



OLGU SUNUMU / CASE REPORT

Solid pseudopapillary tumor and insulinoma in distal pancreatectomy

Distal pankreatektomide solid psödopapiller tümör ve insulinoma birlikteliği

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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, hypoglycemia 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.

Öz

Multipl Endokrin Neoplazi Tip 1 otozomal dominant geçişli nadir bir genetik hastalık olup pankreas, paratiroid ve anterior pitüiter tümörleri ile karakterizedir. Pankreasın adacık hücre tümörleri daha az sıklıkta görülmekte olup bu tümörler içerisinde gastrinoma ve insulinoma daha çok ortaya çıkan tümörlerdir. Solid Psödopapiller Tümör pankreasın diğer bir nadir tümörü olup erken yaşta kadın hastalarda ortaya çıkan sıklıkla benign bir neoplazidir. Distal pankreatektomili, MEN 1 sendromu olan 16 yaşında erkek hastada eş zamanlı görülen solid psödopapiller neoplazi ve insulinoma olgusunu sunuyoruz. Hastanın nöbet şikayetleri mevcut olup acil servise başvurduğunda yüksek insülin düzeyine bağlı olarak hipoglisemi saptandı. Tanısal çalışmalar sırasında pankreas korpusunda multipl lezyonlar saptandı. Ailede, erkek kardeşinde asemptomatik paratiroid adenomu görüldü. Hastaya distal pankreatektomi uygulandı. Makroskopik incelemede bir neoplazi proksimal cerrahi sınıra yakın diğeri distal pankreas yerleşimli iki adet neoplazi bulundu. Histomorfolojik ve immünohistokimyasal değerlendirmede proksimaldeki lezyonun solid psödopapiller neoplazi, distaldeki lezyonun fonksiyonel nöroendokrin tümör (insulinoma) olduğu dikkati çekti.

Anahtar kelimeler: Solid psödopapiller tümör, insulinoma, MEN1, 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

pancreas, benign or malignant". 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.

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Pancreatic neuroendocrine tumors (PanNETs) are a rare group of neoplasms arising from the endocrine pancreas and incidence is increasing⁴. They are associated with poor clinical outcomes, with a 10-year overall survival of 45%^{5,6}. PanNETs occur most frequently in adults between 40 and 60 years of age and equally seen in females and males⁷.

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, hypoglycemia 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^{8,9}. Most of these tumors are found in young women in the second or third decade¹. Approximately 25% of these tumors may be seen in children³. The most common clinical presentation is a palpable abdominal mass and abdominal pain¹⁰. These tumors have a low malignant potential, and their prognosis is extremely good unlike other tumors of the pancreas¹¹.

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 tumor¹². In our case, the patient was male, and showed progesterone reseptor 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)¹³ after gastrinoma. In contrast, only 4%-6% of patients with insulin- oma will develop MEN1¹⁴⁻¹⁶. 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^{17,18}.

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^{19,20}. 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.

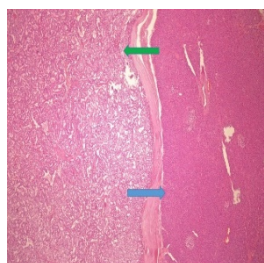


Figure 1. Solid Pseudopapillary Tumor, H&E, X40

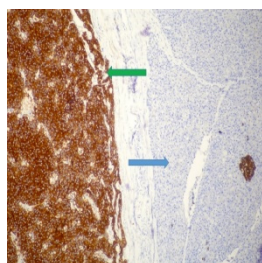


Figure 2. Solid Pseudopapillary Tumor, Synaptophysin, X100

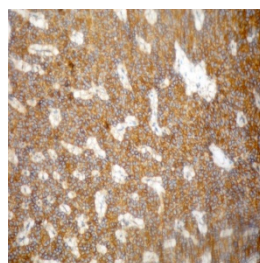


Figure 3. Solid Pseudopapillary Tumor, Beta Catenin, X200.

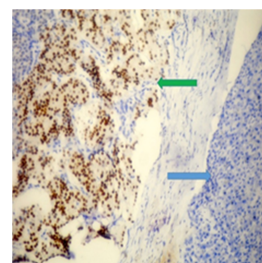


Figure 4. Solid Pseudopapillary Tumor, Progesteron, Reseptor, X200.

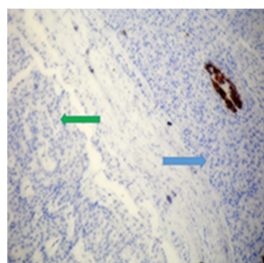


Figure 5. Solid Pseudopapillary Tumor, Insulin, X200.

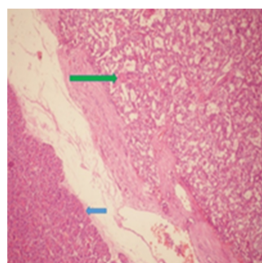


Figure 6. Insulinoma, H&E, X100.

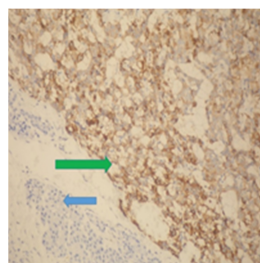


Figure 7. Insulinoma, Insulin, X200.

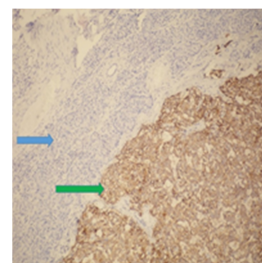


Figure 8. Insulinoma, Chromogranin, X100.

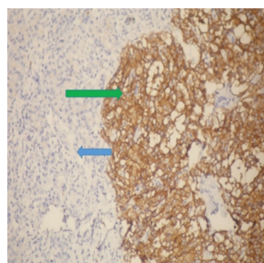


Figure 9. Insulinoma, Synaptophysin, X200.

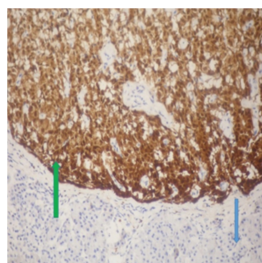


Figure 10. Insulinoma, PGP 9.5, X200.

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