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Synchronous granular cell tumors of the pancreas and cecum

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ABSTRACT

Granular cell tumors (GCT) are rare and typically benign. Diagnosis is challenging due to nonspecific imaging characteristics and symptomatology. Herein, we report a combination of pancreatic/cecal GCTs in a 43-year-old man.

Contrast enhanced MDCT demonstrated a 1.5 cm well-defined homogeneous intraluminal cecal mass and a 1.6 cm slightly hypervascular pancreatic body mass. On MRI, the pancreatic mass showed increased enhancement on post-gadolinium delayed sequences. Diagnosis was confirmed by excisional pathology (S100 and CD68, PAS-D positive).

Radiologists, gastroenterologists, and surgeons should ponder the possibility of GCTs in the differential diagnosis of any small, pancreatic or cecal well-defined tumor.

1. Introduction

Granular cell tumors (GCT) are very uncommon tumors that usually appear as solitary, small neoplasms and that habitually follow a benign course [1]. Malignancy is reported in < 2% of cases. Even though a clear histopathogenesis of the tumor remains challenging, recent authors [2–4] suggest a derivation from Schwan cells. A clear positivity to S100 and CD68, PAS reactivity and diastase-resistance [5], a lack of immunoreactivity for Ki-67 [6], and a low p53 activity [2] makes a GCT diagnosis plausible.

GCT can occur at any age [7], and in virtually every organ of the body [8] but the soft tissues of the trunk and the tongue are reportedly the most common locations [9]. In the gastrointestinal tract, GCTs are seldom and most commonly found in the esophagus, followed by the colon [10,11]. Most are identified unexpectedly during endoscopic studies [12]. Due to their rare occurrence and only few reports in the medical literature, imaging characteristics for GCTs in the gastrointestinal tract are not definite [4,13]. Similarly, pancreatic GCTs are exceedingly rare with only 8 reported cases thus far [13]. Tumors presented as well-defined, hypovascular masses possibly causing duct obstruction, mimicking a hypovascular pancreatic endocrine tumor [14].

Herein we report, to the best of our knowledge, the first case of pancreatic and cecal GCTs in the radiologic literature as well as their synchronic presentation. Familiarity with this entity and its imaging appearances may help recognize GCT as a possible differential diagnostic consideration in a patient with a solid well-defined pancreatic mass, especially if similar lesions are present in other organ systems.

2. Case report

A 43-year-old Caucasian man presented to our institution for surgical consultation for management of a presumed granular cell tumor involving the right colon. His medical history was notable for seven months of heartburn, chest pain, increasing stooling and one episode of hematochezia along with an unintended 10 pounds weight loss. Optical colonoscopy with biopsy and staging CT were obtained at the outside institution prior to transfer. The abdominal CT scan with oral contrast demonstrated a homogeneous 1.5 cm intraluminal cecal lesion with well-defined borders, but also segmental pancreatic ductal dilation and relative atrophy in the distal body and tail of the pancreas. No clear pancreatic mass was observed (Fig. 1).

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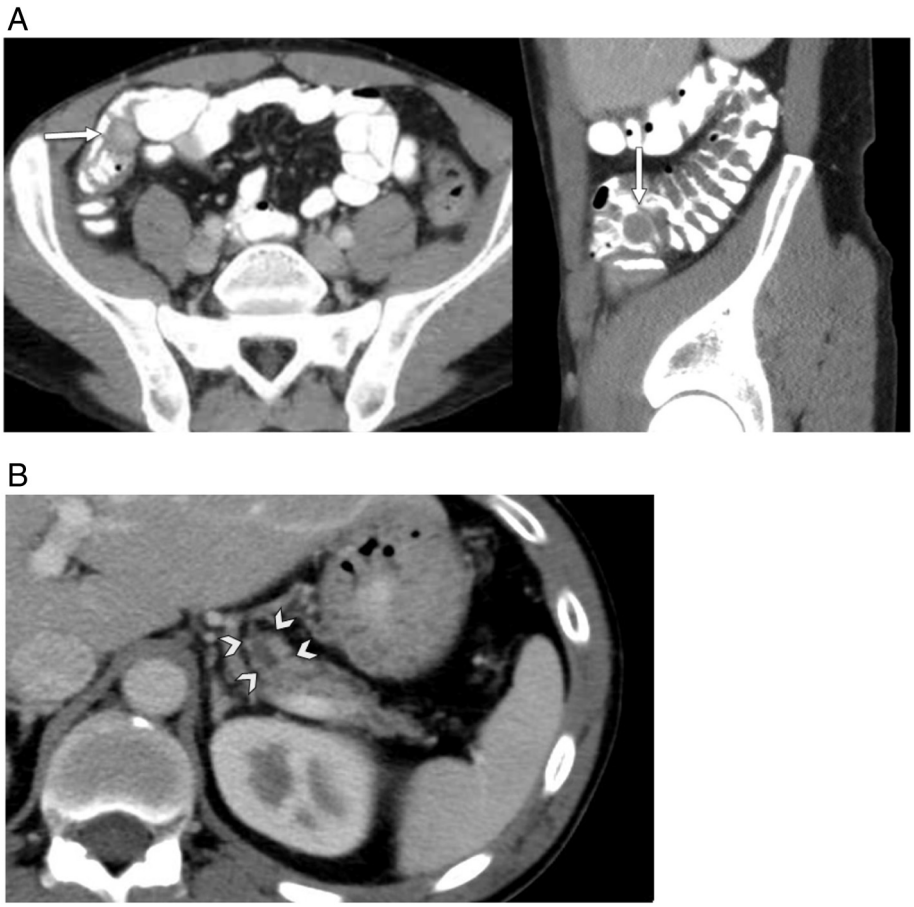

Fig. 1. Contrast-enhanced computed tomography of the abdomen and pelvis.
A. Axial and sagittal images show a 1.5 cm well-defined homogeneous intraluminal mass (white arrows) in the caecum next to the ileo-cecal valve.
B. Axial image shows pancreatic ductal dilation in the body and tail with relative atrophy of the parenchyma (white arrowheads).

Pathological evaluation of the cecal lesion, following endoscopic biopsy, revealed a tumor positive for periodic acid Schiff (PAS), periodic acid Schiff diastase (PAS-D) and S-100 without prominent atypia or necrosis; findings were consistent with a granular cell tumor (GCT).

A subsequent MRCP examination was performed at our institution to evaluate the cause of dilation of the main pancreatic duct (MPD). Both intravenous (7 mL of Gadavist) and oral contrast (1 cc of Gadavist mixed with 50 cc of water) were administered. The examination was notable for segmental dilation of the MPD up to 5 mm within the pancreatic tail with irregularity and abrupt non visualization of the MPD at the body-tail junction. Associated parenchymal tail atrophy was found, along with loss of intrinsic T1 hyperintensity, due to chronic inflammation and duct obstruction. A small pancreatic body enhancing focus was noted on delayed post-gadolinium fat-suppressed T1-weighted images (Fig. 2). On DWI, no restriction was observed. A presumed diagnosis of a small pancreatic endocrine tumor was rendered.

A PET scan after the administration of Ga-68 labeled dotatate yielded physiologic uptake throughout the body without pathological uptake in the pancreatic body or tail. Finally, a preoperative multiphasic CTA, 11 months following the initial presentation, showed mild atrophy of the distal pancreas with duct dilation and a 1.6 cm slightly hypervascular pancreatic mass located in the proximal pancreatic body; no local vascular invasion was evidenced (Fig. 3).

An intraoperative ultrasound of the pancreas was performed and showed a well-defined sub-centimeter hypoechoic nodule with a dilated MDP (Fig. 4). The patient then underwent a combined robotic-assisted minimally invasive distal pancreatectomy, splenectomy and right hemicolectomy. The postoperative course was uneventful. By immunohistochemistry, both masses were positive for S100 and CD68 as well as PAS-D positive. Tumor proteins p53 and Ki67 showed weak, patchy positivity (wild-type) and a low proliferative index (< 5%) respectively. No significant mitotic activity or necrosis was seen (Fig. 5). The findings were consistent with a multi-centric synchronous granular cell tumor involving both the pancreas and the cecum.

3. Discussion

This case study presents the unusual presentation of a 43-year-old Caucasian man with pathological proven granular cell tumors (GCT) of both the pancreas and the cecum. The patient was referred to our institution with a colonoscopic biopsy that revealed an uncommon histologic diagnosis in the cecum near the ileo-cecal valve. Additionally, MDCT showed a 1.6 cm slightly hypervascular pancreatic body mass. On MRI, the pancreatic mass presented as a small focus of enhancement and duct obstruction. Pathological diagnosis was confirmed by surgical excision. Both tumors stained positive to CD68 and S100 with positive PAS-D staining.

Granular cell tumors (GCT) are rare neoplasms of neuro-ectodermal nature with a very low malignant potential [9]. GCT were first described by Abrikossoff in 1926 [15] as a tumor of muscular origin, hence its original name of “myoblastoma” [4,16]. A prior study by Vered et al., suggests that GCT might be a lesion that echoes metabolic imbalances or inflammatory reactive changes rather than being a true de novo neoplasm [17]. However, current immunohistochemistry data suggests a clear neuronal derivation [4]. Cells in GCTs indeed show positivity in staining for S100 [6] which is a particular acidic protein that exists in peripheral Schwann cells and satellite cells of ganglia [13]. These immunological findings are congruent with our patient's pathological findings of S100 positivity in both his cecal and pancreatic masses.

GCTs typically present as painless and circumscribed nodules