Adult-Onset Nesidioblastosis, A Diagnostic and Management Dilemma

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Keywords

Nesidioblastosis, hypoglycemia, case report, hyperinsulinism, pancreatectomy

Case Report

We describe a case of a 49-year-old female patient with recurrent postprandial symptomatic hypoglycemia. The hypoglycemia was combined with increased insulin and C peptide but all diagnostic modalities for localizing an insulinoma were negative. Medical management didn't control symptoms and thus the patient underwent subtotal distal pancreatectomy. Surgical tissue audit confirmed the diagnosis of noninsulinoma pancreatogenous hypoglycaemia syndrome (NIPHS) or nesidioblastosis.

Description

Nesidioblastosis (NB) could also be a disease characterized by hyperfunctioning pancreatic β cells, usually associated with hypoglycemia within the presence of high endogenous insulin levels. it is necessary to rule out insulinoma or exogenous insulin of sulfonylurea administration when making the diagnosis. most commonly described in neonates and kids, but recent studies have shown an increased number of cases in adults. Focal NB: confined to a minimum of one discrete area of the pancreas. Diffuse NB: All better islet cells are affected. commonest explanation for persistent hyperinsulinemic hypoglycemia in neonates

Synonym(s): noninsulinoma pancreatogenous hypoglycemia (NIPHS); persistent hyperinsulinemic hypoglycemia of inception (PHHI); congenital hyperinsulinism; islet cell hyperplasia

Incidence

- Infants/children

o Sporadic: 1/30,000 to 1/50,000 live births

o Familial: 1/2,500 births are seen.

- Adults: 0.5–15% of organic hyperinsulinemias

o Annual incidence is 0.09/100,000.

o Peaks in 5th decade of life

Discussion

Nesidioblastosis was first reported in infants by George F. Laidlaw in 1938. He described nesidioblastosis as neoformation of Langerhans islets from the pancreatic ductal epithelium, and it's now known to be the primary explanation for persistent hyperinsulinemic hypoglycemia in infants. In Adults, hyperinsulinemic hypoglycemia is particularly caused by insulinomas, other causes are rare and typically because of adult-onset nesidioblastosis, which was first described in 1975. Since then fewer than 100 cases are reported but it seems to be increasing in frequency.

The explanation for adult-onset nesidioblastosis is unknown, but there seems to be an association with gastric bypass surgery. The excellence between insulinoma and nesidioblastosis preoperatively can sometimes be difficult, because the clinical presentation might be similar, and imaging studies are often equivocal. In patient with nesidioblastosis symptoms occur mainly postprandially and only rarely while fasting. In contrast, max patients with insulinomas have fasting hypoglycemia. Moreover, radiological localization studies, like CT scan, endoscopic ultrasound, octreotide scan, and selective arterial calcium stimulation test (SACST) with hepatic venous sampling, are often performed to differentiate between a focal abnormality (insulinoma) and a diffuse process (nesidioblastosis). Conventional radiologic testing isn't reliably helpful in differentiating an insulinoma from nesidioblastosis. While the diagnosis of nesidioblastosis should be considered when imaging studies (CT, resonance imaging, ultrasonography, and angiography) don't localize a discrete lesion of the pancreas, it should not be relied on. Many insulinomas are small) and even in experienced hands, the sensitivity of these radiologic studies for an insulinoma is simply 50% to 80%, and false-positive results occur also.

The most promising imaging technique for localization of an insulinoma is transgastric ultrasonography of the pancreas. Within the absence of detecting an insulinoma using this technique, the suspicion of a diagnosis of adult nesidioblastosis should be heightened. One should then proceed to percutaneous transhepatic portal venous sampling, or better yet, to a calcium-stimulated arteriogram of the pancreas. during this latter test, a selective arteriogram is performed of the gastroduodenal artery, splenic artery, superior mesenteric artery, and hepatic artery. After dye is injected into a given artery, a bolus of calcium is then injected. Insulin is sampled from a catheter placed during a hepatic vein. When an insulinoma is present, calcium injection results in the discharge of insulin as long because the precise artery that feeds the planet of the pancreas that contains that tumor is tested. On the other hand, within the presence of nesidioblastosis, calcium injection into all of the pancreatic vessels may end in insulin release from the entire pancreas, because the abnormal islets are usually found throughout the entire pancreas, this finding would be highly implicational nesidioblastosis within the absence of the MEN-1 syndrome, during which multiple insulinomas are often present.

While pancreatic resection represents the definitive treatment for an insulinoma, patients often require medication to manage

their hypoglycemia preoperatively or postoperatively. Diazoxide is that the foremost frequent medication used, but its use is amid troublesome adverse effects, including fluid retention, hypotension, hypertrichosis, and bone marrow suppression. Other medications that are used with varying success include somatostatin analogs and glucocorticoids. There are reports regarding the use of calcium channel–blocking agents to treat patients with insulinoma, neonatal nesidioblastosis, and reactive hypoglycemia.

Although there's limited data regarding the efficacy of pancreatectomy for nesidioblastosis, successful resolution of hypoglycemia has been reported after partial or subtotal pancreatectomy. On the other hand, recurrent hypoglycemia has also been reported which can require total pancreatectomy.

Other possible etiologies of hyperinsulinemic hypoglycemia include drug induced hypoglycemia, gastric dumping syndrome and exogenous insulin administration.

Summary: Nesidioblastosis could also be a rare explanation for persistent hyperinsulinemic hypoglycemia in adults. The incidence of nesidioblastosis in adults is typically thought to be very low. The β cell changes in adult nesidioblastosis suggest a dysregulation of the function of the cell. Histologically nesidioblastosis is almost always characterized by a diffuse islet cell hyperplasia throughout the entire pancreas. Clinically and biochemically, it's impossible to differentiate between diffuse nesidioblastosis and insulinoma. If all highly selective noninvasive imaging techiques fail to identify a tumor, selective arterial calcium stimulation testing should be performed. The last word diagnosis relies on the histopathologic evaluation and immunohistochemical confirmation as was exhausted our case.

Conclusion

Nesidioblastosis is an uncommon but clinically important explanation for hypoglycemia within the adult population, and will be considered during a patient with a presumptive preoperative diagnosis of insulinoma. Subtotal distal pancreatectomy could provide clinical benefit in refractory cases.

In summary, adult nesidioblastosis resulting in hypoglycemia may be a crucial entity that need to be understood by all surgeons who operate for an insulinoma. The optimal treatment of this condition requires further study. Whether or not a nonoperative approach to this disease will ever be practical remains to be determined.

Biography

Dr Pankaj Baweja has completed his MD Pathology from KMC Manipal, MANIPAL University in April 2007. He has more than 13 yrs. of oncopathology experience in various reputed hospitals in New Delhi, INDIA. He has published papers in reputed international and national journals. He is active member of Indian Association of Pathologists and Microbiologist (IAPM) and young Oncologist group (YOG). Presently he is working as Senior Consultant Histopathologist in Indraprastha Apollo Hospitals, New Delhi, India.