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Case Report

Surgically curable Pancreas Enigma: Solid Pseudopapillary Tumor. Report of a case and literature review

Abstract

Solid pseudopapillary tumor (SPT) is a rare pancreatic neoplasm, classified as low-grade malignant tumor by the World Health Organization in 2000. Surgical treatment usually provide curative intent. Here in we report a 30 years-old woman whom had been operated upon possible benign pancreatic head mass diagnosed histopathologically as solid pseudopapillary tumor. And we aimed to review the literature on the basis of the case. In conclusion, solid pseudopapillary tumors are slow-growing tumors, with unpredictable potential for malignancy. Aggressive surgery yields favorable results even in the presence of vascular invasion and metastatic disease.

Introduction

Solid pseudopapillary tumor (SPT) is a rare neoplasm of the exocrine pancreas. The incidence of this tumor is rises from 0.5% to 2.7% of all pancreatic tumors in recent years depend on the recognition of the clinicians and the development of radiologic modalities [1,2]. It was first described by Frantz in 1959 and it is also known as Frantz's tumor [3,4]. It was renamed as SPTs and categorised under exocrine pancreatic tumors by the World Health Organization in 2000 [5]. It predominantly affects young women and children [3,4]. The tumor may progress to very large dimension (30 cm) without any metastasis or ascites. Cure had been reported even in the basis of very large dimention or metastasis [3,4,6-9]. As well, liver transplantation for liver metastasis of SPT had been reported previously [10].

Case

Patient: Thirty years-old woman admitted to our clinic after the diagnosis of pancreatic head mass by computed tomography (CT) in another hospital. When she came to our clinic her biochemical tests, tumor markers CEA, CA 19-9 and neuron specific enolase (NSE) levels are all in normal range and hypervascular, cystic and solitary well-capsulated roundy 4 cm pancreatic head mass was seen in the contrast enhanced abdominal CT. She was very young, she had no malignancy risk, the mass is wellshaped (not seem infiltratively and invaded any structure) with cystic parts. So the diagnosis is the possible benign or premalignant behavior pancreatic mass. Solid pseudopapillary tumor or neuroendocrine tumor are the suspicious diagnosis. Findings are the similar in magnetic resonance ımaging (MRI). When CT (Figure 1) and MRI (Figure 2) evaluated upon surgical decision there is no neighbor organ infiltration or vasculary invasion, surgical removal with negative surgical margins should be possible, so we did not prefer to take potential risk of biopsy, even endoscopically. Whipple operation performed with the pylorus preservation. Pathology reported that the tumor was well differentiated, well circumferentially solid pseudopapillary tumor of the pancreas, resected with all negative surgical margins and without any lymph node metastasis. At the 44th month of the surgery she is following up without any recurrent disease.

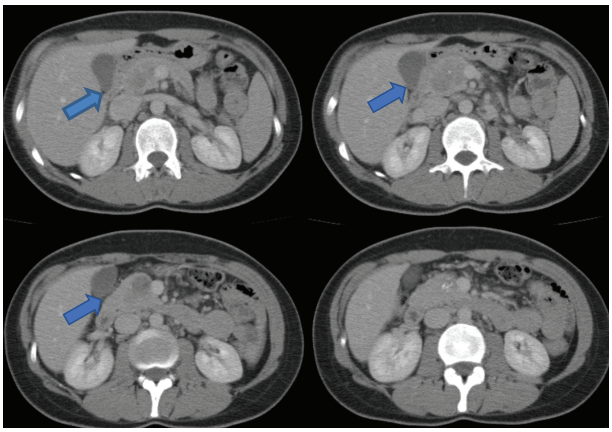

Figure 1: CT findings of the mass without vasculary invasion.

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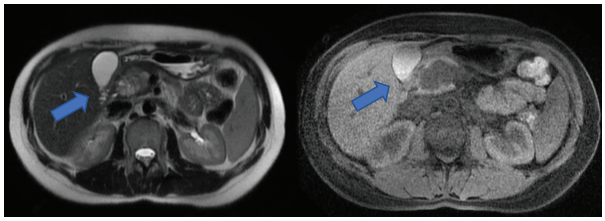

Figure 2: MRI findings of pancreatic head mass.

Discussion

SPT has shown a progressive increase in incidence, from 0.2-2.7 % of all exocrine tumors of the pancreas in the 1980's, to 6% in more recent reports, being higher in Asia [1,11,12]. Some authors have related this effect to specialization, improvement in diagnostics and wider recognition of the disease [10]. Although male patients have been reported, it predominantly affects young women, especially black and Asian, between 20 and 30 years of age [1,9,13,14]. Our patient was 30 years-old woman with mild symptoms at the diagnosis.

SPT is often asymptomatic until the tumor reaches larger size. In our case, she had 4 cm mass at the pancreatic head without any specific symptom and without invasion to surrounding structures. Abdominal pain or tenderness, palpable mass and jaundice are the most common reported symptoms [9,11,15-17]. Preoperatively, radiological diagnosis can be possible, a well localized and encapsulated, complex large masses (with solid and cystic components) without dissemination may encountered a suspicion [2,9,13,18,19]. In some patients all the tumor could be seem solitary, cystic part is not a rule. These findings especially in young females recedes experiments from adenocarcinoma, and also from neoadjuvant treatment choice.

Percutaneous biopsy may facilitate preoperative diagnosis [9,16,20,21]. But we know that it is not an easy and uncomplicated procedure such an complex-risky area, some authors reported the potential role of previous biopsy or trauma (drainage because of misdiagnosis etc.) in seeding and development of peritoneal carcinosis in pancreatic tumors [22,23]. And also additional contribution to the biopsy in this situation is contrary.

The treatment choice is basicly focused on the removal of the tumor totally. Reports have demonstrated resectability and long term survival with isolated portal vein and arterial resection and reconstruction [1,24].

Local invasion of peripancreatic tissue is the most common malignant pathologic feature [8,2,5]. Although angioinvasion and perineural invasion are regarded as evidence of malignant potential according to the WHO classification [5]. Old age, male sex, and tumor size larger than 5 cm has been reported as predictor factors of the malignant form [25,26].

Local recurrence or metastasis is unusual, and have been reported to be particularly rare in elderly patients [8,14]. The most common organ for metastasis is the liver, but lymph node, adrenal or peritoneal metastases have been reported [1,8,10,13,27]. Even in the presence of disseminated disease, patients have a good prognosis as long as the lesions are resected completely [1,10,27]. Tang et al. reported that the overall 5 year survival was 97% when metastases were removed successfully [14]. A case of liver transplantation was reported from Nagoya for unresectable multiple liver metastases of SPT (at the second year of the operation, she was disease free) [10].

In conclusion, due to the slow-growing character and low malignant potential, and favorable prognosis even in the presence of dissemination, an aggressive surgical approach from metastasectomy to liver transplantation, is advisable and removal of the tumors with negative surgical margins may result in curable disease [24,28,29]. Resectability rate and expectance for disease-free survival is high even in the presence of major vascular invasion or distant metastases [10,24,28,30]. Late recurrences were reported and this may indicate long term follow-up for these patients [9].

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