Diabetic non-ketotic hyperglycaemia presenting as chorea — a case report

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Summary

We report a patient with hyperosmolar non-ketotic hyperglycaemia who presented with chorea and septic arthritis on his knee. The chorea resolved completely and quickly with correction of the metabolic disturbance, only to return just as quickly when his metabolic disturbance subsequently deteriorated as a result of overwhelming septicaemia, suggesting coexisting cerebreal ischaemia, although the basis of focal neurological sign in non-ketotic hyperglycaemia remains controversial.

Key words: Hyperosmolar non-ketotic hyperglycaemia, chorea, focal neurological sign.

Introduction

Hyperosmolar non-ketotic hyperglycaemia is a well-known complication affecting elderly non-insulin dependent diabetics. It may develop insidiously or be precipitated by an acute stress like an infection and may make its first appearance in patients with undiagnosed diabetes.

We report here an elderly patient with undiagnosed diabetes, who presented with chorea due to hyperosmolar non-ketotic hyperglycaemia. Such a presentation has only been previously reported once in the medical literature. Lack of awareness of such conditions may be an important factor in the underdiganosis of hyperosmolar non-ketotic hyperglycaemia coma.

Case Report

A 56 year old Malay man presented with acute onset of jerky, irregular involuntary movements of the right upper limb. The movements occurred almost continuously every five to ten seconds. The patient was otherwise fully alert and was able to give full history. He had been having increasing pain and swelling of his left knee over the last two weeks associated with fever and nocturia. The patient had been treated for hypertension. However, he had stopped taking his medication for one month. He was not known to have diabetes.

On examination at admission, he was afebrile. His blood pressure was 130/110, pulse rate 108/minute. His left knee was red, swollen and tender. Apart from the persistent irregular jerky movements of his right upper limb, he was fully conscious and the rest of the neurological and systemic examination were unremarkable.

A diagnosis of cerebrovascular accident with hypertension was made by the admitting doctor and the patient was treated with metoprolol 100mg bd, soluble aspirin 75mg daily, and indomethacin 50mg tds for the painful knee. Over the following twelve hours, his condition deteriorated. He became increasingly drowsy and tachypnoeic. The jerky choreaform movements of his right upper limb persisted though less frequently. The deterioration was interpreted as progressive cerebral infarct. Intravenous diazepam 10mg was given to control the involuntary movements without success. Intravenous dexamethasone was also given.

The physician was asked to review the patient's condition. It was noted the patient was grossly dehydrated. The glucostix test for blood glucose was 'high' (> 22 mmol/l) and the patient's urine contained 2% glucose, though urinary ketones were negative. A tentative diagnosis of hyperosmolar non-ketotic hyperglycaemia was made and treatment commenced with rehydration, insulin, potassium replacement and intravenous heparin. Further intravenous diazepam 10mg was given to control the involuntary movement again without success.

Investigations at this stage revealed blood glucose 30 mmol/l, haemoglobin 17.6 gm/dl, packed cell volume 50%, blood urea 26 mmol/l, serum sodium 129 mmol/l, serum potassium 2.3 mmol/l, serum chloride 90 mmol/l. The calculated serum osmolarity was thus 318.6 mosm/kg.

Within twelve hours, the patient improved dramatically. He was fully conscious and able to take his breakfast. The jerky movements had disappeared. His state of hydration as well as his blood profile had improved:–
blood urea 20 mmol/l, serum sodium 140 mmol/l, serum potassium 3.5 mmol/l, blood glucose 13.8 mmol/l, haemoglobin 13 gm/dl, and packed cell volume 35%.

However, the next day, he had intermittent high-fever and became increasingly toxic. The jerky movements relapsed and his hyperglycaemia worsened. Frank pus was aspirated from his swollen left knee and gram positive cocci isolated. Intravenous cloxacillin was added to his treatment. He failed to improve and died in septicaemic shock with renal failure two days later.

Discussion

Non-ketotic hyperglycaemia is probably underdiagnosed.[1] In Mentakab District Hospital, not a case was diagnosed for at least the last three years. Early diagnosis and treatment are important because late diagnosis with alteration in consciousness is associated with worse prognosis.[2] This probably accounts for the reported high mortality rate of 40%–70%.[1]

Hyperosmolar coma represents only one extreme of the spectrum of hyperglycaemia. At the lesser extreme of hyperglycaemia and hyperosmolarity, other neurological manifestations had been well described.[3–6] When the underlying hyperosmolarity remained undetected and untreated, coma eventually followed. The types of neurological manifestations included focal and generalised seizure, focal neurological signs (hemiparesis, Babinski reflex, hemisensory defects, hyperreflexia, aphasia, heminopia), visual hallucination, myoclonic twitch, nystagmus, and signs of meningeal irritation. There is previous report of choreoathetosis occurring in non-ketotic hyperglycaemia.[7] This report represents the second.

All these neurological manifestations were readily reversible with correction of the metabolic disturbance.[1,3–6] In patients who presented with seizure, they did not respond to anticonvulsant.[3–6] In our patient, the chorea failed to be suppressed by sedation, but

responded dramatically to correction of the hyperosmolar hyperglycaemia. When control slipped again as a result of the overwhelming septicaemia, the chorea returned.

A lack of awareness of the association between hyperosmolar non-ketotic hyperglycaemia and focal neurological signs may lead the signs interpreted to be due to cerebrovascular disease and the hyperglycaemia to be due to stress or coexisting uncontrolled diabetes, thus missing the diagnosis. This was obvious in this case and indeed, the most common admitting diagnosis in patients with non-ketotic coma is stroke.[2,5] The presence of coexisting structural lesion such as cerebrovascular disease which may occur in elderly patient, especially in the setting of non-ketotic hyperglycaemia which predisposes to intravascular thrombosis.[8,9] Further, many elderly diabetics may have preexisting vascular lesion such as old cerebral infarct[11] which is more susceptible to the metabolic disturbances in non-ketotic hyperglycaemia. This then provides a substrate for the focal neurological sign.

However, in many patients presenting with neurological signs, neuropathological and angiographic studies[3,10] had failed to disclose any structural abnormalities. Further, the rapid reversibility of the neurological signs with correction of the metabolic disturbance points to a direct causal relation between hyperosmolar hyperglycaemia and the neurological signs. The mechanism underlying this is however difficult to explain. Hyperosmolarity in itself is an insufficient explanation as most patients with non-ketotic hyperglycaemia do not have marked hyperosmolarity and neurological manifestation is rare in diabetic ketoacidosis which have similar hyperosmolarity and hyperglycaemia.

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