Malignant Transformation of an Epidermoid Cyst in an Intrapancreatic Accessory Spleen: A Case Report

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Abstract

A 33-year-old man was evaluated because of an incidentally found cyst in the pancreatic tail, which was first seen 6 years ago. The cyst was a unilocular cystic mass, 13.0 cm in diameter, and had increased in size in last 2 months. On F-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT), the cystic wall showed increased FDG uptake. The patient underwent distal pancreatectomy with suspicion of mucinous cystic neoplasm of the pancreas. The mass turned out to be a squamous carcinoma arising from an epidermoid cyst in an intrapancreatic accessory spleen (ECIPAS). FDG PET/CT may assist recognition of a potential malignant lesion arising from an ECIPAS.

Keywords Epidermoid cyst · Intrapancreatic accessory spleen · Malignant transformation · Fluorodeoxyglucose

Introduction

An accessory spleen is a small nodule of spleen which is separate from the main body of spleen. It is observed in about 10% of the population, with one out of six cases showing in the pancreatic tail [1]. Splenic cysts are found in 0.07% of the population [2], and splenic epidermoid cysts constitute approximately 20% of all splenic cysts [3]. Epidermoid cysts in intrapancreatic accessory spleen (ECIPAS) is rare, with less than 60 cases, and is thought to be a benign entity and harbor no malignant potential [4]. Here, we describe a case of ECIPAS with malignant transformation.

Case Report

A 33-year-old male was referred for evaluation of a pancreatic cyst. Six years ago, the cyst was first found on abdominal ultrasound, which was performed as a health checkup, and the patient

had no symptom. The long diameter of the cyst was 3.0 cm and had not changed until 2 months ago. Recently, the cyst had grown to 13.0 cm. He had no significant past medical history. He did not complain any specific symptom, and no abnormality was found on physical examination. Serum carbohydrate antigen 19–9 (CA 19–9) was elevated to 50.0 U/mL, and serum carcinoembryonic antigen (CEA) was within normal range (0.6 ng/mL). On computed tomography (CT) and magnetic resonance imaging (MRI), the cyst was seen as a unilocular cystic mass in the pancreatic tail with irregular wall thickening (maximal thickness, 8.0 mm). The pancreatic duct was not communicated with the cyst and was not dilated.

The patient underwent F-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) imaging. Briefly, FDG PET/CT images were acquired from the cerebellum to the mid-thigh (3 min/bed) with using a Biograph TruePoint 40 scanner (Siemens Medical Systems, Knoxville, TN). The patient fasted for 13 h before imaging, and peripheral blood glucose level was 95 mg/dL prior to the injection of FDG. Low-dose CT (36 mA, 120 kVp) was acquired, and PET images were iteratively reconstructed with CT-based attenuation correction. FDG PET/CT revealed that the cyst had increased FDG uptake along its wall, which corresponded to the irregular wall thickening, and maximum standardized uptake value (SUVmax) was 23.5. No other abnormality was found in the rest of the body (Fig. 1).

Endoscopic ultrasonography-guided fine needle aspiration biopsy (EUS-FNA) failed to make a diagnosis. CA 19–9 and CEA

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Fig. 1 F-18 fluorodeoxyglucose (FDG) positron emission tomography/computed tomography revealed a cystic mass in the pancreatic tail. The cystic wall showed increased FDG uptake, and SUVmax was 23.5. Maximum intensity projection image (a) and fusion axial image (b) were shown

in the cystic fluid were elevated to 6316.0 U/mL and 176.7 ng/mL, respectively. The patient underwent laparoscopic distal pancreatectomy and splenectomy, with suspicion of mucinous cystic neoplasm of the pancreas. The pathology revealed an epidermoid cyst in an intrapancreatic accessory spleen, with a 2.5-cm-sized squamous cell carcinoma arising from the cyst (Fig. 2). The carcinoma was limited to the pancreas, with focal invasion into the cystic wall. There was no invasion to pancreatic parenchyma, and no lymphovascular or perineural invasion was identified. After 4 months, serum CA 19-9 was returned to normal range (6.1 U/mL). The patient was followed-up with abdominal CT in every 3 months for 2 years, and no recurrence was observed.

Fig. 2 Gross appearance of the epidermoid cyst in an intrapancreatic accessory spleen. A squamous carcinoma was arising from the cyst, and the lesion showed increased fluorodeoxyglucose uptake on positron emission tomography/computed tomography

Discussion

An ECIPAS is an extremely rare entity. There were 56 patients in 47 case reports of ECIPAS in English literature [4]. ECIPAS was more common in female than male (32 females and 24 males), and the median age was 47 years (range, 10–70 years). A majority of patients were asymptomatic (33 out of 56) or had nonspecific symptoms such as abdominal pain. Elevated levels of serum CA 19-9 (20 out of 37) and CEA (1 out of 27) were also observed. A typical ECIPAS is a unilocular cyst (89%) in the pancreatic tail, with median long diameter of 3.4 cm (range, 1.3–15.0 cm). And keratinized/non-keratinized stratified squamous epithelium or cuboidal epithelium is covering the cyst, circumscribed by normal spleen tissues [5]. A cyst containing serous or non-serous fluid and lacking skin appendages and mature lymphoid tissues are key features differentiating epidermoid cyst from dermoid cyst and lymphoepithelial cyst [5, 6].

Due to its rarity, preoperative diagnosis of ECIPAS is often difficult. A key clue in diagnosis of ECIPAS is identification of surrounding ectopic splenic tissue. The cystic wall of ECIPAS and spleen shared similar enhancement patterns on dynamic contrast-enhanced CT or MRI [7, 8]. Tc-99 m heat-damaged RBC scan also can be used to detect ectopic splenic tissue, and combined SPECT/CT may improve diagnosis [9]. However, as with our case, the usefulness of Tc-99 m heat-damaged RBC scan has not been explored in the diagnosis of ECIPAS. Also, depiction of the ectopic spleen in the pancreas depends on the amount of ectopic splenic tissue [10]. There was only one case of ECIPAS who underwent FDG PET/CT, and the lesion showed mild FDG uptake with a SUVmax of 1.9, which is consistent with a benign disease [11].

As there is no specific feature of ECIPAS other than presence of ectopic splenic tissue, making a diagnosis of ECIPAS

based on CT, MRI, or FDG PET/CT can be difficult. Of note, differentiation between mucinous cystadenoma of the pancreas and ECIPAS can be difficult, because both can occur in the pancreatic tail, especially in middle-aged females [12, 13]. Although it is a rare entity, recognition of ECIPAS as a differential diagnosis in addition to mucinous cystadenoma, mucinous cystadenocarcinoma, and intraductal papillary mucinous neoplasms may facilitate accurate diagnosis.

To our knowledge, our case is the first case with malignant transformation of an ECIPAS. Previous studies have suggested that ECIPAS is a benign disease, and by making an accurate diagnosis of ECIPAS, unnecessary surgery could be avoided [4]. Although malignant transformation of an epidermoid cyst is rare, it is previously reported in epidermoid cysts of the brain and skin [14, 15]. We showed that an ECIPAS can be transformed to a squamous carcinoma, and combined with spleen-specific imaging, FDG PET/CT may identify malignant transformation of an ECIPAS [16].

Conclusion

We described a case with malignant transformation of an ECIPAS. Although an ECIPAS was considered as a benign entity, we showed that malignant transformation can occur from an ECIPAS, and FDG PET/CT may aid the diagnosis of malignant transformation from an ECIPAS.

Compliance with Ethical Standards

Conflict of Interest Jiyoung Wang, Won Jun Kang, and Hojin Cho declare that they have no conflict of interest. There is no source of funding.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee.

Informed Consent The institutional review board of our institute approved this retrospective study, and the requirement to obtain informed consent was waived.

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