

Pancreatic inflammatory myofibroblastic of the pancreatic neck: a case report and literature review

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Case report

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Abstract

Background

Pancreatic inflammatory myofibroblastic tumor (IMT) is a relatively rare disease, which is often confused with pancreatic cancer or pancreatic neuroendocrine tumor. Its histological features are fibroblast and myofibroblast proliferation, accompanied by varying degrees of inflammatory cell infiltration.

Case presentation

Herein, we presented the management of IMT occurring at the neck of pancreas. A 66-year-old female patient was diagnosed as pancreatic neck mass after a series of tests. Then she underwent enucleation of pancreatic neck tumor followed by pathological diagnosis of IMT. Meanwhile, we reviewed the clinical features and pathological diagnosis and treatment of pancreatic IMT which were previously reported. To our knowledge, this is the unique case of enucleation of pancreatic IMT. In the management of pancreatic IMT,

Conclusion

we concluded that enucleation may be a safe and efficient surgical method for the management of pancreatic IMT and may also provide a better prognosis. Further accumulation of cases is required to explore the surgical measure of pancreatic IMTs.

Introduction

Inflammatory myofibroblastic tumor(IMT)is an uncommon mesenchymal tumor of unknown pathogenesis that is malignant and aggressive potential, with a global incidence of less than 1%^{1,2}. IMT most commonly occurs in children's or young adults' lungs, followed by the head and neck³, liver⁴, pancreas⁵, genitourinary tract⁶ and thyroid⁷. The clinical presentation caused by pancreatic IMT varies depending on its anatomic location and the final diagnosis of most lesions requires pathological examination. The most common site for pancreatic IMT is the pancreatic head and surgical resection may be the first choice at present. To our knowledge, only 29 cases of pancreatic IMT have been reported in the English literature, and none of which was performed the enucleation of the tumor. Herein, we presented an unusual pancreatic neck IMT occurring in a 66-year-old female patient. It may be the first case of enucleation of pancreatic IMT. Due to its relatively lower incidence and unspecific manifestations, the clinical and histological features, as well as diagnosis and treatment of pancreatic IMT are discussed.

Case Report

A 66-year-old female patient was admitted to Shulan (Hangzhou) Hospital on January 13, 2020 for complaining of finding a pancreatic mass. 4 days ago, she went to the local hospital for follow-up examination and abdomen ultrasonography showed a hyperechoic foci in the neck of pancreas,then the patient was transferred to our department for further treatment. She had a history of right pulmonary wedge resection for adenocarcinoma in 2014 and right hemicolectomy for colon cancer in 2018. Physical examination was unremarkable and laboratory results including complete blood count, c-reactive protein and tumor markers were all within the normal range. The ultrasound scan revealed a 2.5×1.5 cm mass in the neck of pancreatic. Dynamic contrast enhanced magnetic resonance imaging(MRI)scan showed an abnormal soft-tissue heterogeneous mass in the neck of pancreas, which appeared hyperintense on T1-weighted image and mild hyperintense on T2-weighted image. Centripetal enhancement pattern was observed during the delayed phase of contrast imaging. (Fig. 1). Whole body ¹⁸F-fluorodeoxyglucose (FDG) positron emission tomography/PET/CT examination revealed a 2.3×1.4

cm mild-to-moderate FDG uptaked nodule in the front of the pancreatic neck (SUVmax 3.87) with normal scans of her head, neck, chest, and colon(Fig. 1). The imaging findings were highly suggestive of pancreatic IMT. However, the possibility of metastatic tumor could not be ruled out basing on the history of lung and colon cancer. Patient underwent the enucleation of pancreatic mass with a tentative diagnosis of pancreatic IMT after MDT discussion. During the laparotomy, a hard protruding mass with a size of 2.3×1.5 cm was observed on the pancreatic neck. A subsequent enucleation was performed on the patient. The entire mass was fleshy with a grayish-white cut surface and the intraoperative frozen section confirmed that it was IMT. We decided to preform enucleation only. Therefore, post-operative detailed histopathological examination was performed revealed that the carcinoma cells were positive for staining for desmin vimentin, CD34, CD31, BCL2 and β -catenin, and negative for S-100 Pan-CK (AE1/AE3) Caldesmon, DOG1, CD117 smooth muscle actin, P53. A diagnosis of pancreatic neck IMT was established based on the histopathological results(Fig. 2). The postoperative recovery was uneventful, and the patient was discharged on postoperative days 11(Fig. 3). No adjuvant treatment was administered and no obvious evidence of metastasis or recurrence within the next 10 months of follow up.

Discussion

IMT is a rare special type of disease, which was first reported in the lungs^{8,9}, the name IMT is often termed differently in primary research, including plasma cell granuloma, plasma cell pseudotumor, inflammatory pseudotumor, inflammatory fibroxanthoma and histiocytoma¹⁰. IMT can occur almost anywhere in the body, including lungs, liver, bladder, mesenteric, neck¹¹⁻¹³. However, IMT arising from the pancreas are extremely rare. To date,only 29 cases have been reported in the English literature. We conducted a brief literature review of reported cases with pancreatic IMT so as to better understand the pancreatic IMT, as summarized in Table 1^{5,10,14-34}. Of these cases 20 were male(20/29, 69%) and 9 were female (9/29, 31%) with obvious male predilection. The tumor diameter for all reported cases ranged from 1.5 to 15 cm. Almost tumors were pancreatic head (21/29 patients) followed by the pancreatic tail (4/29 patients) or pancreatic body (3/29 patients), which may represent that pancreatic IMT was more common in pancreatic head, however, more evidence is required.

Pancreatic IMT can occur at all ages, but prefer children and young adult³⁵. We found in the English literature that the age range for all reported cases is from 6 months to 82 years (mean age 42 years). The current patient is 66 years old, which matches the age reported in the literature.

As described previously, the clinical presentation caused by pancreatic IMT varies depending on its anatomic location, from asymptomatic to hemorrhagic shock due to rupture of the spleen^{19,22}. Almost all of pancreatic IMT have similar nonspecific symptoms regardless of their site of tumor, that is, abdominal discomfort, abdominal distension, abdominal pain, general fatigue and wight loss have also been recorded at the time of presentation. Obstructive jaundice may be noted in typical patients with pancreatic head IMT. It can also obstruction of the pancreatic duct and induced chronic pancreatitis with abdominal discomfort, diarrhea, and indigestion²³. The IMT arising from the pancreatic tail also can obstruction of blood vessels of the spleen, resulting in rupture of the spleen with severe abdominal pain and hemorrhagic shock²². However, our patient was located at the neck of the pancreas, she came to the hospital without any special symptoms.

The preoperative laboratory findings of pancreatic IMT were nonspecific for diagnosis of disease. Only few patients with a solitary mass occurring in the head of the pancreas may be elevated in both serum total bilirubin, amylase, and carbohydrate antigen 19 – 9 due to obstruction of the bile duct or pancreatic duct²⁶. The radiological features are often deceptive. Ultrasound, CT and MRI examinations show a mass lesions mimicking pancreatic cancer or pancreatic neuroendocrine tumor. Also similar to other malignant tumors, whole body ¹⁸F-FDG PET/CT also showed elevated SUVmax³⁶, this feature can distinguish IMT from non-neoplastic lesions, such as pancreatic pseudocyst, swollen lymph nodes. In addition, whole body 18F-FDG PET/CT are best tools in detection of tumor recurrence or distant metastasis. More interestingly, even with standard intraoperative frozen pathology may not provide definitive information to distinguish pancreatic IMT from pancreatic inflammatory pseudotumor.

The definitive diagnosis of IMTs rely on histological evaluations and immunohistochemical tests³⁷. The histological features of IMTs are spindle-shaped cells accompanied by varying degrees of inflammatory cell^{38,39}. Coffin et al. suggested that clonal cytogenetic abnormalities involving the anaplastic lymphoma kinase (ALK) gene on the short arm of chromosome 2 at 2p23, which occur in approximately 50% of IMTs³⁷, its can be a useful test in the definitive clinicopathologic diagnosis and when positive. In addition, most extrapulmonary IMTs display immunohistochemical reactivity for SMA, Desmin, tissue cell marker CD68 and vascular markers CD34⁴⁰.

To date, no standard consensus regarding the treatment of pancreatic IMT has been established. However, almost all of authors have recommended surgical resection of the lesion as the primary therapeutic option for pancreatic IMT. This was based on the analysis of 29 cases of pancreatic IMT. The surgical approach is related to the location of the lesion on the pancreas. For pancreatic head IMT, a pancreaticoduodenectomy is recommended, while a distal pancreatectomy is recommended for pancreatic body or tail IMT. Pancreatic IMT often invasion surrounding organs, such as colon, duodenum, stomach, surgery with invasion of adjacent organ may remain the therapeutic option. However, no widely accepted theory exists for such low-grade malignant lesions, whether radical surgery is necessary or not requires a large number of further clinical studies to be verified. Radiation therapy, chemotherapy and high-dose steroid therapy have also been used in patients who resection is neither complete nor possible or in patients with malignant disease status postsurgical resection^{20,28,41}. The spontaneous regression of pancreatic IMT has been reported only infrequently²⁸. Given that our patient was an elderly, infirm female who had pancreatic neck IMT only, sparing the head and body, MDT discussion suggests that enucleation may be more benefit therapeutic option. No adjuvant treatment was administered following enucleation of pancreatic IMT, the patient remained symptom-free and healthy without evidence of recurrence tumor or metastasis was observed during the ten months following surgery. Although there is only 1 reported case of surgical measure with enucleation in patients with IMT, such surgical measure could be considered in the future. Further accumulation of cases is required to explore the surgical measure of pancreatic IMTs.

Pancreatic IMT regarded as low-grade malignancy, the prognosis is generally favorable. However, its exhibits the potentiality of malignancy, distant metastasis and recurrence, close and long-term follow-up after surgery must be carried out.

Conclusion

In summary, IMT of the pancreas is rare, we reported a rare pancreatic neck IMT, upon confirmation of the diagnosis, a decision was made to management the patient with enucleation to confirm if radical surgery could be avoided. This is the first reported case whereby enucleation use has resulted in favorable prognosis of pancreatic IMT. By referring to previously reported cases, we concluded that surgical resection may be the preferred treatment and may provide a better prognosis. In addition, using enucleation as a surgical measure for treatment of patients with IMT may also have a good prognosis. This avoids radical surgery with its associated serious complications.

Abbreviations

| | |
|-----|------------------------------------|
| CT | computed tomography |
| IMT | inflammatory myofibroblastic tumor |
| MRI | magnetic resonance imaging |
| ALK | anaplastic lymphoma kinase |
| MDT | multidisciplinary team |
| DWI | diffusion-weighted imaging |

FDG fluorodeoxyglucose

PET positron emission tomography

H&E hematoxylin and eosin

Declarations

Consent for publication

Written permission to publish this case report was obtained from the patient.

Competing interests

The authors declare that they have no competing interests

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Authors' contributions

All authors read and approved the final manuscript. Review of patient data and critical comments were performed by ZTC, YXL, MXL, TZ, LLZ, DLW, and SZL. The literature review was completed by ZTC and DLW. TZ reviewed and described the pathologic findings. The manuscript was written by ZTC, YXL, DLW, and LLZ.

Availability of data and materials

Not applicable.

Ethics approval and consent to participate

Not applicable as this is not a study.

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Tables

Table1

Reported cases of pancreatic inflammatory myofibroblastic tumor (IMT) in the English literature.

| Cases | Sex | Age(y) | Location | Diameter (cm) | Symptoms | Treatment | Follow-up | References |
|-------|-----|--------|----------|---------------|-----------------------------|---|---|------------|
| 1 | M | 70 | PT | 3.8 | asymptomatic | DP + splenectomy | Disease-free at 10 months | 29 |
| 2 | M | 62 | PH | 3 | jaundice | PD | Disease-free at 6 years | 14 |
| 3 | M | 56 | PH | no | jaundice | PD | Disease-free at 5 years | 14 |
| 4 | M | 50 | PH | 5 | Jaundice, abdominal pain | PD | Disease-free at 4 years | 14 |
| 5 | F | 57 | PH | Not available | jaundice | PD | Disease-free at 3 years | 14 |
| 6 | M | 45 | PH | Not available | jaundice | PD | Disease-free at 10 years | 14 |
| 7 | F | 32 | PH | 3 | abdominal pain | PD | Disease-free at 12 years | 14 |
| 8 | F | 42 | PB | 7 | abdominal pain, weight loss | DP | Disease-free at 6 months | 15 |
| 9 | F | 8 | PBT | 10.7 | abdominal mass | DP | Disease-free at 2 years | 16 |
| 10 | M | 35 | PH | 5×4×3 | abdominal pain, weight loss | PD | Lung metastasis at 6 years | 17 |
| 11 | M | 55 | PH | 1.5 | asymptomatic | PD | Disease-free at 28 months | 10 |
| 12 | M | 69 | PBT | Not available | abdominal pain | DP + splenectomy + colon splenic flexure | die after 7 months of hospitalization due to sepsis | 18 |
| 13 | M | 65 | PB | 2 | asymptomatic | DP + splenectomy | Disease-free at 3 years | 19 |
| 14 | M | 56 | PT | 5×7 | melenia | DP + splenectomy | Disease-free at 18 months | 30 |
| 15 | F | 13 | PH | 3 | Vomiting, weight loss | PD | Disease-free at 7 years | 20 |
| 16 | M | 10 | PH | 2.2 | abdominal pain, anepithymia | prednisolone, cefuroxime | Disease-free at 6 years | 20 |
| 17 | M | 19 | PT | 8.2×6.5×6.0 | abdominal pain | DP + splenectomy | Disease-free at 6 years | 22 |
| 18 | M | 44 | PH | 6×4 | abdominal pain, vomiting | PD | Disease-free at 1 years | 23 |
| 19 | M | 65 | PH | Not | abdominal | PD | Not available | 25 |

| | | | | available | pain | | | |
|----|---|-----|----|-----------|-----------------------------|--|---------------------------|---------|
| 20 | M | 0.5 | PH | 4 | jaundice | PD | Disease-free at 3.5 years | 31 |
| 21 | F | 32 | PH | 4.8×3.2 | abdominal pain | PD | Disease-free at 2.5 years | 26 |
| 22 | M | 46 | PH | 8×6×5 | jaundice | PD | Not available | 27 |
| 23 | M | 69 | PH | 4×3 | Vomiting, anepithymia | PD | Disease-free at 3 years | 21 |
| 24 | M | 15 | PH | 5×5×4.3 | abdominal pain, fever | PD | Not available | 24 |
| 25 | M | 1 | PH | 4×3 | asymptomatic | PD | Not available | 34 |
| 26 | F | 82 | PH | 5 | abdominal pain | None | Disease-free at 9 months | 28 |
| 27 | M | 61 | PT | 15×13×7 | abdominal pain | DP+ left surrenalectomy + left hemicolectomy + splenectomy | Disease-free at 8 months | 5 |
| 28 | F | 11 | PH | 3.4 | abdominal pain, weight loss | PD | Not available | 32 |
| 29 | F | 13 | PH | 2.5 | abdominal pain | PD | Disease-free at 4 years | 33 |
| 30 | F | 66 | PN | 2.3×1.5 | asymptomatic | enucleation | Disease-free at 9 months | current |

*F*female, *M*male, *PN*pancreatic neck, *PH*pancreatic head, *PT*pancreatic tail, *PB*pancreatic body, *PBT*pancreatic body and tail,

PD pancreaticoduodenectomy, DP distal pancreatectomy.

Figures

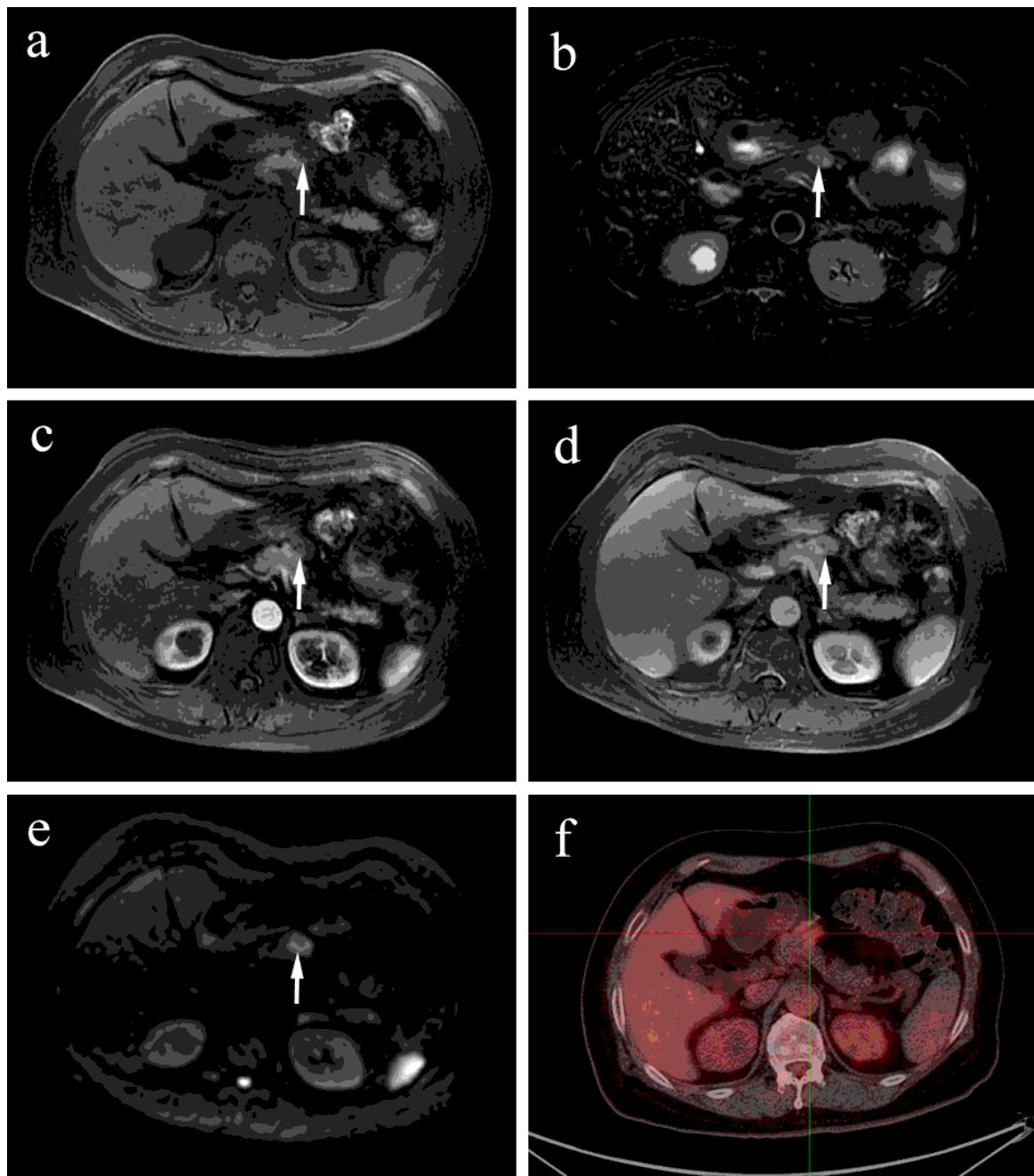


Figure 1

Dynamic contrast enhanced MRI and 18F-FDG PET/CT reveal one irregular lesion in the pancreatic neck. (a) The lesion of pancreatic neck (white arrow) was hyperintense in T1-weighted imaging. (b) The irregular lesion (white arrow) has slightly hyperintense in T2-weighted imaging. (c) Arterial phase imaging revealed slightly enhancement of the pancreatic lesion (white arrow). (d) The lesion (white arrow) showed persistent enhancement in venous phases imaging. (e) The pancreatic mass (white arrow) has significant hyperintense in diffusion-weighted imaging (DWI). (f) 18F-FDG PET/CT revealed hypermetabolic pancreatic neck nodule measuring 2.3×1.4 cm ($SUV_{max}=3.87$).

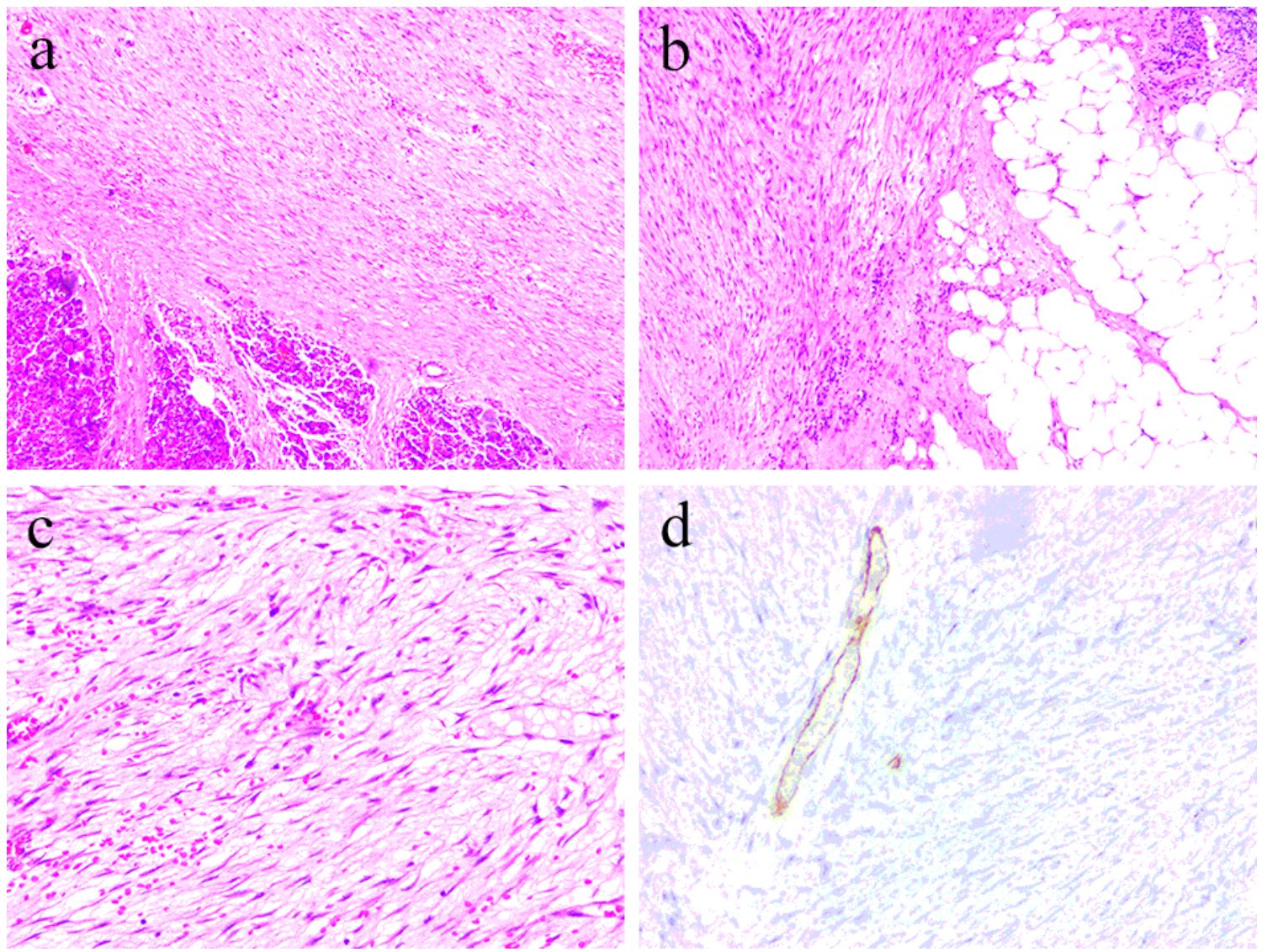


Figure 2

10 days after the surgical resection, Computed tomography showed the pancreatic neck IMT has been enucleated and the tissue of pancreas remained intact.

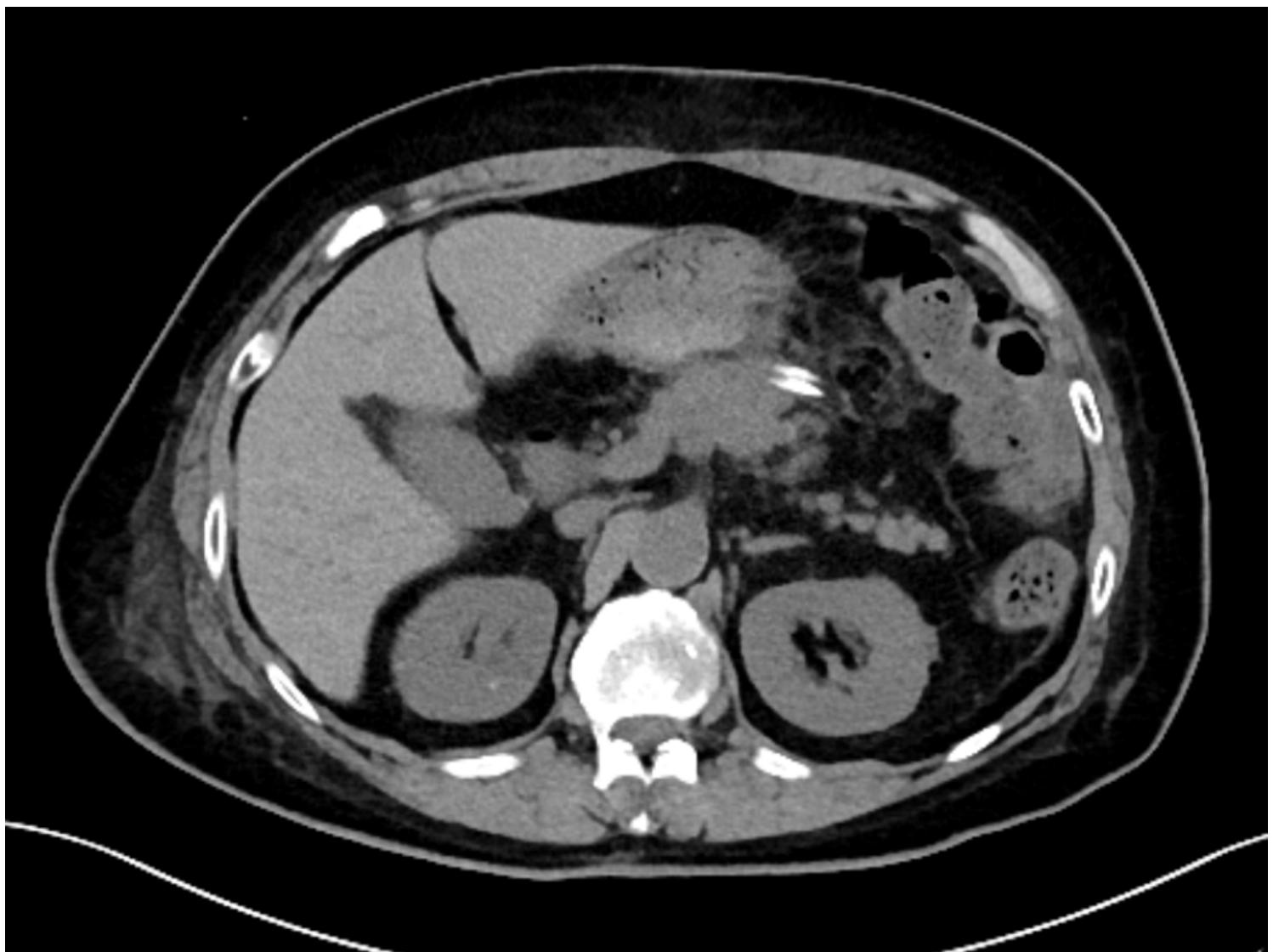


Figure 3

The pathological findings of the resected specimen revealed IMT. (a), (b) and (c) histologic images showed mixed components of dense myofibroblastic tissues and few inflammatory cells, with neoplastic cells infiltrate the surrounding fat tissue (hematoxylin and eosin [H&E] staining). (d) Immunohistochemical studies show positivity for smooth muscle actin.