OLGU SUNUMU / CASE REPORT

Solid pseudopapillary tumor and insulinoma in distal pancreatectomy

Distal pankreatektomide solid psödopapiller tümör ve insulinoma birlikteliği

Deniz Tuncel¹, Banu Yılmaz Özgüven¹, Ayşe Ayşim Özagarı¹, Muharrem Battal², Ahmet Uçar³, Elif Devecioğlu¹, Fevziye Kabukcuoğlu¹

¹Şişli Hamidiye Etfal Training and Research Hospital, Department of Pathology, ²Department of General Surgery, ³Department of Endocrinology, İstanbul, Turkey

Abstract

Multiple endocrine neoplasia type 1 (MEN 1) is a rare autosomal dominant inherited endocrine disease characterized by pancreatic, parathyroid, and anterior pituitary tumours. Pancreatic islet tumours occur less frequently, among them gastrinomas and insulinomas are the most prevalent. A solid pseudopapillary neoplasm (SPN) is another extremely rare tumour of the pancreas that frequently occurs in young females and is mostly benign. We report a case of a 16-year-old male MEN 1 syndrome with insulinoma and solid pseudopapillary neoplasm in distal pancreatectomy. The patient presented with seizures. When admitted to the emergency room, hypoglisemia has been detected due to the high levels of insulin. Multiple lesions have been observed in the corpus of the pancreas during the diagnostic work-up. According to the parathyroid scintigraphy, an adenoma has been detected. In the family, an asymptomatic parathyroid adenoma has also been seen in his brother. Distal pancreatectomy has been performed. In the gross examination, one neoplasm close to the proximal margin, one in the distal part of the pancreas have been found. The histomorphological and immunohistochemical evaluation revealed the proximal neoplasm as solid pseudopapillary neoplasm and the distal one as functioning neuroendocrine tumor (insulinoma).

Key words: Solid pseudopapillary neoplasm, insulinoma, MEN 1, pancreas.

INTRODUCTION

Solid pseudopapillary neoplasm of the pancreas is an uncommon tumor seen in approximately 1-2 % of the exocrine pancreatic tumors ¹ It has been described by Frantz as a papillary tumor of the pancreatic islet cell types and benign or malignant.² WHO declared the neoplasm as the solid pseudopapillary tumor in the classification of tumors of the exocrine pancreas.² The tumor mostly occurs in young female with a mean age of 27.2 ¹. It is commonly asymptomatic and incidentally found in 15% of the cases.³

Anahtar kelimeler: Solid psödopapiller tümör, insulinoma, MEN1, pancreas.
Pancreatic neuroendocrine tumors (PanNETs) are a rare group of neoplasms arising from the endocrine pancreas and incidence is increasing. They are associated with poor clinical outcomes, with a 10-year overall survival of 45%. PanNETs occur most frequently in adults between 40 and 60 years of age and equally seen in females and males.

PanNET and Solid Pseudopapillary Neoplasm appearing together is an enormously rare occasion. Thus the purpose of this article is to present the case with SPT and PanNET with the detailed clinical and pathological information.

CASE

A 16-year-old male patient who had MEN 1 syndrome with insulinoma and solid pseudopapillary neoplasm in distal pancreatectomy was presented. The patient was brought to hospital with seizures. When admitted to the emergency room, hypoglisemia has been detected due to the high levels of insulin. Multiple lesions have been observed in the corpus of the pancreas during the diagnostic work-up. According to the parathyroid scintigraphy, an adenoma has been detected. In the family, an asymptomatic parathyroid adenoma has also been seen in his brother. In the DNA sequence analysis heterozygous deletion pW183S has been detected. Same mutation has been identified both in his brother and his father. In the medical history, asymptomatic hiperparathyroidism has been observed in the patient’s brother. Distal pancreatectomy has been performed. In the histopathological and immunohistochemical evaluation, the proximal region mass was composed of small and medium size tumor cells, which had no obvious atypia. Pseudopapillary structures were found in most of the areas (Figure 1,2). The tumor was positive for vimentin, CD56, B-Catenin, Alpha-1 Antitrypsin, progesterone receptor, chromogranin, synaptophysin and negative for insulin (Figure 3-7), Ki-67 index was 6-7%. The distal region mass was composed of tumor cells possessing round or oval nuclei with “salt and pepper” chromatin and eosinophilic granular cytoplasm. The tumor nests are arranged in trabecular, insular, or sheet-like patterns. The tumor was positive for chromogranin, synaptophysin, PGP 9.5, insulin. Ki-67 index was < 1%. The tumors were diagnosed as neuroendocrine tumor Grade 1 (insulinoma) in the distal region and solid pseudopapillary tumor in the proximal pancreatic surgical in the specimen. In the follow-up period the patient is alive with no evidence of recurrence.

DISCUSSION

Solid-pseudopapillary neoplasm of the pancreas is a rare exocrine pancreatic tumor, which comprises only 1-2% of all tumors of the pancreas, first described by Frantz in 1959. Most of these tumors are found in young women in the second or third decade. Approximately 25% of these tumors may be seen in children. The most common clinical presentation is a palpable abdominal mass and abdominal pain. These tumors have a low malignant potential, and their prognosis is extremely good unlike other tumors of the pancreas.

Immunohistochemically, the SPPNs show positive reactivity for keratin, desmoplakin, trypsin, chymotrypsin, amylase and vimentin. In addition, focal positivity has been found for NSE and various islet cell hormones such as insulin and glucagons. This might suggest that SPPN of the pancreas arises from primitive pancreatic epithelial cells with a predominance of exocrine features but having capacity for dual (endocrine and exocrine) differentiation. The presence of progesterone receptors and its well-known predilection for females suggest that it is a hormone-dependent tumor. In our case, the patient was male, and showed progesterone receptor positivity and insulin negativity. Insulinoma is the most common cause of hyperinsulinemic hypoglycemia (1-4/million patients) and the second most common (10%–30%) functioning pancreatic islet cell tumor associated with multiple endocrine neoplasia type 1 (MEN1) after gastrinoma. In contrast, only 4%-6% of patients with insulinoma will develop MEN1. Unlike sporadic insulinomas that usually develop after the age of 40, MEN-associated insulinomas usually occur before 40 years of age and even sometimes before 20.

MEN is characterized by the occurrence of a tumor involving two or more endocrine glands within a single patient and has an equal sex distribution. MEN type 1 (MEN1) occurs in approximately one in 30,000 individuals. It encompasses tumors of the parathyroids (95% of cases), pancreatic islets (30%-80%), and anterior pituitary (15%-90%). We report a case of a 16-year-old male MEN 1 syndrome with
insulinoma and solid pseudopapillary neoplasm in distal pancreatectomy.

Figure 1. Solid Pseudopapillary Tumor, H&E, X40

Figure 2. Solid Pseudopapillary Tumor, Synaptophysin, X100

Figure 3. Solid Pseudopapillary Tumor, Beta Catenin, X200.

Figure 4. Solid Pseudopapillary Tumor, Progesteron, Reseptor, X200.

Figure 5. Solid Pseudopapillary Tumor, Insulin, X200.

Figure 6. Insulinoma, H&E, X100.

Figure 7. Insulinoma, Insulin, X200.

Figure 8. Insulinoma, Chromogranin, X100.

Figure 9. Insulinoma, Synaptophysin, X200.

Figure 10. Insulinoma, PGP 9.5, X200.

This case report was presented in the XXXI International Congress of the International Academy of Pathology and 28th Congress of the European Society of Pathology in Cologne in 2016.

REFERENCES


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