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Neuroma (schwannoma). A rare pancreatic tumor

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ABSTRACT:

Introduction: Neuroma (Schwannoma in Latin) is an encapsulated, mesenchymal tumor arising from Schwann cells surrounded by nerves. Hence it can be located in any area in the body with passing peripheral nerves. The most common location is the head, neck, and extremities. The tumor arising from Schwannoma cells was first described by Stout and Carson in 1935. Pancreatic schwannomas are extremely rare tumors. Until 2017, in English literature 68 cases have been described. Surgical treatment is the most common way of treating pancreatic schwannomas, and postoperative prognoses are good.

Case report: A 63-year-old patient was admitted to the Clinical Department of Gastroenterological Surgery and Transplantation of the Central Clinical Hospital at the Ministry of Interior and Administration in Warsaw due to pancreatic head cancer. Needle biopsy–both ultrasound-guided and CT-guided as well as open biopsy for lesions in the pancreas did not show tumor cells in any of the collected samples. Abdominal CT in a projection of the uncinate process of the pancreas revealed an oval lesion highly suspected of neoplastic process. Next, diagnostics was extended by abdominal MRI which revealed a retroperitoneal tumorous thick-walled cystic mass filled with fluid. The patient was qualified for surgical treatment. Pancreaticoduodenectomy (Whipple Procedure) was done on August 22, 2017. Material sent for histopathological examination revealed Schwannoma capitis pancreatis. In surgical practice, pancreatic schwannoma occurs extremely rare, but in centers which conduct large numbers of surgical procedures in the pancreas, a case like this may occur.

KEYWORDS: pancreas, pancreatoduodenectomy, schwannoma, tumor of pancreas

INTRODUCTION

Neuroma (Schwannoma in Latin) is an encapsulated, mesenchymal tumor arising from Schwann cells surrounded by nerves. Hence, it can be located in any area in the body with passing peripheral nerves. The most common location is the head, neck, and extremities. The tumor arising from Schwannoma cells was first described by Stout and Carson in 1935 [15]. The texture if the tumor is made up of spindle cells in a vorticose arrangement palisadic nuclei forming Verocay cells (Antoni A) or a loose system of star-shaped cells in a loose, mucosal stroma (Antoni B). In general, it is a benign tumor and surgical treatment is definitive, while in rare cases it develops into a malignant form, i.e., sarcoma neurogenes. The malignant form of neuroma coexists with von Recklinghausen's disease in 5% of cases. In mild cases, monosomy 22 or 22q deletion occurs [13, 14].

Pancreatic schwannomas are extremely rare tumors. For the last 40 years, English literature has described 68 cases of such cases. [14] The ratio of men to women is 1:1.26, mean age is 55.7 years, while the average tumor size is 6.1 cm. About ⅓ of the patients did not report any complaints, the remaining patients experienced abdominal pain, weight loss, back pain, nausea, and vomiting.

Forty percent of the tumors were located in the head of the pancreas, 4.62% on the borderline of the head and body, 23.08% in the body, 10.78% on the borderline of the body and tail, 10.78% in the tail, 10.78% in the uncinate process of the pancreas. Surgical treatment is the most common treatment for pancreatic neuromas and postoperative prognosis is good [1–3, 7–9, 11, 12, 14]. The most frequently performed procedure was pancreatoduodenectomy (34%), distal resection of the pancreas was performed in 22%, and tumor enucleation in 12% of patients [14].

CASE REPORT

A 63-year-old patient was admitted to the Clinical Department of Gastroenterological Surgery and Transplantation of the Central Clinical Hospital at the Ministry of Interior and Administration in Warsaw due to pancreatic head cancer. The interview included the patient's condition after past cholecystectomy performed 5 years ago. Based on the available documentation of the patient's treatment in other centers, it was revealed that in 2014 she had underwent a needle biopsy – both ultrasound- and CT-guided and open biopsy of lesions in the pancreas, the collected materials did not reveal any tumor cells. Abdominal and pelvic CT scan with contrast performed on June 20, 2017 visualized an oval lesion of the uncinate process of the pancreas with dimensions of 71x45x56mm with 12mm-thick parietal tissue areas, central hypodensity (possibility of liquid or decay) – image highly suspected of tumorous process. Next, the diagnosis was extended to include abdominal MR performed on July 5, 2017, in which a tumorous thick-walled cystic mass filled with fluid was found. According to the description, the lesion most likely arose from the uncinate process of the pancreas and had a size of 61x50x57mm (in the MR study of June 9, 2014. 49x39x41mm). A suspicion of cystadenoma mucinosum (MCN) arose as the conclusion of the study, the lesion had increased in comparison to the study from 2014 [Fig. 1.]. The patient was qualified for surgical treatment. The surgery was performed on August 22, 2017. A cystic tumor was found in the head of the pancreas intraoperatively. It was decided to excise the head of the pancreas with the duodenum. The Whipple procedure (pancreaticoduodenectomy) was performed. Perioperative and postoperative periods were uneventful. The patient was discharged home on the 8th day after surgery, in good general condition. In the follow up study done 3 months after the surgery, no signs of tumor recurrence were found. The material sent for histopathological ex-

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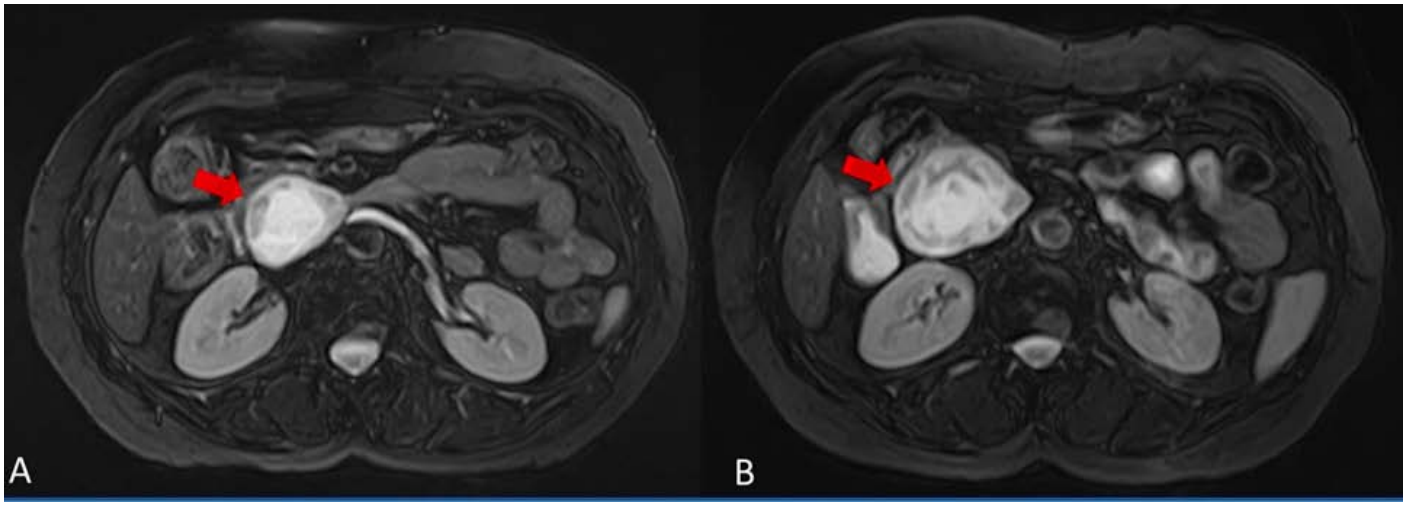

Fig. 1. The results of imaging examinations performed before the operation.
A – 2014.06.09.MR AX T2 FSAT; B – 2017.07.05. MR AX T2 FSAT

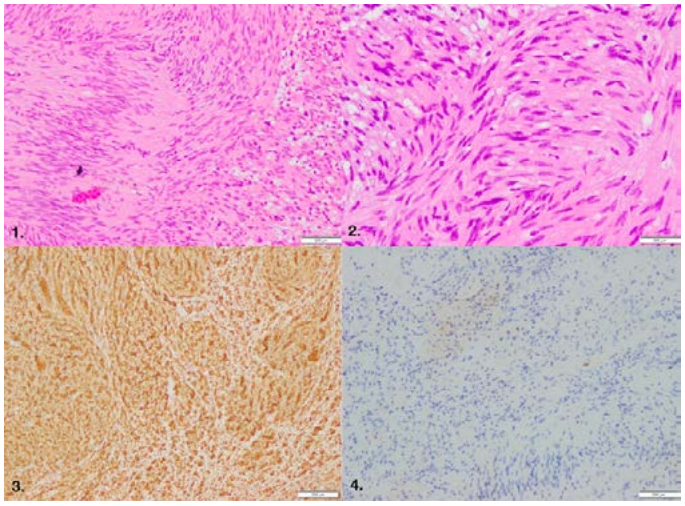

Fig. 2. The results of imaging examinations performed before the operation.
1. HE stain (x20). 2. HE stain (x40). 3. S100(+) positive reaction in tumor cells.
4. CD117(-) exclusion of GIST tumor.

amination revealed Schwannoma cells, the resection margin was R0, while none of the nine lymph nodes contained in the preparation revealed metastatic lesions. A solid, well-demarcated polycyclic expanding tumor with dimensions of 60x47x56mm delineated from the pancreatic parenchyma was described macroscopically. The remaining pancreatic parenchyma without focal lesions. Immunohistochemical study revealed S100 (+), CD34 (-), desmin (-), CD117 (-) [Fig. 2.]. The remaining pancreatic parenchyma showed PanIN-1, PanIN-2 type intraductal hyperplasia.

DISCUSSION

In the described case, over three years of tumor observation could suggest its benign character. The MR study aroused a suspicion of MCN, confirmed by the lack of pathognomonic sings of neuroma in imaging studies. In the available literature, it was often stated that diagnostic imaging such as ultrasound, CT, MRI and visualize neuroblastomas as cystic tumors, solid tumors, mucinous tumors or pseudocysts. The more and more frequently performed FDG-PET/CT examinations reveal marker cumulations, however as far as 68 cases described in the English-language literature are concerned, only 3 patients completed the study [1]. Histopathological examination remains a study that can determined the nature of tumor. In the immunohistochemical study, Schwannoma is confirmed in the case of positive protein S100, Vimentin, and CD56. Schwannoma occasionally demonstrates expression in CD57 and GFAP, while expression remains negative in cytokeratin AE1/3, CD34, c-kit, desmin, and myosin [14]. Therefore, EUS examination with biopsy may prove very useful in pre-operative diagnosis. In surgical treatment of neuroma, consideration may be given to performing tumor enucleation as a less aggravating procedure for the patient, but the risk of malignant transformations of lesions should be taken into account in a small percentage of cases [1, 3, 4, 5]. Until 2010, of 47 cases of pancreatic neuromas, only 5 cases were described as malignant [10]. Three tumors were located in the head of the pancreas, two in the distal part of the pancreas, one of which infiltrated the colon [10]. In the last case, en bloc resection was applied. There was no recurrence in 28-month follow-up. Radiotherapy was used as adjuvant therapy in pancreatic neuromas, but chemotherapy was not applied in the perioperative period [6]. It is believed that with a tumor size over 10 cm, particular attention should be paid to the potential malignant character of the tumor [10]. Schwannomas are tumors which occur individually. To date, only one case of three synchronously occurring tumors with neuroma morphology has been described – in the liver, porta hepatis, and gall bladder [7].

According to the latest National Comprehensive Cancer Network (NCCN) guidelines of 2017 for pancreatic cancer, every focal lesion in the pancreas should be treated as potentially cancerous. To date, we have performed 1,627 pancreatic resections at the Clinical Department of Gastroenterological Surgery and Transplantation of the Central Clinical Hospital at the Ministry of Interior and Administration in Warsaw specializing in pancreatic surgery. The material collected during surgeries was dominated by ductal carcinomas, IPMN, MCN, SCN, less frequently neuroendocrine tumors and metastatic tumors, whereas the described case is the first Schwannoma lesion. In surgical practice, pancreatic neuroma (schwannoma) is extremely rare, but in centers performing large numbers of pancreatic surgeries, the possibility of this cancer should be taken into account.

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