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Case Report

Nesidioblastosis In A Patient In Adulthood.

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Abstract

Nesidioblastosis is a rare condition among adults, affecting between 0.5 and 5.0% of the age range, producing persistent organic hyperinsulinemic hypoglycemia. Postprandial hypoglycemia is a common clinical presentation, and because there is no clinical context or very specific examination, the preoperative diagnosis is difficult and requires exclusion criteria. We describe a 47-year-old male patient who was diagnosed with neoblastosis in 2014 following repeated episodes of unconsciousness that had nothing to do with fasting. Following the failure of previous operations, the patient was submitted to Frey's approach. The patient's endocrine issue was successfully resolved by the chosen approach, which also had no negative effects on the gastrointestinal system.

Keywords : *Nesidioblastosis, Insulin, Hypoglycemia.*

INTRODUCTION

Not a common clinical issue, hypoglycemia in non-diabetic patients might present diagnostic and treatment challenges [1]. Both babies and adults can develop persistent organic hyperinsulinemic hypoglycemia (PHH), an endocrine pancreatic disease. PHH in neonates is primarily brought on by β-cell dysfunction, which is referred to as congenital hyperinsulinism (CH) [2,3] or nesidioblastosis [4,5]. In adulthood, nesidioblastosis or insulinoma are the two unusual causes of PHH [6–8].Nesidioblastosis in adult patients is defined by developing islets that emerge from the periductal epithelium, β-cell hypertrophy, and islets with larger and hyperchromic nuclei. It is also characterized by spontaneous hyperinsulinemic hypoglycemia that is not brought on by an insulinoma [10–12]. Postprandial hypoglycemia and biochemical findings similar to those of insulinoma are the main clinical features of this illness. Adults account for 0.5 to 5.0% of nesidioblastosis cases, according to published figures [13]. An adult male with a two-year history of sporadic episodes of symptomatic hypoglycemia has been reported to have nesidioblastosis as a result, necessitating multiple surgical procedures for a successful course of treatment. Case Presentation A 47-year-old male patient with a mean body mass index (BMI) of 41.4 kg/m² began seeing a doctor in 2012. He reported experiencing episodes of unconsciousness roughly six to eight times per day, along with a blood glucose test. capillary, frequently below 30 mg/dL, sporadic, and unconnected to fasting. As the symptoms grew worse, he sought medical attention, which led to an investigation and the possibility of nesidioblastosis.He was sent to Hospital Sírio Libanês (São Paulo-SP, Brazil) in 2014, where diagnostics revealed nesidioblastosis affecting the pancreatic head and tail. Consequently, he began receiving therapeutic pharmaceutical treatment using Diazoxide and Octreotide. But as his symptoms worsened, the patient had a Roux-en-Y gastric bypass and a body-caudal pancreatectomy in 2015. He claims that following the procedure, he was asymptomatic for three months before experiencing hypoglycemia symptoms once more. Due to intestinal invagination, the Rouxen-Y gastric bypass had to be undone in one of the two surgical procedures the patient needed in 2016 after presenting with semi-intestinal occlusion. He experienced acute hypoglycemia in 2017 and went back to look for professional medical care. Associated comorbidities were refuted by him. Given the circumstances, a selective calcium stimulation test was conducted [Figure 1], revealing an excess of insulin synthesis in the area that is irrigated by the gastroduodenal artery (GDA) and the superior mesenteric artery (SMA). Furthermore, an abdominal computed tomography was conducted [Figure 2], which revealed no neoplasic processes but rather only alterations in the abdominal anatomy brought on by prior surgeries. In addition to the surgeries previously

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discussed in 2015 and 2016, there was also a partial left nephrectomy in 1999 to remove a sarcoma.Following that, a subtotal pancreatectomy with duodenal preservation was selected as an open surgical reintervention to decrease the amount of pancreatic tissue that remained. The procedure is performed under general anesthesia while the patient is in a supine position. After the front scar was removed during a Chevron laparotomy, the retractors were positioned, and the dissection process was started by releasing any prior adhesions. In the subsequent

Following the completion of a broad Kocher and Catell procedure, the superior mesenteric vessels were dissected and the vessels that connected with the pancreas were tied off. The head and the pancreatic uncinate process were resected using the Frey technique, while the dissection was carried out using UltraCision throughout the process [Figure 3].The patient's blood glucose was checked every 30 minutes during the procedure, and it rose to levels above 200 mg/dL following the removal of the uncinate process and a sub-total part of the pancreatic head. Cholecystectomy and cholangiography [Figure 4] were then carried out to confirm the integrity of the resected area after biological glue had been applied. of the bile duct retro-pancreatic. Due to the complexity of the case, a 24 Fr Blake drain was also placed near the resection and exited through the counter-opening, connecting to the reservoir. Additionally, an appendectomy was conducted. The abdominal wall was closed at last. Pathological analysis of the seven pancreatic sections (head and uncinate process) excised after surgery revealed nuclear hypertrophy of beta cells and hyperplasia of pancreatic islets. The following antibodies were found to be positive in immunohistochemistry: cytokeratins AE1/AE3, chromogranin A, synaptophysin, CD56, and insulin; glucagon and Ki-67 were found to be negative. The diagnosis of neoblastosis is supported by these results as well as the clinical information. The patient reported that the hypoglycemia episodes had stopped following the surgery. using insulin, dapaglifozin, and pancreatin, as well as a regulated diet.

DISCUSSION AND CONCLUSION

Nesidioblastosis is an uncommon condition that causes HPH in adults, with 0.5 to 5.0% of cases occurring in adults, according to published figures [13]. In 1938, the first incidence of nesidioblastosis was reported in children [14], while the first case in adults was reported in 1975 [15]. Nesidioblastosis in this age range is defined by a beta-cell dysfunction that can arise following Roux-en-Y gastric bypass surgery or as a symptom of noninsulinoma pancreatogenous hypoglycemia syndrome (NIPHS), both of which result in endogenous hypoglycemia. However, local or numerous insulinomas are the most frequent cause of this illness in adulthood [16].Postprandial hypoglycemia is the most common clinical characteristic. Postprandially, two to four hours after meals, and infrequently during fasting, symptoms appeared in a group of 18 Mayo Clinic patients, ages 16 to 78, with a BMI of 25.7 kg/m² and a 70% male preponderance [10,11]. On the other hand, fasting hypoglycemia is seen in the majority of insulinoma patients. The patient in the case study did not exhibit distinct hypoglycemia episodes; instead, they happened at random. Adult nesidioblastosis is difficult to diagnose preoperatively since there are no highly specific functional tests, clinical signs, or history to determine [18]. Since the patient in the instance had already been diagnosed with nesidioblastosis, functional testing had not been conducted and the patient's complaints were vague. In addition to guiding the identification of the disease and the direct excision of relevant pancreatic areas, a selective arterial calcium stimulation test can reveal hyperactive β-cell activity [19]. This analysis found that the areas irrigated by AMS and GDA, which correlate to irrigation of the head and uncinate process, produced more insulin.The exclusion of an insulinoma and a number of histological characteristics are part of the diagnostic criteria for adult nesidioblastosis put forth by Klöppel et al. [17]. Therefore, following a histological study, the final diagnosis of adult nesidioblastosis can be more precisely determined. The pathological examination of the excised portion demonstrated all of the criteria put forward by Kloppel et al. [17]. Additionally, immunohistochemistry helped to diagnose nesidioblastosis because it revealed diffuse neuroendocrine tissue.A reasonable first line of treatment for nesidioblastosis in patients with mild to severe symptoms is nutritional adjustment. nutritional adjustment, including cutting back on free carbohydrates and distributing them equally throughout the day. We recommend the alpha-glucosidase inhibitor Acarbose if mild to severe symptoms continue. Other drugs, including octreotides, verapamil, acarbose, and diazoxide, helped patients who had hypoglycemia following a Roux-en-Y bypass. But for severe and moderate cases that don't respond to alternative treatments, surgery is the only option. Surgery was decided since the patient in this case did not respond well to the first clinical treatment that was suggested. The majority of cases were resolved by partial or subtotal pancreatectomy, despite the lack of a conventional surgical method [10,11]. The degree of pancreatic resection necessitates attention to maintain enough β-cell volume to prevent diabetes because of these numerous procedures. According to the Frey procedure, which Frey et al. [20] proposed for the treatment of chronic pancreatitis, the reported patient's condition behaved similarly since he required two treatments, the final one being a subtotal pancreatectomy with duodenal preservation. Because a little portion of the patient's pancreas was still intact, this technique was successful in reducing symptoms

and improving glucose management. Therefore, it is clear that the surgical method (Frey's procedure) was effective in controlling the patient's blood sugar levels and did not result in any gastrointestinal system complications. Nevertheless, even though the pancreatic tissue was preserved, the case that was described evolved hyperglycemia following resections of pancreatic tissue.

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