the CT findings as diagnostic. It was identical to that described by Thomas et al (18) in whose case the post-mortem features were characteristic. We were able to define the lesion on more than one CT slices and to show it on three scans performed six weeks and six months apart. In the absence of mass effect and displacement, the differential diagnosis of the CT scan include a large infarct, syringobulbia, large area of demyelination from multiple sclerosis. However, these were unlikely when the clinical picture was considered.

Given the usual fatal cause of the disease, early treatment is essential for survival. Wiederholt et al (20) stressed the importance of suspecting CPM when the clinical picture is appropriate, since patient may recover if vigorously treated. This report demonstrates the usefulness of CT scan for early diagnosis of CPM, and thus facilitates early treatment. However, the brain stem is a difficult area to visualise clearly on CT, and the CT may be normal even in autopsy proven cases (17). In the treatment of CPM, the electrolyte disturbances must be reversed without increasing the cerebral oedema, and concomitantly anti-oedema measures must be taken in order to be successful in the management.

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