Multiple adenomatosis presenting with psychiatric manifestations

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Though hyperinsulinism is a common manifestation of multiple adenomatosis and the association between hypoglycaemia and mental disturbance is well recognized, only one instance of multiple adenomatosis presenting as an acute psychosis—depression—has been reported (Fullerton, Lohrenz & Holrey, 1967). There is no record of an association with mental retardation.

For this reason we present a case of multiple adenomatosis presenting with confusional episodes and violent behaviour in a mentally retarded patient.

Case Report

The patient, a 27-year-old warehouseman, was referred (March 1967) as a psychiatric emergency because of several bouts of aggressive and destructive behaviour. He was disorientated and behaved inappropriately in the attacks, and also sweated and showed a violent tremor. There were no convulsions. He had no subsequent recollection of the attacks.

History. He had always been backward. Nine years previously, when examined after a head injury with transient unconsciousness, he had shown unsteadiness, tremor, difficulty in reading, writing, calculating and memorizing (only three digits forwards and backwards)—signs not attributed to head injury but linked with his subnormality—and pes cavus.

Family History. His father had suffered from similar bouts of violent behaviour, particularly if (according to his mother) the evening meal was late. He had died following operation and necropsy had shown an 'islet cell secreting tumour of the pancreas and calcification of the kidneys'. No record of the condition of the parathyroid glands was made.

On examination, he was orientated and co-operative, but slow in grasp. Blood pressure was 130/80.

Investigation. Hourly blood sugars were taken during a prolonged fast. At 16 hr he became violent and the blood sugar was found to be 20 mg/100 ml. Glucose immediately restored the level to normal and the violent behaviour ceased. Electro-encephalography showed on hyperventilation a paroxysm of slow activity, maximal on the left side.

Serum insulin was measured by the radio immuno assay method (Dr Vincent Marks). The fasting...
serum insulin was high (86 μU/ml) and rose to over 200 μU/ml after glucagon 1 mg i.v. Sodium tolbu-
amide 1 g i.v. produced a rise of serum insulin from a fasting level of 75 μU/ml to over 200 μU. Serum calcium was 12 mg/100 ml, serum phosphate 2.8 mg/100 ml and 24-hr urinary calcium excretion 370 mg. Overnight gastric secretion was 590 ml (31.3 mEq hydrochloric acid). Ratio of basal to histamine-stimulated acid secretions was 4.6/21.9.

Laparotomy. (Mr H. Smith:) a number of islet cell tumours were found. The largest, in the head of the pancreas, was excised, and the distal half of the pancreas resected. The patient had no further attacks and returned to work.

Three months after the operation serum and urinary calcium were unchanged. A tolbutamide tolerance test was in the upper limits of normal.

Postoperative psychometric investigation. His verbal IQ was 75, and his performance and full-scale IQ's 71. There was a striking impairment of psychomotor organization. It was concluded that he was of borderline subnormal intelligence, the pattern of responses was 'rather organic' and, since there was no elevation of performance over verbal ability, there had been no spectacular increase since the operation.

Discussion

Though the initial differential diagnosis included aggressive psychopathy, subnormality with behaviour disturbance and post-traumatic epilepsy, the occurrence of confusion with a lowered blood sugar and its relief by restoration of the blood sugar to normal, showed that these episodes were due to spontaneous hypoglycaemia. Furthermore, no EEG epileptogenic focus was seen. Multiple endocrine adenomatosis is transmitted as a dominant autosomal trait and characterized by adenomas in pancreatic islet cells and in the pituitary, adrenal and thyroid glands: a gastrin-secreting tumour of the pancreas is sometimes associated (Harrison, 1967). In the present patient the diagnosis was made by the demonstration of multiple islet cell tumours and hyperparathyroidism (from the serum calcium and phosphorus and the urinary calcium excretion values), and the family history. The overnight gastric secretion test results excluded a gastrin-secreting tumour and no evidence of pituitary, adrenal or thyroid dysfunction was found.

Backwardness had apparently been present from early infancy and the 1956 neurological findings and the psychometric test results suggested a chronic cerebral abnormality. Though infantile hypoglycaemia can give rise to mental retardation, neurological sequelae and fits, multiple adenomatosis is an uncommon cause of hypoglycaemia below the age of four (Marks & Rose, 1965). It is thus possible that other endocrine abnormalities were aetologically significant. Unlike a case of chronic psychos- remain after removal of an insulinoma (Markwitz, Slanetz & Frantz, 1961), operation did stop the acute psychotic episodes in our patient; nevertheless it is noteworthy that the mental retardation was not affected.

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References


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