Case Report

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Xanthogranulomatous pancreatitis combined with multifocal necrosis: A case report and literature review

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Abstract

Xanthogranulomatous inflammation is rare and occurs in several viscera in the body, such as the kidney, gallbladder, bone, and stomach. The pathogenesis of Xanthogranulomatous pancreatitis (XGP) is not well understood, and the disease is characterized by lipid-laden histiocytes deposited at various sites in the organs. XGP associated with multifocal necrosis is extremely rare. In this report, we described a case of XGP associated with multifocal necrosis and included a review of the literature.

Keywords: Pancreas, Xanthogranulomatous, Inflammation

Xanthogranulomatous inflammation is rare circumstance in which lipid-laden histiocytes are deposited at various sites in the organs. Xanthogranulomatous pancreatitis (XGP) combined with multifocal necrosis is extremely rare. We report a case of XGP with multifocal necrosis. To our knowledge, 2 cases in China have been published in Chinese journals. This is the first reported case in China in the English-language literature.

Case presentation

A 76-year-old man was referred to our hospital (Huzhou First People's Hospital, Huzhou, China) for further examination of persistent upper abdominal pain associated with vomiting of stomach contents after eating that lasted for 2 weeks. The patient reported that his weight was unchanged over the past 3 months. He had upper abdomen tenderness through physical examination. There was no history of fever, chills, or chest pain. His medical and surgical history was unremarkable except for hypertension without controlled. His social history was unremarkable. His carbohydrate antigen 125 (CA125) was elevated to 40.5 U/mL, serum lipase was elevated to 148.6 UL, and serum amylase was 100 UL (high normal). Total protein was reduced to 64.8 g/L, and albumin was reduced to 36.9 g/L. Other laboratory data, including serum carcinoembryonic antigen, carbohydrate antigen 19-9, CA153, C-reactive protein, bilirubin, and leukocytes, were normal. Abdomen 2-dimensional ultrasound revealed a 75 mm × 60 mm homogeneous hypoechoic mass in the uncinate process of the pancreas (**Fig. 1A**). Double contrast-enhanced ultrasound (DCEUS) with oral contrast-enhanced ultrasound agents and intravenous contrast-enhanced ultrasound (CEUS) agents were given to the patient at the same time. The peripheral mass began to undergo rapid high enhancement at 20 s before the pancreas parenchyma (**Fig. 1B**). Microbubbles in the mass began to wash out at 36 s (**Fig. 1C**). Part of the mass did not enhance over time. The size of the mass was calculated as 8.5 cm × 6.7 cm after enhancement. The duodenum wall was thick around the mass, enhanced and washed out at the same time as the distal wall. There were lymph nodes above 3 around the mass. Based on our experience and the rich microvasculature revealed by CEUS imaging, we inferred that what occurred at the duodenal wall was similar to edema and not a malignant neoplasm infilitration; however, the properties of the mass were uncertain. An abdominal computed tomography (CT) scan demonstrated that the lesion was strongly associated with liquefaction in the pancreatic parenchyma and distinction from the duodenum (**Fig. 2A**). Contrast-enhanced CT revealed that the mass had irregular ring enhancement and was likely a malignant tumor (**Figs. 2B, C**).

The mass was obtained by biopsy through ultrasonic guidance, and the pathology result was pancreatic parenchyma chronic inflammation associated with fibrous tissue hyperplasia and multifocal necrosis (**Fig. 3A**). Whipple's operation (pancreaticoduodenectomy) was performed, and a yellow lump lesion was found to be 8.5 × 6.0 cm in size (**Fig. 3B**). On gross examination, the mass had an unclear border and adhered to the partial transverse mesocolon, greater omentum, and duodenal mesocolon. Microscopically, the lesion was composed of an aggregation of several foam cells, lymphocytes, and multinucleated giant cells as well as necrosis (**Fig. 3C**). The final diagnosis of XGP associated with multifocal necrosis was made. On postoperative day 20, the hemodynamics was stabilized, no gastrointestinal hemorrhage, and the patient was extubated. The patient was subsequently discharged. After 6 months, the patient did

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This case has reported work in accordance with SCARE 2018 standards.

This manuscript has been peer reviewed.

Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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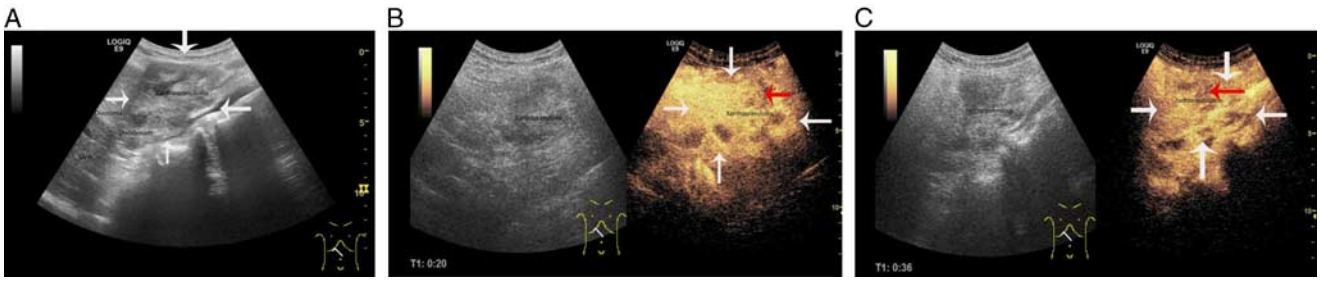

Figure 1. Images of the pancreatic lesions. A, Gray-scale US: the mass was homogeneously hypoechoic in the uncinate process of the pancreas (arrows) and closely related to the duodenum; the border was unclear. B, Enhancement phase (20 s): enhancement started from the edge of the mass (white arrow). C, Regression phase (36 s): the mass began to wash out (white arrows), and the area was not enhanced at all times (red arrow).

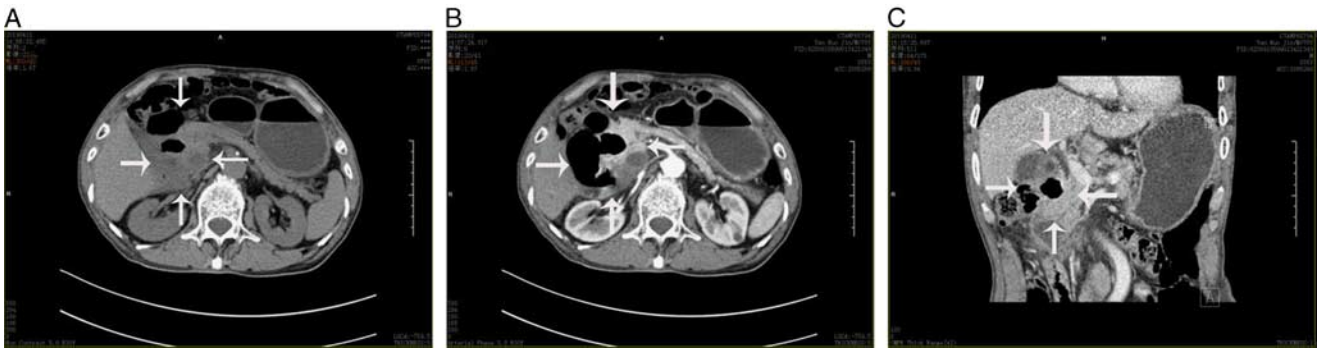

Figure 2. A, Computed tomography (CT) plain scan: solid mass associated with liquefaction in the pancreatic parenchyma. B, Contrast-enhanced CT (cross section of abdomen): the mass was a ring of irregular enhancement, and the cyst area had not enhanced. C, Contrast-enhanced CT (sagittal position): the mass was closely related to the duodenum. The arrows are CT manifestations of the mass.

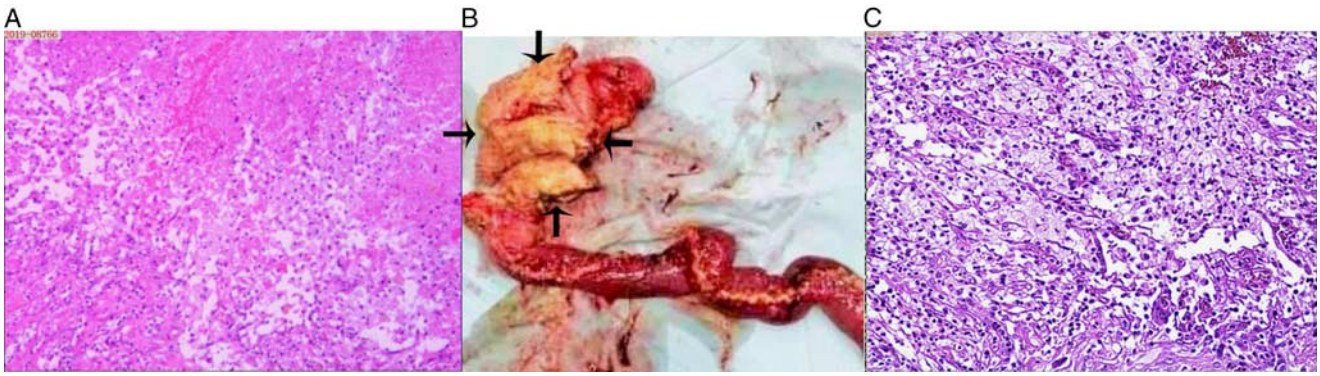

Figure 3. A, Light microscopic examination: showing lymphocytes, fibrous tissue hyperplasia and multifocal necrosis [hematoxylin and eosin (HE) staining; magnification, × 100]. B, Pathologic gross specimens: a yellow nodular lesion (arrows) was found, with adherence to duodenum. C, Light microscopic examination for gross specimens: the mass contained an aggregation of foamy histiocytes, lymphocytes, and plasma cell granulocyte cells (HE staining; magnification, × 100).

Table 1

Imaging and clinical features of Xanthogranulomatous pancreatitis.

Case First/ References	Sex/ Age (y)	Symptom	Country	Site	SIR	Tumor markers	US	CT	MRI	Preoperative diagnosis	Appearance	Operation	Tumor size (cm)
Uneo et al[5]	42/M	Abdominal pain	American	Body	CRP +	–	Homogeneous echotexture	Hypoattenuating	↓T1,↑T2	Pseudo cyst	Cystic	DP	8 × 5
Iyer et al[6]	50/M	Jaundice	India	Head	NA	NA	NA	NA	NA	Malignancy	Nodular	Mass excision	3
Iyer et al[6]	36/M	Stone	India	Tail	NA	NA	NA	NA	NA	Malignancy	Massive	DP + S + PG	NA
Kamitani et al[7]	82/M	Epigastric pain	Japan	Body	–	–	NA	Ill-defined enhancing	↓T1,↑T2	IPMT	Cystic	DP + S + PG	3 cm
Okabayshi et al[12]	60/M	Abdominal pain and pancreatic cystic mass	Japan	Tail	CRP +	–	NA	Hypoattenuating	NA	Pseudo cyst	Cystic	DP	NA
Okabayshi et al[12]	69/M	Abdominal pain	Japan	Tail	CRP +	–	NA	Hypoattenuating	NA	Pseudo cyst	Cystic	DP	NA
Shima et al[13]	66/M	Epigastric pain	Japan	Body	CRP +	CA199 +	NA	Hypoattenuating	Isointense T1, ↑T2	Malignancy	Solid	DP	4
Iso et al[9]	82/M	Weight loss	Japan	Tail	CRP +	–	Heterogeneous echotexture	Hypoattenuating	NA	IPMC	Nodular	DP	NA
Ikeura et al[14]	73/M	Abdominal pain	Japan	Body	–	–	Heterogeneous (mostly solid, partially cystic)	Hypoattenuating	Mildly ↑T1	SPPT	Cystic	PPPD	4 × 4
Uguz et al[15]	30/M	Abdominal pain	Turkey	Head	–	–	NA	NA	NA	Chronic pancreatitis	NA	DPP	NA
Uguz et al[15]	34/F	Abdominal pain	Turkey	Head	–	–	NA	NA	NA	Chronic pancreatitis	NA	DPP	NA
Kim et al[16]	72/F	Abdominal pain	Korea	Body	–	–	NA	NA	Subtle, focal↓T1	IPMT	Cystic	PPPD	1.5 × 1
Kim et al[4]	70/F	Abdominal pain	Korea	Head	CRP +	–	NA	Hypoattenuating	Isointense T1, ↑T2	Malignancy	Solid	WPD	2.2 × 2.0
Nishmimura et al[17]	76/M	Abdominal pain, Weight loss	Japan	Body	White blood cell +	–	NA	Cystic lesion, thick wall	↓T1,↑T2	IPMC	Cystic	DP + S	5.2 × 2.6
Atreyapurapu et al[18]	60/M	Abdominal pain	India	Head	Normal	Normal	NA	Heterogeneously	NA	Malignancy	Solid	WPD	NA
Hanna et al[19]	50/F	Abdominal pain and nausea	UK	Body/ tail	CRP +	CA199 + , CEA +	NA	Hypoattenuating	NA	Malignancy	Cystic	DP	14 × 14 × 14.5
Kwon[20]	60/M	Pancreas MASS	Korea	Body	–	CEA +	NA	Low-attenuated	↓T1, ↑T2	uncertain	Solid	DP	1.6 × 1.5
Navarro et al[10]	86/F	Epigastric pain and weight loss	Spanish	Head	–	–	Heterogeneous	Hypermetabolic nodule	N/A	Malignancy	Solid	conservative	NA
Becker et al[21]	56/M	Prostate adenocarcinoma	American	Tail	–	CEA +	Heterogeneously hypoechoic	Hypoattenuating	↓T1, ↑T2	IPMN	Cystic	DP + S	2.8 × 2.5
Gaur et al[22]	58/M	Abdominal pain and weight loss	India	Head	CRP +	CA199 + , CEA +	Solid cystic lesion	Heterogeneously enhancing	NA	Malignancy	Solid and cystic	WPD	6.1 × 5.2 × 4.0
Present case	76/M	Abdominal pain, vomiting	China	Head	–	CA125 +	Mostly solid, partially cystic; ringed enhanced	Irregular enhancing	NA	Malignancy	Solid	WPD + PG	8.5 × 6.0

DP indicates distal pancreatectomy; DPP, duodenum-preserving pancreatectomy; NA, not applicable; PG, partial gastrectomy; PPPD, pylorus preserving pancreatoduodenectomy; S, splenectomy; SIR, systemic inflammatory response (high fever, elevated white blood cell count, or elevated C-reactive protein); WPD, whipple pancreaticoduodenectomy.

not experience any recurrence. This case is reported in accordance with the SCARE 2018 standard[1].

Discussion

Xanthogranulomatous changes are considered a rare chronic inflammation characterized by an aggregation of foamy histiocytes and inflammatory cells. The lesion has been reported in various cases in the gallbladder and kidney[2]. According to one proposed etiology for xanthogranulomatous cholecystitis, bile enters the gallbladder wall and is engulfed by macrophages, resulting in a chronic granulomatous inflammatory response[3]. The etiology of XGP is not entirely clear but likely results from a combination of duct obstruction, infection, and repeated hemorrhage, as is seen in the kidney and gallbladder[4–7]. Xanthogranulomatous lesions in the pancreas are a benign condition. Kwon et al[8] concluded the lesion was a progressive enhancement pattern in contrast-enhanced CT and MRI with 10 cases. PET/CT examinations showed focal hypermetabolism, which was one of the diagnostic impressions of pancreatic cancer on PET/CT[8]. XGP manifests neoplasm-like characteristics, such as being poorly defined with local tissue invasion and destruction[7,9]. It is difficult to discriminate this condition from pancreatic cancer with clinical and radiologic results[9]. As a result, most cases were overtreated with surgical resection, and there have been only 2 reported cases of conservative management following a non-operative diagnosis[10,11].

Twenty-one cases had been reported in English literature before 2019.A review of the existing literature was performed (**Table 1**). All cases were mostly from Asian countries, especially Japan (15/21). The patient age was 61.3 years (range, 30–86 y), and most were men (16/21). They were referred to the hospital for epigastric pain (16/21), accompanied by weight loss in 4 cases. Some patients with an inflammatory response had an elevated white blood cell count or elevated C-reactive protein (9/19). The lesions appeared hypointense on T1-weighted images and hyperintense on T2-weighted images (5/9) in which MRI examination was performed. Contrast-enhanced CT showed that the lesion was hypoattenuating (10/16). The result of 2-dimensional ultrasound was homogeneous echotexture (5/7), and most cases had no CEUS examination performed. The preoperative diagnosis in 15 cases (15/21) was malignancy. Of all cases, 2 cases underwent conservation treatment, and the others (included ours) were treated with surgery. In our case, the lesion was large because the patient had not been examined previously. The mass was solid and was associated with liquefaction in the pancreatic parenchyma and a ring of irregular enhancement with an indefinite border on CT scan in our case. DCEUS showed that the lesion was enhanced before the pancreatic parenchyma, and part of the lesion had no enhancement at all times; thus, we thought it was necrotic. It was also noteworthy that the duodenal wall manifested as inflammatory edema and not tumor infiltration. Based on the above information, we concluded that a malignant lesion could not be excluded. However, we did not perform other imaging, such as MRI and PET scans, as we had shunted the lesion. To alleviate the severe symptoms of the patient, we decide to explore abdominal cavity. During the exploration, we found a solid mass with a diameter of about 8 cm at the level of the uncinate process of the pancreas, the boundary of which was unclear, and part of the transverse mesocolon, omentum, and duodenum were involved and densely adhered, so we performed a Whipple's operation.

Conclusions

Although XGP associated with multifocal necrosis is rare, it is important for surgeons, pathologists and radiologists to recognize this uncommon entity, as the lesion may manifest clinical features that mimic those of pancreatic tumors.

Ethical approval

Written informed consent for publication was obtained from the patient.

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None.

Author contribution

X.T. and W.L. contributed the same to this article. They contributed to write the manuscript, to gather the clinical data as well as the relevant imaging information. All authors read and approved the final manuscript.

Conflict of interest disclosure

The authors declare that they have no financial conflict of interest with regard to the content of this report.

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