Pancreatic sarcoidosis mimicking neoplasia: Case report

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ABSTRACT

INTRODUCTION: Although sarcoidosis rarely involves the pancreas, such involvement may mimic pancreatic cancer. We herein report a case of pancreatic sarcoidosis giving rise to a cancer-mimicking retention cyst, concomitant with a neuroendocrine adenoma.

PRESENTATION OF CASE: A 47-year-old Caucasian male presented to follow-up for a benign-appearing cyst of the tail of the pancreas, detected incidentally on CT scan done for a urinary stone in 2017. He had been asymptomatic since his last presentation. The lesion was found to have increased in size from 1 cm to 3 cm in greater diameter. Yet, a CT angiography showed no evidence of invasion of surrounding organs, vessels, or lymph nodes. The patient had previous medical history of treated sarcoidosis, hypertension, recurrent nephrolithiasis, and gout. Due to the size increment a neoplastic cystic lesion was considered and distal pancreatectomy was performed. Pathologic examination revealed a retention cyst associated with chronic pancreatitis and the presence of non-caseating granulomas consistent with sarcoidosis. In addition, a neuroendocrine adenoma, and an adjacent focus of pancreatic intraepithelial neoplasia-1 and 2 were noted.

DISCUSSION: Such presentations may be asymptomatic, as in this case, and a multidisciplinary workup is often required. Care must be taken to rule out pancreatic cancer. A possible relationship between pancreatic sarcoidosis and pancreatic cancer merits further study.

CONCLUSION: The diagnosis of pancreatic sarcoidosis is difficult, and conclusive diagnosis requires histopathologic assessment.

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1. Introduction

Sarcoidosis is an inflammatory disease characterized by non-caseating granulomas, often multisystemic, and of contentious etiology [1]. The disease was first described in 1877, and sarcoidosis involving the pancreas was not reported until 1937. Pancreatic sarcoidosis is a rare entity, found in only 1–6% of patients with systemic sarcoidosis [2]. We report a case of sarcoidosis manifesting as a pancreatic retention cyst, with the absence of relevant pancreatic symptoms. This case is reported in line with SCARE criteria [3] [Agha].

2. Presentation of case

Here we report a case of a 47-year-old Caucasian male who was incidentally found to have a benign-looking 1 cm cystic lesion of the tail of the pancreas, on CT scan done as part of nephrolithiasis work-up in 2017.

The patient was previously diagnosed with sarcoidosis which was treated with methotrexate. He also had hypertension, treated with Coversyl, and gout, treated with Allopurinol. He had previous episodes of nephrolithiasis for which he underwent a ureteroscopy with lithotripsy. The patient was also known to be lactose intolerant.

The patient was scheduled for follow-up for surveillance of his pancreatic findings. In May 2019, the patient presented to follow-up and was asymptomatic. Serum chemistries revealed normal serum amylase and lipase of 61 U/L and 45 U/L respectively. CT with abdominal angiography showed that the cystic lesion increased in size to 3 cm with an enhancing wall and mural nodules (Fig. 1). The pancreatic angiography at the level of the lesion appeared to be narrowed down with the development of epiploic varices. There

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was no evidence of communication with the pancreatic duct or of invasion of surrounding organs or lymph nodes. The gallbladder had a normal density with no focal lesions seen and no intra or extrahepatic biliary ductal dilation. There was no intra-abdominal or retroperitoneal lymphadenopathy. Other gastrointestinal findings were only significant for the presence of diverticula in the sigmoid colon. These findings raised the possibility of a neoplasm and distal pancreatectomy was performed by the surgery department, after the patient’s consent and the surgical specimen consisting of a 6 cm × 3.8 cm × 2 cm portion of distal pancreas was sent to pathology. The cut sections showed a diffusely indurated and atrophic pancreatic parenchyma with a 2 cm × 1 cm simple cyst located 2 cm from the closest transection margin. The cyst nature was confirmed by histopathologic examination as a retention cyst lined by a low cuboidal epithelium with associated chronic pancreatitis, along with the presence of numerous non-caseating granulomas consistent with sarcoidosis (Figs. 2 and 3). In addition, two incidental lesions were identified: a 1.5 mm neuroendocrine adenoma, and an adjacent 4 mm focus of pancreatic intraepithelial neoplasia-1 and 2. Post-operatively, the patient suffered from splenic artery thrombosis and a splenic abscess that were treated, and he has fully recovered.

3. Discussion

We report a case of sarcoidosis manifesting as a retention cyst of the pancreas. Pancreatic sarcoidosis in a living patient is a rare entity; the characteristic granulomas are usually found postmortem [4]. This particular patient did not seem to show any symptoms of his previously diagnosed sarcoidosis, despite the lesion in the pancreas. In fact, the lesion was incidentally discovered during radiologic workup for an unrelated condition. To our knowledge, this is the first reported case of pancreatic sarcoidosis causing a retention cyst that mimicked a neoplastic process along with concomitant neuroendocrine adenoma of the pancreas.

The retention cyst in this patient was secondary to the sarcoidosis-induced chronic inflammation and fibrosis. In fact, retention cysts in the pancreas are often associated with chronic pancreatitis [5]. The underlying fibrosis, in addition to the expanding nature of the cyst and the diffuse granulomas, may well account for the narrowing of the pancreatic vein.

Cohen et al. discussed the relation between sarcoidosis and neoplasia [6]. A retrospective cohort study of Swedish patients with sarcoidosis [7] showed that sarcoidosis patients had an increased risk of various kinds of cancer. The authors suggested that the increased risk of cancer following sarcoidosis may be due to the fact that sarcoidosis causes chronic inflammation, which eventually gives rise to malignancy [7]. Moreover, similar (but not identical) combinations of tumors identified in this case, have been previously reported, such as pancreatic neuroendocrine tumors with
extra-pancreatic sarcoid [8,9]. However, a definite link between sarcoidosis and malignancy is yet to be confirmed. More studies should aim at exploring the pathophysiology that may connect the two pathologies.

Pancreatic sarcoidosis is an elusive diagnosis to establish. Radiology is a valuable tool when working up this disease, but definitive diagnosis depends on pathology. More ominous diagnoses such as adenocarcinoma must be ruled out since symptoms may be non-specific and mimic cancer, and pancreatic sarcoidosis can present concomitantly with pancreatic cancer [10]. Furthermore, sarcoidosis that does not involve the pancreas can co-present with primary tumors of the pancreas, including neuroendocrine tumors [8] and pseudopapillary tumors [11].

4. Conclusion

Pancreatic sarcoidosis is rare and its clinical presentation may be deceptive. Definitive diagnosis remains challenging and requires histopathologic assessment.

Conflicts of interest

None.

Sources of funding

None.

Ethical approval

We have reported a single case, not a clinical study, with no requirement for ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Georges Chedid: writing – original draft.
Jad Gerges Harb: writing – original draft.
Hussein A. Noureddine: writing – original draft.
Claude Tayar: investigation, writing – review & editing.
Selim M. Nasser: conceptualization, investigation, writing – review & editing.
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Guarantor

Dr. Selim M. Nasser.

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