A rare case of focal nesidioblastosis causing adult-onset hypoglycemia

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Abstract. Nesidioblastosis is a major cause of persistent hyperinsulinemic hypoglycemia of infancy and is caused by hypertrophy of the pancreatic endocrine islands. The disease can be categorized histologically into diffuse and focal forms. The condition rarely occurs in adults and only one adult case of suspected, but not histologically confirmed, focal nesidioblastosis has been reported. The present study describes the case of a 62-year-old man suffering from symptomatic hypoglycemia for 3 years and exhibiting a nodule in the pancreatic tail. Pathological evaluation following surgical enucleation of the pancreatic body and tail revealed focal nesidioblastosis. The hypoglycemic symptoms of the patient disappeared postoperatively. To the best of our knowledge, this is the first histologically-confirmed case of focal adult nesidioblastosis, suggesting that the possibility of nesidioblastosis should be taken into account in adult patients with persistent hypoglycemia.

Introduction

Hypoglycemia in nondiabetic patients is not a common clinical problem and can be a diagnostic and therapeutic challenge (1). Persistent hyperinsulinemic hypoglycemia (PHH) is a functional disorder caused by aberrant insulin release by pancreatic β cells (2). Nesidioblastosis is the major cause of PHH in infants and children, but in adults it is usually a consequence of a solitary insulinoma. Nesidioblastosis has been reported infrequently in adults (1-3). It should be noted that in pediatric patients nesidioblastosis may be classified histologically as either diffuse or focal, but only diffuse lesions have been reported in adult patients with histologically-confirmed nesidioblastosis. A single case of suspected focal nesidioblastosis in an adult was reported by McElroy et al in 2010 (4), but was not confirmed histologically. Due to the lack of evidence, most physicians do not take a possibility of focal nesidioblastosis into account when confronted with an adult patient with PHH.

The present study focused on the case of a 62-year-old man with a 3-year history of intermittent episodes of symptomatic hypoglycemia. A 72-h fasting test, elevated levels of insulin and C-peptide, concomitant with decreased blood glucose levels and imaging, led to the discovery of a nodule in the pancreatic tail. The pancreatic corpus and tail were enucleated laparoscopically and the presence of focal nesidioblastosis was confirmed histologically. We propose that focal nesidioblastosis should be taken into consideration when confronted with PHH, even in middle-aged patients.

Case report

Case presentation. A 62-year-old man with a body mass index of 26.99 presented with a 3-year history of intermittent episodes of dizziness, weakness and sweating, which were apparently associated with work load and subsided upon food intake. Previous clinical evaluations had not included blood glucose measurements. The patient had no history of diabetes, pancreatic diseases, von Hippel-Lindau or multiple endocrine neoplasia syndromes and denied using insulin or medications associated with hypoglycemia.

Based on the clinical presentation of the patient, a 72-h fasting test was performed to address the possibility of endogenous hyperinsulinemic hypoglycemia, but was discontinued after 61 h due to complaints of fatigue and dizziness. During the hypoglycemic episodes, laboratory tests revealed that the glucose, insulin and C-peptide levels were 48 mg/dl (normal range, 70-100), 162.20 pmol/l (normal range, 17.0-173.0) and 1.394 nmol/l (normal range, 0.37-1.47), respectively. A nodule in the pancreatic tail was observed on abdominal computed tomography (CT) (Fig. 1A), which prompted further examination using other imaging modalities. Magnetic resonance imaging (MRI) revealed the presence of an unevenly enhanced lesion in the pancreatic tail, measuring 2.2 cm in diameter (Fig. 1B and C). The patient underwent laparoscopic enucleation of the pancreatic body and tail following an initial diagnosis of a distal pancreatic tumor.

Investigations. The collected pancreatic specimen measured ~3.5x2.5x1.5 cm. This specimen was fixed in formalin, and

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hematoxylin and eosin-stained serial sections were prepared for microscopic examination by an expert pathologist. Immunohistochemical staining was also performed, using rabbit monoclonal antibodies against synaptophysin (1:1; #RMA-0537) and chromogranin A (1:1; #MAB-0202), and mouse monoclonal antibodies against neuron-specific enolase (1:1; #MAB-0584) and Ki-67 (1:1; #Kit-0005-2) purchased from Fuzhou Maixin Biotechnology Development Co., Ltd.
Persistent hyperinsulinaemia associated with neonatal hypoglycaemia (PHH) typically occurs in the first year of life (1). In adults, PHH is much less common, but is rarely encountered in adults. The term nesidioblastosis was coined by Laidlaw (5) in 1938 to describe the neoplastic nature of the islets. However, it is rare in adults, and its cause is unknown. In the present study, a case of nesidioblastosis is presented, and the histological characteristics of the disease are described. The patient of the present study developed diabetes mellitus following the resection of the pancreatic tissue. In the aforementioned case of suspected focal nesidioblastosis, euglycaemia was restored following simple enucleation of the lesion, which comprised 5% of the total pancreatic mass (4). In patients with focal nesidioblastosis, partial pancreatectomy would be expected to control the hypoglycaemia and result in a good long-term outcome.

In conclusion, focal nesidioblastosis is rare in adults and can be difficult to diagnose preoperatively. The present case report, however, suggests that physicians should consider focal nesidioblastosis when confronted with patients with PHH, since early detection may prevent unnecessary full resection of the pancreatic corpus and tail. Imaging can be useful, particularly when combined with moderately high levels of insulin following fasting; patients exhibiting these characteristics should be considered for partial pancreatectomy.

**References**


