'noninsulinoma pancreatogenous hypoglycemia syndrome' (NIPHS); A case report of adult nesidioblastosis from Turkey

Soner Cander, Ozen Oz Gul, Erdinc Erturk
Bursa Sevket Yilmaz Education and Reserach Hospital,
Uludag University Medical School, Endocrinology and Metabolism

Introduction

The most common reason for the rare condition of hyperinsulinemia-related hypoglycaemia is insulinoma, a tumor of pancreatic islet cells. However, nesidioblastosis characterized by diffuse or focal hyperplasia of the pancreatic islet cells is the most common cause of hyperinsulinemic hypoglycemia in newborns. Nesidioblastosis seen in newborns is now called 'persistent hyperinsulinemic hypoglycemia of infancy' (PHHI) while the condition in adults is called 'noninsulinoma pancreatogenous hypoglycemia syndrome' (NIPHS) as a separate entity. It is impossible to clinically differentiate insulinomas from NIPHS.

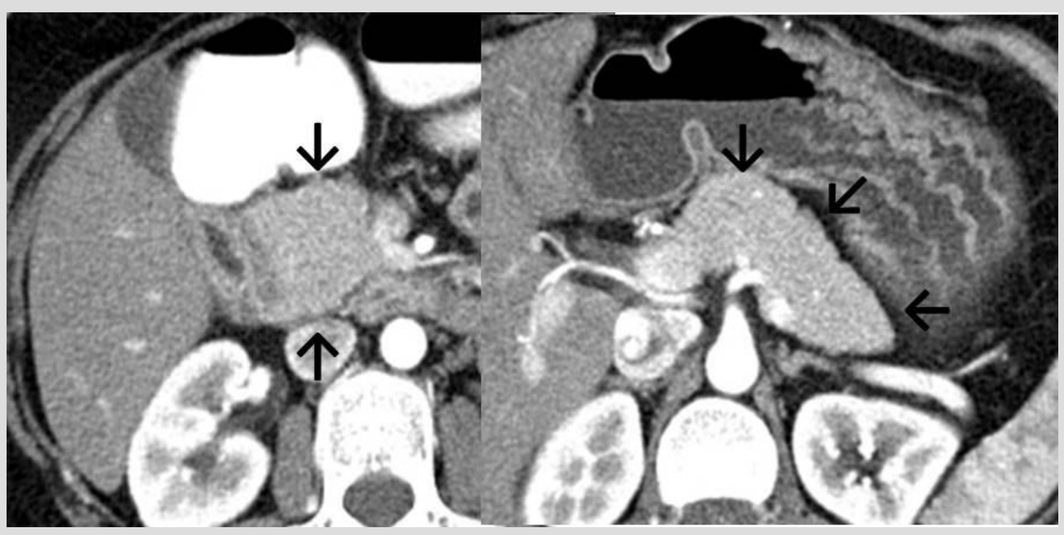
Case

- A 38-year-old female presented with neuroglycopenic symptoms in the form of drowsiness, inability to speak, numbness in the mouth, and nausea in the last 3-4 months.
- Endogenous hyperinsulinemia was found with recurrent neuroglycopenic symptoms (the glucose level was 25 mg/dl, insulin 43.9 µ/ml, C-peptide 5.54 ng/ml).
- The selective arterial calcium stimulation test (SACST) result was consistent with a diffuse disease in the body and tail.

Table: Preoperatively performed selective calcıum infusion test results of patient.

	SAD		SAP		GDA		HA		SMA	
	Ins	Glu	Ins	Glu	Ins	Glu	Ins	Glu	Ins	Glu
zero sec	14	59	19	57	17	51	22	51	12	53
30th sec	41	63	60	52	49	53	64	53	11	54
60th sec	54	62	56	56	52	52	47	53	12	55
90th sec	56	63	44	55	35	50	41	53	14	57
120th sec	50	63	37	52	28	52	33	53	15	56

SAD: Splenic Artery Distal part SAP: Splenic Artery Proximal part
GDA: Gastroduodenal Artery HA: Hepatic Artery
SMA: Superior Mesenteric Artery



- No lesion was found on imaging tests including enhanced computed tomography (CT) methods performed with a preliminary diagnosis of insulinoma. A suspicious hyperperfusion was present in the pancreatic tail on the perfusion CT examination performed after obtaining approval.

Figure 1: Head and corpus-tail sections of the pancreas (black arrows) were painted homogeneous in the 40th sec images after the application of water and injection of intravenous contrast material. Focal lesion was not determined and pancreas was stained uniformly in the other phases (images not added).

- The patient underwent partial (75%) pancreatectomy and is now followed up as a diabetes patient on intensive insulin treatment at the postoperative 38th month.

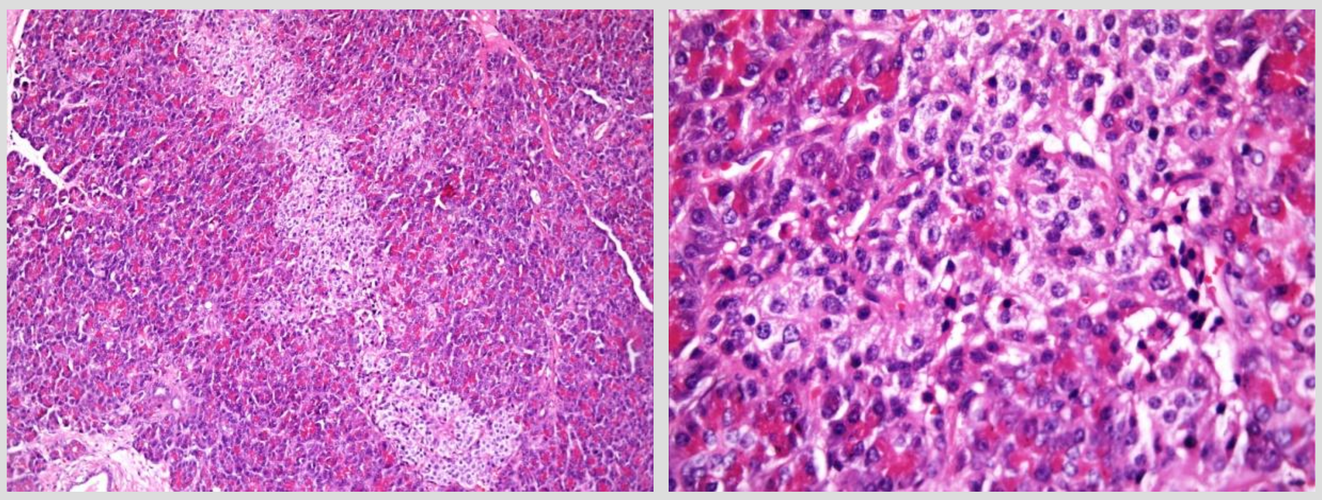


Figure 2: A) Microscopic image of an enlarged islet of endocrine pancreas cells with irregular margin from surrounding pancreas tissue (Haematoxylin&Eosinx100) B) Pleomorphic islet cells show increased nuclear size with abundant clear cytoplasm and prominent nucleoli (Haematoxylin&Eosinx200)

Conclusions

The NIPHS is rarely seen in the adult age group. SACST seems to be the most suitable test to differentiate diffuse or multiple disease from insulinoma and to guide the surgery when advanced radiological imaging methods are inadequate to detect the presence of insulinoma. Regarding perfusion CT, it would be more appropriate to wait for comparative data to be put forward in a more consistent manner. When no response to medical treatment, partial/total pancreatectomy is appropriate treatment option as it enables recovery from the hypoglycemic episodes despite leading to diabetes.

1.García-Santos EP, Manzanares-Campillo Mdel C, Padilla-Valverde D et al. Nesidioblastosis. A case of hyperplasia of the islets of Langerhans in the adult. Pancreatology. 2013;13(5):544-548.
2.Won JG, Tseng HS, Yang AH et al. Clinical features and morphological characterization of 10 patients with noninsulinoma pancreatogenous hypoglycaemia syndrome (NIPHS). Clin Endocrinol (Oxf). 2006;65(5):566-78.

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