—Case Reports—

A 13-year-old girl with a Preoperatively Diagnosed Solid Cystic Tumor of the Pancreas

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Abstract

We report on a 13-year-old girl with a solid cystic tumor of the pancreas. She was referred to our hospital with abdominal pain in the left upper quadrant. Physical examination revealed a firm mass in the abdomen. Ultrasonography showed a clearly demarcated round mass with a mixture of solid and cystic components. Computed tomography confirmed that the mass measured 5 × 5 cm with low density in the cystic region and soft-tissue density in the solid region. Magnetic resonance also showed a clearly demarcated mass with a solid portion of low intensity on T1-weighted images and high intensity on T2-weighted images. We diagnosed a solid and cystic tumor of the pancreas and subsequently performed distal pancreatectomy. A firm, well-encapsulated tumor was found in the pancreas tail. The cut surface of the tumor consisted of a solid area with hemorrhage and a cystic area. Light microscopy of the tumor confirmed small neoplastic cells. Pathological diagnosis was solid pseudopapillary tumor (solid cystic tumor) of the pancreas. Surgery was successful, and the postoperative course was uneventful.
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Key words: solid and cystic tumor, pancreas

Introduction

Solid and cystic tumor of the pancreas (SCT) is believed to arise from acinar cells and has also been described as solid and cystic acinar cell tumor[1,2]. This tumor is usually observed in adolescent girls or young women, and the main symptom is an abdominal mass with or without abdominal pain[3]. To date only 180 cases of SCT have been reported in Japan[4]. Thus, this tumor is difficult to diagnose definitively before surgery because it is rare and the chief complaint is abdominal tumor with lack of definitive tumor markers or biochemical abnormalities[3]. We have previously reported on a 12-year-old boy with SCT[5]. Here, we report on a 13-year-old girl with a preoperatively diagnosed SCT.

Case Report

A 13-year-old girl was admitted to our hospital with sustained abdominal pain and a firm, smooth

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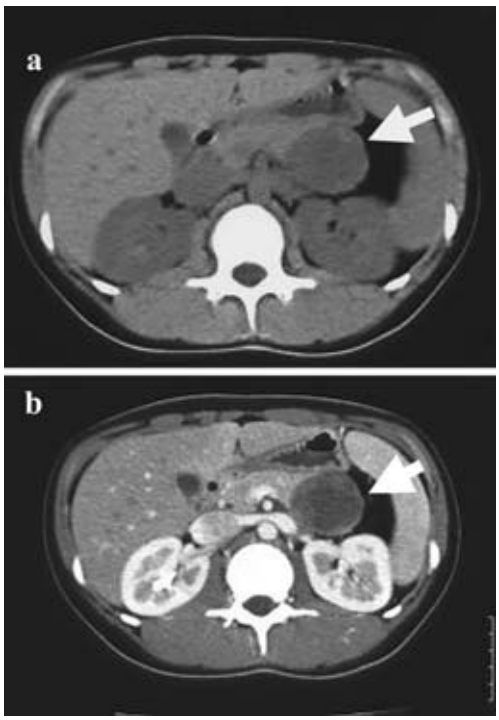

Fig. 1 **a**: CT findings of the tumor: A 5×5-cm tumor **(arrow)** with low density in the cystic region and soft-tissue density in the solid region was observed. **b**: Enhanced CT findings of the tumor: Irregular enhancement was observed.

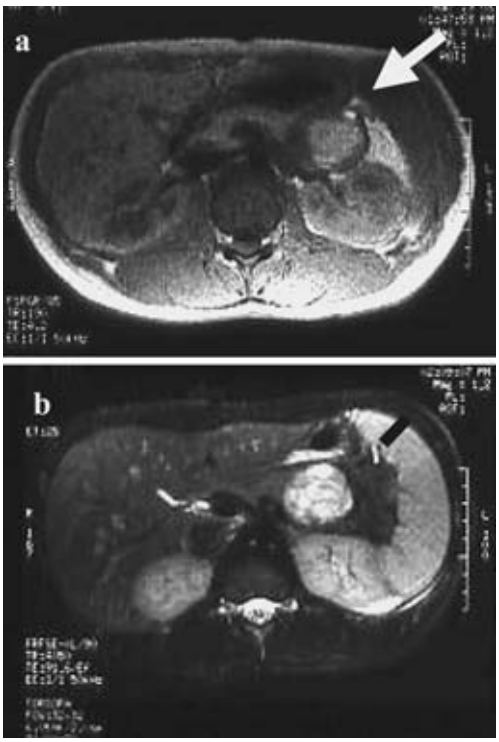

Fig. 2 **a**: MR findings: A 5×5-cm, well-demarcated tumor was observed with low intensity on T1-weighed images **(arrow)**. **b**: A 5×5-cm, well-demarcated tumor was observed with high intensity on T2-weighed images **(arrow)**.

mass in the left upper quadrant of the abdomen. The mass measured 3 × 3 cm. Physical examination revealed no anemia or jaundice and no swelling of supraclavicular, axillary, or inguinal lymph nodes. Hematological and biochemical examination revealed no abnormalities. Tumor markers, including serum carcinoembryonic antigen, alpha-fetoprotein, human chorionic gonadotropin, CA19-9, IAP, elastase 1, neuron-specific enolase, and urine vanilmandelic acid and homovanilic acid, were not elevated. Ultrasonography showed a clearly demarcated round mass with a diameter of 4 cm comprising a mixture of solid and cystic components in the left upper quadrant of the abdomen. Computed tomography (CT) confirmed that the mass was located on the left kidney, adjacent to the hilar portion of the spleen **(Fig. 1a)**. The mass measured 5 cm in diameter and exhibited low density in the cystic region and soft-tissue density in the solid region. CT with a contrast agent showed partial enhancement of the tumor but no enhancement in the cystic region **(Fig. 1b)**. Magnetic resonance (MR) also showed a clearly demarcated mass on the left kidney, adjacent to the hilar portion of spleen **(Fig. 2a)**. The solid portion of the tumor showed low signal intensity on T1-weighted MR and high intensity on T2-weighted images **(Fig. 2a, b)**. Gadolinium enhancement revealed dim ring enhancement of the tumor. ^{99m}Tc-MDP-scintigraphy, ^{67}Ga-scintigraphy, and ^{123}MIBG-scintigraphy demonstrated no abnormal accumulation in the tumor area. On the basis of these observations, we believed the tumor to be an SCT and subsequently performed distal pancreatectomy.

A firm, well-encapsulated tumor (size, 48 × 50 × 36 mm; weight, 48 g; **Fig. 3a**) was found in the pancreas tail. The cut surface of the tumor consisted of a solid area with hemorrhage and a cystic area **(Fig. 3b)**. Light microscopy of the tumor showed small neoplastic cells with eosinophilic or vacuolated cytoplasm and pseudo papillary features with a fine fibrovascular stalk. The tumor was also composed of a cystic lesion lined by foamy or hemosiderin-laden macrophages **(Fig. 3c)**. The surgical margin of the

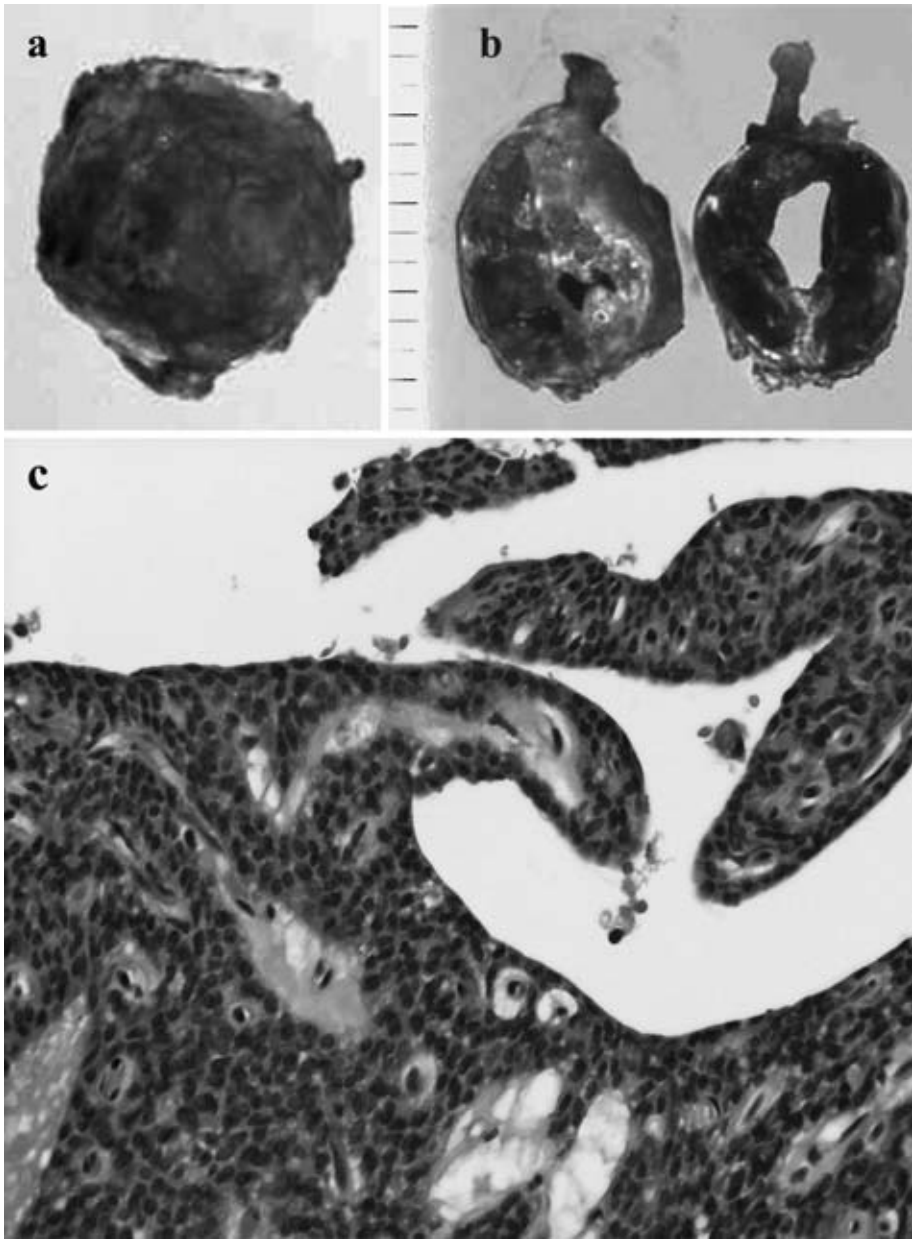

Fig. 3 **a**: Macroscopic findings of the tumor: The tumor measured 6×7 cm and was well defined with a fibrous capsule. The cut surface of the tumor consisted of a solid area with hemorrhage and a cystic area. **b**: Cut surface of the tumor: A solid portion with hemorrhage and a cystic portion were observed. **c**: Microscopic findings of the tumor: Small neoplastic cells having eosinophilic cytoplasm with a solid and pseudopapillary pattern and variable stromal sclerosis were observed.

pancreas tail showed no tumor cell invasion. We did not perform splenectomy. The pathological diagnosis was solid pseudopapillary tumor (solid cystic tumor) of the pancreas tail, which was encapsulated by a fibrous capsule. The tumor was considered to be a low-grade malignancy because of continuous invasion into the fibrous septa. However, the margin of the pancreas tail showed no tumor cell invasion microscopically. The postoperative course was uneventful, and the patient remains free of disease 5 years after surgery.

Discussion

To date, only 180 cases of SCT have been reported in Japan[4], indicating that this tumor is rare and may be difficult to diagnose before surgery. At our hospital, three cases have been pathologically

diagnosed as SCT; these cases were initially diagnosed as cystadenocarcinoma of the pancreas or pancreas tail tumor, but the present case was the only one to be correctly diagnosed before surgery. For the differential diagnosis of pancreatic tumors, we must consider pancreatoblastoma and microcystic adenoma, as well as SCT. Pancreatoblastoma usually occurs in 5- to 8-year-old boys, who are younger than typical patients with SCT. Microcystic adenoma usually occurs in the elderly, and CT findings reveal hypervascularity and sun-burst-like calcification in the center of the tumor. In addition to the above findings for differential diagnosis, a sharply demarcated large mass, a solid and cystic portion with CT numbers higher than those of water and the clear absence of internal septation are indicative of SCT[6]. Characteristic ultrasonographic findings of SCT are a clearly demarcated large pancreatic mass with a thick capsule, calcification, and a mixed solid and cystic pattern, depending on the degree of hemorrhage and necrosis[7]. A clearly demarcated tumor of the pancreas with areas of high signal intensity, suggesting hemorrhage, on T1-weighted images also strongly suggests SCT[8].

The treatment of first choice is complete surgical resection, which is true for the primary tumor, metastases, and local recurrences. Adjuvant therapy in patients who have undergone curative resection is unnecessary and does not appear to improve the prognosis[9]. The prognosis is generally good in cases of complete resection, although some cases of multiple metastasis or invasion to adjacent organs have been reported[10].

In the present case, SCT was diagnosed because an earlier patient of ours was a boy with SCT[5]. For diagnosing SCT, it is important to take into consideration the differential diagnosis for abdominal tumors near the pancreas in adolescent girls. If SCT can be diagnosed preoperatively, necessary surgery may be performed and postoperative chemotherapy is not necessary.

In conclusion, we must consider the differential diagnosis for abdominal tumors adjacent to the pancreas and adrenal gland region, particularly in adolescent girls.

Important diagnostic points for SCT are as follows:

1) Preferentially occurring in young (adolescent) girls

2) Abdominal tumor with or without abdominal pain

3) No abnormal findings on hematological and biochemical examination

4) Mixed solid and cystic pattern with hemorrhage and necrosis on ultrasonography, CT, and MR

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