UNUSUAL NEUROLOGICAL MANIFESTATIONS IN HYPEROSMOLAR HYPERGLYCAEMIA NONKETOTIC COMA

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SYNOPSIS

A middle aged woman who presented with neurological signs suggestive of brain stem infarction was subsequently found to have hyperosmolar hyperglycaemic non ketotic coma (HHNK). She recovered completely following treatment of the underlying metabolic abnormality. This report highlights the bizarre neurological manifestation of HHNK and the value of routine glucose estimation in cerebrovascular accident.

INTRODUCTION

Modern awareness of hyperosmolar hyperglycaemic non-ketotic (HHNK) coma is generally attributed to Sament and Schwartz.[1] Since then several reviews have attempted to bring this complex medical problem into clear focus.[2,3,4] Maccario was the first to highlight the bizarre neurological manifestations of this condition.[5] We report an unusual case of HHNK which presented with signs suggestive of brain stem infarction and which recovered completely following treatment of the underlying metabolic abnormality.

CASE REPORT

ZBA, a 56 year old Indian female housewife was admitted on 22 August 1978 because of stupor. The patient was well until four days prior to admission when she developed motor seizures that started in her right leg and became generalised about 5 minutes later. During the seizures she remained conscious and was able to talk. There were more than ten seizures daily. On the day of admission, she was noticed by a general practitioner to have focal seizures and a temperature of 105° F (41° C). There was no past history of fits, head injury or diabetes mellitus. She was on irregular treatment for hypertension of two years' duration.

On admission the patient was unconscious, not responding to painful stimuli. Blood pressure was 170/80 mmHg, pulse rate 100/min, temperature 103° F (39.5° C). There was no acidotic respiration. Skin turgor was poor. Both pupils were pin-point, oculocephalic reflex was absent and all four limbs were flaccid. She was areflexic with bilateral upgoing plantar response. There was no neck stiffness. Kernig's sign was negative. While in the ward, she developed focal seizures starting from the right foot and involving mainly the right side of her body. These seizures were not controlled by intravenous diazepam. She was diagnosed to have cerebropontine haemorrhage. Laboratory data revealed: Hb 13.8 g/dl, blood sugar 54 mmol/L, blood urea 23.3 mmol/L, Se Na^+ 137 mmol/L, Se K^+ 4.8 mmol/L, Se C1 96 mmol/L.

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There was pronounced glycosuria 4+ and no acetonuria. Calculated serum osmolality was 342 mOsm/Kg, blood pH 7.245, Pco_2 38 mmHg, PaO_2 70 mmHg, standard bicarbonate 16 mmol/L. Fasting cholesterol and triglycerides were normal. In view of the marked hyperglycaemia, hyperosmolality, gross glycosuria and no acetonuria, she was thought to have hyperosmolar hyperglycaemic non-ketotic coma secondary to an underlying stroke.

She required a total of 50 units of soluble insulin administered 5 units intramuscularly hourly to correct her hyperglycaemia from 54 mmol/L to 11 mmol/L. As she was not grossly dehydrated, fluid replacement consisted of 500 mls of normal saline, followed by 2000 ml of hypotonic (½ strength) saline. Biochemical abnormalities returned to normal 14 hours after insulin and fluid replacement but her clinical improvement was apparent only 48 hours later. She became more conscious, her pupils dilated and motor power returned.

On the fourth admission day, there was no neurological deficit apart from a left concomitant squint. At this stage electroencephalography showed mild diffuse slow wave activity which persisted for two weeks. She was discharged well on lente IZS 36 units every morning.

DISCUSSION

The entity of hyperosmolar hyperglycaemic non ketotic (HHNK) diabetic coma is characterised by extreme elevation of blood glucose level and absent or minimal ketonaemia. The patient normally is elderly with either nondiagnosed or mild diabetes mellitus and presents with signs of frank coma or milder degrees of central nervous dysfunction.[2,3] Mortality approaches 60% in some series.[2] However the extent to which neurological dysfunction can result from HHNK coma is not commonly recognised. These can vary from alteration of consciousness to hallucinations, seizures, focal neurological signs, abnormal tone, vestibular dysfunction and signs of meningeal irritation.[5] Because of the neurological findings, patient's age and lack of diabetic history, an initial diagnosis of cerebrovascular accident is common.

This case is reported because of the unusual neurlogical signs. To our knowledge there have been no reports of patients presenting with neurological deficits similar to our patient. The combination of deep coma, pinpoint pupils, absent oculocephalic reflex, hypotonia, hyporeflexia, bilateral Babinski response and hyperpyrexia suggests that there is a brain stem lesion. Maccario in his review on HHNK pointed out that deep coma is the rule and seizures, when present, are frequent, focal in nature and resistant to anti-convulsant therapy.[5]

Our patient had focal seizures for four days before she was admitted in stupor. During these seizures she remained conscious and was able to talk. This presentation was present in two out of seven patients with HHNK coma reported by Macarrio in 1968.[5]

Hyperthermia in HHNK coma is usually a terminal event[7] and pupillary abnormalities[8] are not reversed by treatment of the underlying hyperosmolality. Although our patient had pupillary changes and hyperpyrexia (41°C) she made a complete recovery indicating that these changes are not necessarily terminal events in patients presenting with hyperosmolar coma.

Neurological dysfunction associated with HHNK is not commonly recognised. The presence of overt focal neurological signs in these patients have been interpreted as evidence that 'strokes' are an aetiological factor in the development of hyperglycaemia.[9]

However, the reversibility of neurological symptoms, signs and electroencephalographic abnormalities with control of the metabolic imbalance on one hand and on the other, the fact that neuropathological findings have been absent and angio-graphic studies unrevealing,[6] all point to a direct causal relationship between hyperglycaemia and the neurological signs. Serum hyperosmolality leading to intracellular dehydration and disturbance of electrolyte equilibrium of the intracellular and extracellular compartment is the probable etiologic factor.[2,3]

This case emphasises the importance of routine blood sugar estimation in all patients who present as 'cerebrovascular accidents'. The aim of treatment should be the smooth restoration of disordered clinical and biochemical state with minimal morbidity and mortality. It is indeed satisfying to note that hyperthermia which was thought to be a terminal event in previous cases can be reversed with appropriate therapy.

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