Original Article

Pancreatic head tumor revealing as a Schwannoma: A case report

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ABSTRACT

Pancreatic schwannomas are rare neoplasms with an equal gender distribution. These tumors commonly vary in size and undergo degenerative changes; moreover, about two thirds of them are partially cystic. Pancreatic schwannoma tumors may occur at any age with a slightly higher incidence in females than in males. In general, malignancy is observed in few cases of pancreatic schwannoma. These tumors are reported to locate in the head of the pancreas in most of the patients. However, they have been also observed in the head, body, and tail of the pancreas in some cases. All these issues have made the clinical diagnosis of pancreatic schwannoma confusing and difficult. The initial evaluation of choice for pancreatic schwannomas is computed tomography, which usually shows a well-defined, round mass with multiple, low-attenuation, and cystic necrotic areas. Pancreaticoduodenectomy and distal pancreatectomy are the best selective treatments for these tumors. Herein, we presented a 64-year-old female with pancreatic schwannoma referring with a long-term history of abdominal pain.

Keywords: Pancreatic Schwannomas, neurofibroma, immunohistochemical, pancreatectomy.

Introduction

Schwannomas, also known as neurilemmoma or neurofibroma, are benign neoplasms, derived from the Schwann cells, which line the nerve sheath and commonly found in the extremities and soft tissues of the head and neck. They can also appear in the trunk, retroperitoneum, mediastinum, pelvis, and even rectum [1, 2].

Pancreatic schwannomas are rare tumors with an equal gender distribution [3]. They mostly presents in form of cystic, thin-walled, and hemorrhagic masses [4]. So far, more than 50 cases with pancreatic schwannomas have been reported in English literature. Pancreatic schwannoma has been reported in patients at different age groups, and they have a slightly higher incidence in females than in males [1]. The majority of pancreatic schwannoma tumors are large in diameter; accordingly, there are few reports on small solid pancreatic schwannomas [5-7].

These neoplasms exist in remarkably different sizes and commonly undergo degenerative changes. Cyst formation, calcification, hemorrhage, hyalinization, and xanthomatous infiltration are reported in the majority of cases with pancreatic schwannomas [3]. These comorbidities may result in the inaccurate diagnosis of pancreatic schwannomas because these tumors may mimic cystic pancreatic lesions in radiographic findings.

In this report, we presented a case of pancreatic schwannoma with attention to clinical presentation, diagnosis, treatment options, and outcomes.

Case report

A 64-year-old female referred with the complaints of night fever and long-term history of abdominal pain, especially when coughing. She had been prescribed oral omeprazole for 4 months. The patient's symptoms intensified, and she reported a lot of pain in the gastric area when eating, which spread to the back and felt a mass located 3 cm above the umbilicus. The

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patient was subjected to endoscopy three years ago, which revealed gastroesophageal reflux disease and gastritis dilating the second part of the duodenum.

She had a weight loss of 8 kg in the past 3 months; however, she showed no such symptoms as nausea, vomiting, itching, or yellow skin. She had a past surgical history of cataract. There was no history of taking medicine or smoking. Furthermore, she had no family history of malignancy or allergy.

Regarding the vital signs, she had the blood pressure of 120/80 mmHg, heart rate of 78 beats per min, and respiratory rate of 16 breaths per min. There was no sign of scleral icterus. Supraclavicular lymphadenopathy was not observed in the anterior and posterior cervical. In urine test, bilirubin and ketone were negative, and urine glucose was obtained as +1. Table 1 summarizes the results of other laboratory tests.

The results of the chest exam were normal, and the abdomen was soft. A mild tenderness was reported in the area between the epigastrium and umbilicus. A mass was detected in the epigastric region through abdominal palpation.

The patient had no anemia or leukocytosis, and hepatic enzymes were normal.

The tumor markers were within the normal range. Amylase and cancer antigen levels were 70 U/L and 9-19 U/mL, respectively. Furthermore, the levels of carcinoembryonic antigen were obtained as 1.98 and 0.88 ng/ml in the first and second tests, respectively. The possible diagnoses suggested for the patients included serous/mucinous cystic neoplasm, pseudopapillary neoplasm, or other cysts.

Ultrasound showed a solid cystic tumor with a diameter of 126×88 mm encapsulated in a well-defined hard wall of high density was found near the pancreatic head at the retroperitoneum and Gall stone in Gall bladder. CT scan was done and revealed a solid-cystic tumor measuring 82×86 mm with calcified walls in the vicinity of the pancreatic head at the retroperitoneum (Figure 1). Moreover, several lymph nodes, 9 mm in diameter, were found between tumor, inferior vena cava, and third part of the duodenum.

Based on magnetic resonance imaging, a solid mass with a diameter of 113×89×94 mm was detected in the pancreatic head. Upper endoscopy led to the observation of a solid cystic mass in the pancreatic head without vascular invasion.

Endosonography was done and reported a large 98*80 mm well demarcated multicystic lesion with solid components adjacent to pancreatic head with no invasion to vessels, fine needle aspiration was performed and the pathology report was non diagnostic specimen.

The patient was became candidate of surgery with the diagnosis of pancreatic head mass. Prior to the surgery, informed consent was obtained from the patient and her companion. The mass was resected then through pancreaticodeudnectomy procedure. (Figure 2)

The histopathological examination of the resected specimen revealed chronic cholecystitis with cholelithiasis and Schwanoma of pancreatic head mass. Pnacreatic head mass specimen was evaluated for immunohistochemistry and was positive for S100 and CK and weakly positive for Desmin and also negative for CD117, SMA, CD34 and Calretinin.

Finally, the patient was discharged in a good general condition and she could have oral feeding and had normal defecation. She received regular follow-up care and was normal 6 month after surgery.

Discussion

Our case was detected with a solid relatively large schwannoma with a well-defined capsule. The majority of pancreatic schwannoma tumors are large in diameter [5-7]. In a systematic review conducted by Moriya et al., the pancreatic schwannoma size was reported to range within 1-20 cm with a mean of 6.2 cm. However, they found no correlation between symptoms and tumor size or location. In the mentioned study, the majority of the patients with pancreatic schwannoma exhibited cystic tumors, while solid tumors were observed in 34% of the cases. Furthermore, the pancreatic schwannoma was reported to locate in the head of the pancreas in most of the patients (40%); however, this tumor was observed in the head, body, and tail of the pancreas in some cases [1]. In our study, the size of tumor was 126 mm, which is large in comparison to other reported cases of pancreatic schwannoma. Furthermore, in our patient, the tumor was located in the vicinity of the pancreatic head at retroperitoneum.

Pancreatic schwannoma has been reported in patients with the age of 20-87 years; therefore, it may occur at any age [1]. The incidence of pancreatic schwannoma is slightly higher in females than in males (ratio of 26:21). In general, malignancy is observed in few cases of pancreatic schwannoma (less than 10%); furthermore, no mortality has been reported due to this disease [1, 4, 8]. These lesions usually occur as benign and solitary; nonetheless, they may be multiple and malignant when accompanied with von Recklinghausen's disease [4, 9, 10].

Pancreatic schwannoma is typically asymptomatic for a long time and may be clinically noticed with the compression of the surrounding organs in the CT scan findings [4]. The most common symptoms of pancreatic schwannoma are abdominal pain, weight loss, back pain, nausea/vomiting, abdominal mass, melena, and jaundice [1]. However, this tumor may remain asymptomatic in some cases and be only detectable on CT scans.

In the current study, we reported on an old female with benign pancreatic schwannoma and a long-term history of abdominal pain and night fever. In general, schwannoma is known as a benign spindle cell tumor that courses through the pancreas and arises from either autonomic sympathetic or parasympathetic fibers [2]. Secondary degenerative changes (e.g., cyst formation, calcification, hemorrhage, and hyalinization) are more observed in the deep tumors, which grow larger [11].

Histopathologically, schwannoma is composed of Antoni A and Antoni B areas, which consists of densely packed, spindle cells arranged in palisades. However, the other areas have myxoid structures, which are hypocellular and occupied by loosely

arranged tumor cells [1]. Both mentioned areas are observable in the majority of pancreatic schwannoma in different proportions. Degenerative or cystic changes caused by vascular thrombosis and subsequent necrosis occur in the Antoni B area (e.g., calcification or hemorrhage). These degenerative changes may lead to the diagnosis of cystic pancreatic lesions instead of pancreatic schwannoma [12]. Regarding this, the correct diagnosis of pancreatic schwannoma, especially in its cystic type, is very difficult.

These tumors grow slowly and are remarkably different in size. Moreover, about two thirds of them are partially cystic. All these can cause clinical confusion in the diagnosis of pancreatic schwannoma [13]. The initial evaluation of choice for pancreatic schwannomas is CT scan, which usually demonstrates well-defined, round masses with multiple, low-attenuation, and cystic necrotic areas.

In pancreatic schwannoma, the difference between the well-enhanced areas (i.e., Antoni A region) and unenhanced areas (i.e., Antoni B areas) is found in the CT findings. Pathological features are reported to correlate with CT scan findings in this tumor [14]. The presence of low-density region and/or cystic images reflecting the Antoni B component is the most specific feature of pancreatic schwannoma [14]. This tumor in magnetic resonance imaging presents as a hypointensity on T1-weighted images and a hyperintensity on T2-weighted images [15]. However, these imaging characteristics are similar to those observed in the radiological findings of other pancreatic tumors. Therefore, it is of fundamental importance to consider the differential diagnoses of pancreatic schwannomas.

However, the histopathological examination of the lesion with Antoni A and Antoni B areas and immunostaining analysis are necessary for the definitive diagnosis of schwannoma. Anti-S100 protein is the antibody mostly used to identify schwannomas. Fusiform Schwann cells are applied to identify the neural origin of different lesions through showing protein S-100 and nuclear export signal [16]. In addition, schwannoma stains are positive for S vimentin and CD [1].

The use of ultrasound-guided fine needle aspiration biopsy and immunohistochemical staining is a common practice for a more accurate diagnosis of pancreatic schwannoma [17, 18]. However, the diagnostic accuracy of this method is only one in eight histologically proven schwannomas [19, 20]. Consequently, histological examination and complex immunohistochemistry are needed for making a definitive diagnosis [21].

The simple enucleation of pancreatic schwannoma is sufficient because it has a low probability of malignant transformation. Pancreaticoduodenectomy and distal pancreatectomy are the best selective treatments for pancreatic schwannoma. Combined transverse colon resection is another therapeutic option for pancreatic schwannoma. Despite the lack of studies investigating chemoradiation therapy in the management of neurofibroma, radiotherapy has been suggested to decrease tumor growth in neurogenic schwannoma [22]. Given the lack of enough evidence on the efficacy of chemoradiation therapy in the management of pancreatic schwannoma, surgical excision remains the most important treatment, resulting in a low risk of recurrence when aimed at a complete removal [16, 23].

In conclusion, the preoperative diagnosis of pancreatic schwannomas is very difficult. Although pancreatic schwannoma is very rare, it is an important pathological condition. These lesions should be detected with regard to the differential diagnosis of cystic neoplasms of the pancreas.

References

1. Moriya T, Kimura W, Hirai I, Takeshita A, Tezuka K, Watanabe T, et al. Pancreatic schwannoma: case report and an updated 30-year review of the literature yielding 47 cases. World Journal of Gastroenterology: WJG. 2012; 18(13):1538.
2. Paranjape C, Johnson SR, Khwaja K, Goldman H, Kruskal JB, Hanto DW. Clinical characteristics, treatment, and outcome of pancreatic Schwannomas. Journal of Gastrointestinal Surgery. 2004; 8(6):706-12.
3. Almo KM, Traverso LW. Pancreatic schwannoma: an uncommon but important entity. Journal of gastrointestinal surgery. 2001; 5(4):359-63.
4. Stojanovic MP, Radojkovic M, Jeremic LM, Zlatic AV, Stanojevic GZ, Jovanovic MA, et al. Malignant schwannoma of the pancreas involving transversal colon treated with en-bloc resection. World Journal of Gastroenterology: WJG. 2010; 16(1):119.
5. Soumaoro LT, Teramoto K, Kawamura T, Nakamura N, Sanada T, Sugihara K, et al. Benign schwannoma of the pancreas. Journal of gastrointestinal surgery. 2005; 9(2):288-90.
6. Sugiyama M, Kimura W, Kuroda A, Muto T. Schwannoma arising from peripancreatic nerve plexus. AJR American journal of roentgenology. 1995; 165(1):232-.
7. Feldman L, Philpotts LE, Reinhold C, Duguid WP, Rosenberg L. Pancreatic schwannoma: report of two cases and review of the literature. Pancreas. 1997; 15(1):99-105.
8. Móller VP, Hede A, Graem N. A solitary malignant schwannoma mimicking a pancreatic pseudocyst. A case report. Acta Chirurgica Scandinavica. 1982; 148(8):697-8.
9. Goldblum JR, Weiss SW, Folpe AL. Enzinger and Weiss's Soft Tissue Tumors E-Book: Elsevier Health Sciences; 2013.
10. Walsh MM, Brandspigel K. Gastrointestinal bleeding due to pancreatic schwannoma complicating von Recklinghausen's disease. Gastroenterology. 1989; 97(6):1550-1.
11. Akiyoshi T, Ueda Y, Yanai K, Yamaguchi H, Kawamoto M, Toyoda K, et al. Melanotic schwannoma of the pancreas: report of a case. Surgery today. 2004; 34(6):550-3.
12. Weiss S, Langloss J, Enzinger F. Value of S-100 protein in the diagnosis of soft tissue tumors with particular

reference to benign and malignant Schwann cell tumors. Laboratory investigation; a journal of technical methods and pathology. 1983; 49(3):299-308.

13. Kinhal VA, Ravishankar T, Melapure AI, Jayaprakasha G, Gowda BR. Pancreatic schwannoma: Report of a case and review of literature. Indian Journal of Surgery. 2010; 72(1):296-8.
14. Suzuki S, Kaji S, Koike N, Harada N, Hayashi T, Suzuki M, et al. Pancreatic schwannoma: a case report and literature Review with special reference to imaging features. JOP Journal of the Pancreas. 2016; 11(1):31-5.
15. Novellas S, Chevallier P, Saint Paul M, Gugenheim J, Bruneton J. MRI features of a pancreatic schwannoma. Clinical imaging. 2005; 29(6):434-6.
16. Martins MD, De Jesus LA, Fernandes KPS, Bussadori SK, Taghloubi SA, Martins MAT. Intra-oral schwannoma: case report and literature review. Indian Journal of Dental Research. 2009;20(1):121.
17. Li S, Ai SZ, Owens C, Kulesza P. Intrapancreatic schwannoma diagnosed by endoscopic ultrasound-guided fine-needle aspiration cytology. Diagnostic cytopathology. 2009; 37(2):132-5.
18. Hirabayashi K, Yasuda M, Umemura S, Itoh H, Itoh J, Yazawa N, et al. Cytological features of the cystic fluid of pancreatic schwannoma with cystic degeneration. A case report. Jop. 2008; 9(2):203-8.
19. Yu G, Sack M, Baloch Z, Gupta P. Difficulties in the fine needle aspiration (FNA) diagnosis of schwannoma. Cytopathology. 1999; 10(3):186-94.
20. Tafe LJ, Suriawinata AA. Cystic pancreatic schwannoma in a 46-year-old man. Annals of diagnostic pathology. 2008; 12(4):296-300.
21. Eggermont A, Vuzevski V, Huisman M, De Jong K, Jeekel J. Solitary malignant schwannoma of the pancreas: report of a case and ultrastructural examination. Journal of surgical oncology. 1987; 36(1):21-5.
22. Sawamura Y, Shirato H, Sakamoto T, Aoyama H, Suzuki K, Onimaru R, et al. Management of vestibular schwannoma by fractionated stereotactic radiotherapy and associated cerebrospinal fluid malabsorption. Journal of neurosurgery. 2003; 99(4):685-92.
23. Bui TD, Nguyen T, Huerta S, Gu M, Hsiang D. Pancreatic schwannoma. A case report and review of the literature. Jop. 2004; 5(6):520-6.

Table 1: Results of laboratory test and routine hematology

Test	Before resection		After resection	
Blood sugar (mg/dL)	82	Normal	152	High
BUN (mg/dL)	25	High	12	Normal
Creatinine (mg/dL)	1.1	Normal	1.1	Normal
SGOT (IU/L)	14	Normal	88	High
SGPT (IU/L)	14	Normal	78	High
ALK.P (IU/L)	216	Normal	199	Normal
Total Bilirubin	0.3	Normal	1.2	Normal
Albumine	--	--	3.0	Low
Serum Na	144	Normal	142	Normal
Serum K	4.3	Normal	4.5	Normal
Serum Mg	--	--	1.7	low
W.B.C (x1000/mm^3)	3.9	Low	--	--
R.B.C(million/mm^3)	3.83	Low	--	--
Hb (g/dL)	11.8	Normal	--	--
Hct	34.6	Low	--	--
M.C.V (fL)	90.3	Normal	--	--
M.C.H (Pgm)	30.8	Normal	--	--
M.C.H.C	34.1	Normal	--	--
Platelet (x1000/mm^3)	175	Normal	--	--

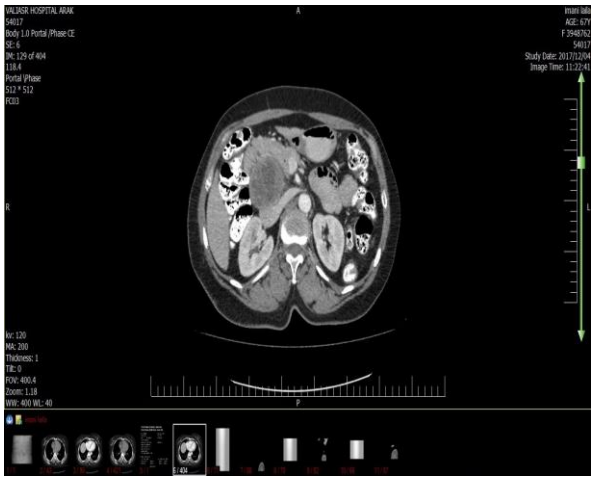

Figure 1: Preoperative CT scan of abdomen showing large cystic mass in head of pancreas.

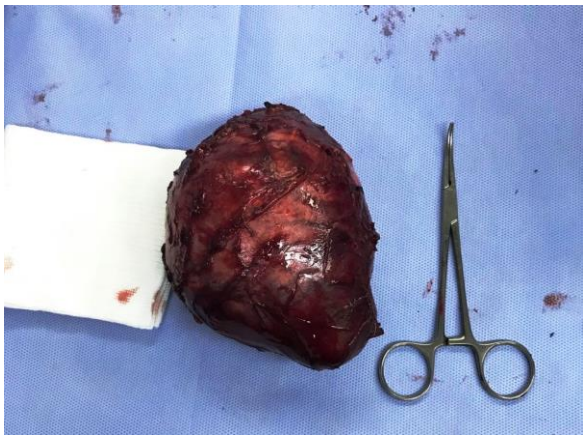

Figure 2: Tumor gross anatomy