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CASE REPORT

# Metastases of pancreatic neuroendocrine tumor to the liver as extremely rare indication for liver transplantation in children. Case report and review of the literature

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**Summary** Neuroendocrine tumors (NET) are extremely rare in children (0.75 cases per 100,000 children and adolescents a year) and the majority of these tumors are benign or present low grade of malignancy. According to the American registry Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute, less than 2% of all neuroendocrine tumors in children occur in the pancreas, making it a rare site for these tumors. The majority of them are found in children over 10 years of age, especially those with malignant potential. Treatment of NET consists of different methods: surgery, somatostatin analogues and chemotherapy. Radical surgical resection remains the standard of treatment; however, it is not always

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feasible because of distant metastases. The authors present a case report of pancreatic NET with multiple metastases to the liver. The patient was treated with pancreatic resection and liver transplantation for liver metastases. Prior to liver transplantation, the patient was treated with somatostatin analogues, sunitinib and chemotherapy. Management of liver metastases with liver transplantation is discussed.


## Introduction

Neuroendocrine tumors are extremely rare in children (0.75 cases per 100,000 children and adolescents a year). The majority of these tumors are benign or of low grade malignancy. Highly malignant NET represent up about 10% of all neuroendocrine tumors in children [1]. According to American registry Surveillance, Epidemiology, and End Results Program of the National Cancer Institute (SEER) less than 2% of all neuroendocrine tumors in children are located in the pancreas [2]. Majority of them occur in children over 10 years of age (mean age at presentation is about 15 years). NET management consists of surgery, administration of somatostatin analogues, chemotherapy, however radical surgical resection remains the key for cure.

More than 60% of patients with malignant abdominal NET tumors (GEP-NET-gastro-entero-pancreatic NET) present with liver metastases at diagnosis [1,2]. Surgical resection remains the gold standard in the treatment of liver metastases.

Local ablative techniques are used in some patients for symptoms and tumor growth control. Liver transplantation is regarded as suitable only in selected group of patients. This may include widespread liver disease without extrahepatic extent, young patients with pain or hormonal symptoms refractory to surgical and systemic therapy [3].

Patients with complete resection of primary pancreatic tumor and multiple or recurrent hepatic metastases may be considered as candidates for liver transplantation. However, according to liver transplantation registry, only about 0.4% of all liver transplants were performed due to metastatic endocrine tumors [4]. We would like to add another interesting case to these reports.

## Case report

A 13-year-old female was referred to our institution with symptoms of repeated hypoglycemic episodes (35—43 mg%) and symptoms of apathy, sweating, headaches occurring since 3 months, particularly after physical efforts. No loss of consciousness and seizures were reported. After the second such episode hypoglycemia was discovered. Fasting test showed hypoglycemia of 40 mg% after 4 h, with increased insulin concentration —86.28 IU/ml ($N < 16$ IU/ml). Ketone metabolites in the urine were not found. Abdominal ultrasound and magnetic resonance imaging (MRI) revealed 15 mm polycystic lesion in the head of pancreas, and 2 smaller nodules localized between pancreas and large vessels, corresponding to lymph nodes. Liver parenchyma was normal. She was treated with Diazoxide in order to prevent episodes of hypoglycemia.

Computer tomography (CT) scan performed two months later showed tumor within pancreatic head, $27 \times 36 \times 43$ mm in size, enlarged lymph nodes in the hepatic hilum, and normal liver. PET CT showed increase of $^{18}$F DOPA metabolism in the pancreatic head, but not within the liver (Fig. 1 A, B). MRI of the brain was not significant.

The patient underwent modified Whipple procedure with resection of pancreatic head and part of the body en block with enlarged lymph nodes and duodenum. A fragment of pancreas $7 \times 4 \times 2$ cm contained firm, solid, white tumor, $3 \times 3 \times 1.5$ cm in size.

Intraoperatively, enlarged lymph nodes in the pancreas proximity and multiple metastases (> 20) in the liver were detected. There was no need to perform ultrasound intraoperatively because metastases were seen well macroscopically.

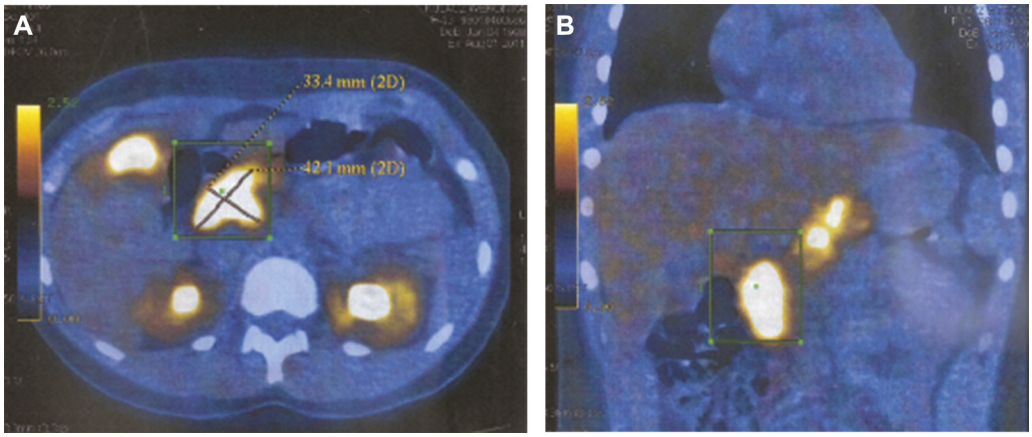


**Figure 1** A, B: positron emission tomography (PET) of pancreatic neuroendocrine tumor before Whipple procedure.