CASE PRESENTATION

The First Report of Subtotal Pancreatectomy for Nesidioblastosis in Bahrain

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ABSTRACT

A case of nesidioblastosis is reported. It occured in a one year old male Bahraini neonate who was a product of full term normal delivery. Soon after birth he was noticed to be jittery, and dextrostix was zero. His blood glucose was persistently low and his serum insulin between 85 and 133 mm/L. Inspite of high rate glucose infusion, hydrocortisone and diazoxide, his hypoglycaemia persisted. Initially the father refused surgical intervention. However subtotal pancreatectomy was performed at nine days of age. Post-operative serum insulin estimation showed a dramatic reduction and blood glucose slowly returned to normal. The baby had an uneventful post-operative recovery. The child is now three years of age. His recent evaluation will be discussed. We present this case to emphasize the necessity of early diagnosis of nesidioblastosis, to differentiate it from other types of neonatal hypoglycaemia, and to denote the importance of subtotal pancreatectomy before cerebral damage occurs.

Hyperinsulinism is the most common cause of persistant hypoglycaemia in children under one year of age [1]. Early and effective treatment is mandatory to avoid neurological sequelae [2]. Banting and Best isolated insulin as an active hormone in 1922 [3]. In 1934 Graham and Hartman cured a one year old girl with hypoglycaemia by 80% subtotal pancreatectomy [4]. Nesidioblastosis, a term coined by Laidlaw in 1938, denotes the formation or budding of new endocrine cells from pancreatic duct epithelium [5]. Thus insulin secreting cells are found outside the normal islet of Langerhans.

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In recent years subtotal pancreatectomy has been advocated for treatment of nesidioblastosis. The result of surgery will depend on the timing of the procedure before onset of neurological deficit, and the amount of the pancreatic tissue which has been removed. We present our experience with a first subtotal pancreatectomy in a Bahraini male neonate treated at Salmaniya Medical Center.

THE CASE

AEH was a product of full term normal delivery. His birth weight was 4730 gms. Soon after birth, the baby was noticed to be jittery. His brother died at one month of age of hypoglycaemia. There was no maternal history of diabetes mellitus. His initial blood glucose was 12 mg/dl, Hypoglycaemia persisted, inspite of dextrose infusion of 15 mg/Kg/min. A full laboratory analysis revealed an inappropriately raised plasma insulin of 133 mu/ml with blood glucose of 12 mg/dl, but all other tests were normal.

Following the diagnosis of hyperinsulinism, he was started on a high carbohydrate intake 15 mg/kg/min, diazoxide and steroids. He continued to have periodic symptomatic episodes of hypoglycaemia. Initially, the father refused surgical intervention. At nine days of age he had a subtotal 95% pancreatectomy. On the first post-operative day his blood glucose increased to 91 mg/dl and plasma insulin to 17.5 mu/ml. He had uneventful post-operative recovery.

At the age of three years, he weighed 14.5 Kg. His height was 101 cm, and his neurological examination showed slow speech and mild spasticity of the limbs.

PATHOLOGICAL EXAMINATION

There were no detectable microscopic abnormalities in the pancreas. Microscopic sections showed features of nesidioblastosis. The islets of Langerhans appeared prominent with many cells showing nuclear enlargement. In some areas the islet cells appeared to have extended between the surrounding exocrine cells, but no definite neoformation of islet cells was recognized. The immunoperoxide staining demonstrated insulin production, but no increase in glucagon.

DISCUSSION

Nesidioblastosis has been reported as a group of morphologic features representing an increase in the amount of endocrine tissue [7], and possibly maldistribution of endocrine cells [8]. In hypoglycaemia with inappropriate production of insulin, medical management is the initial treatment, but when drug treatment has not controlled this disorder subtotal pancreatectomy is recommended.

A 75% subtotal pancreatectomy usually fails to control hyperinsulinic hypoglycaemia. 95% subtotal pancreatectomy gives satisfactory results with few recurrences. Total pancreatectomy is not advisable as a primary procedure because it is more dangerous, diabetes is inevitable, the effect of the other pancreatic hormones in an infant are unknown, and nesidioblastosis cannot be diagnosed on frozen section. Alloxan can be a useful cytotoxic drug specific to B cells. It is recommended for unsuccessful subtotal pancreatectomy [9]. The disadvantage of alloxan is the lack of knowledge of the long term side effect.

CONCLUSION

We conclude by emphasizing the importance of early diagnosis of hyperinsulinic hypoglycaemia and the need for an ealry surgical intervention if the medical therapy failed.

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