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**Columns:** **CASE REPORT**

**Pancreatic mass as an initial manifestation of polyarteritis nodosa: A case report and review of the literature**

Yokoi Y *et al.*Pancreatic mass associated with polyarteritis nodosa

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**Abstract**

Classic polyarteritis nodosa (PAN) that targets medium-sized muscular arteries- and microscopic polyangiitis (MPA) which is characterized by inflammation of small-caliber vessels and the presence of circulating myeloperoxidase anti-neutrophil cytoplasmic antibodies (MPO-ANCA), are distinct clinicopathological entities of systemic vasculitis. A 66-year-old woman presented with fever, cholestasis, and positive MPO-ANCA. Radiologic examination showed a pancreatic mass compressing the bile duct. Therefore, we performed pancreatoduodenectomy. Histopathologic examination revealed that necrotizing vasculitis predominantly affecting the medium-sized vessels spared arterioles or capillaries in the pancreas, a finding consistent with PAN. Unexpectedly, renal biopsy revealed small-caliber vasculitis and glomerulonephritis, supporting MPA. The initial manifestation of a pancreatic mass associated with vasculitis has been reported in only 7 articles. Its diagnosis is challenging, because no reliable clinico-radiologic findings have been observed. Clinicians should be aware of such cases, and early diagnosis followed by immunosuppression is mandatory. Our findings may reflect a polyangiitis overlap syndrome coexisting between pancreatic PAN and renal MPA.

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**Key words:** Classic polyarteritis nodosa; Myeloperoxidase anti-neutrophil cytoplasmic antibodies; Microscopic polyangiitis; Pancreatic mass; Polyangiitis overlap syndrome

**Core tip:** A 66-year-old woman presented with a pancreatic mass accompanied by fever, cholestasis, and positive myeloperoxidase anti-neutrophil cytoplasmic antibodies. The resected pancreas showed extensive fibrosis associated with necrotizing vasculitis targeting medium-sized vessels, but spared small-caliber vessels, a finding compatible with polyarteritis nodosa. Unexpectedly, renal biopsy revealed small-caliber vasculitis and glomerulonephritis, supporting microscopic polyangiitis. The initial manifestation of a pancreatic mass associated with vasculitis has been reported in only 7 articles. Although rare, vasculitis should be included in differential diagnosis for pancreatic masses. Additionally, our findings may reflect a polyangiitis overlap syndrome coexisting between pancreatic polyarteritis nodosa and renal microscopic polyangiitis.

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**INTRODUCTION**

Systemic vasculitis is characterized by a variety of clinical manifestations and courses, depending upon the organ involved. Among the classifications for systemic vasculitis, the Chapel Hill consensus conference (CHCC) nomenclature is widely accepted[1,2]. Vasculitis affecting small-caliber blood vessels (arterioles, venules, or capillaries) often accompanies anti-neutrophil cytoplasmic antibodies which are postulated to play a major pathologic role in developing necrotizing vasculitis[3]. Such ANCA-associated vasculitis includes the following 3 clinicopathologic variants: microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis[2]. Among them, MPA is characterized by non-granulomatous inflammation, few or no immune deposits (pauci-immune), glomerulonephritis, and presence of myeloperoxidase (MPO)-ANCA[2].

Another category of vasculitis, classic polyarteritis nodosa (PAN), targets small- and medium-sized muscular arteries, spares small-caliber vessels, and causes diffuse vascular inflammation, ischemia, or rupture of affected organs[4]. Although PAN frequently complicates the skin, joints, kidneys, gastrointestinal systems, and kidneys, initial and symptomatic involvement of the pancreato-biliary systems has only been reported in rare cases[5-8].

Herein, we report a patient presenting with fever, cholestasis, and a pancreatic mass compressing the bile duct as a clinical feature of PAN.

**CASE REPORT**

A 66-year-old woman presented with a 2-week history of intermittent high-grade fever (approximately 39°C). She did not report arthralgia, myalgia, or abdominal symptoms. Approximately 1 month before admission, she underwent tympanotomy for left otitis media. Her medical history was noncontributory. She denied alcohol and drug use. Laboratory examination showed elevated biliary enzyme levels including alkaline phosphatase level of 717 U/L (115– 359 U/L) and gamma glutamyl transpeptidase levels of 238 U/L (10- 47 U/L), and C-reactive protein (CRP) levels of 8.30 mg/dL (< 0.30 mg/dL). Serum levels of amylase, aspartate aminotransferase, alanine aminotransferase, blood urea nitrogen, creatinine, carcinoembryonic antigen, carbohydrate antigen 19-9, and procalcitonin were normal. The levels of glycated hemoglobin was slightly elevated. Leukocytosis and eosinophilia were not present. Immunological data showed slight elevations of IgG [1902 mg/dL (820-1740 mg/dL)] and IgA [628 mg/dL (90- 400 mg/dL)], but IgM and IgG4 levels were normal. Autoimmune investigations showed elevated MPO-ANCA levels [473 IU/mL (< 3.5 IU/mL)], in addition to a slight elevation of anti-nuclear antibodies (1:64) and rheumatoid factor. Proteinase 3- ANCA, serum hepatitis B surface antigen, and hepatitis C virus antibodies were not detected. No bacteria grew on blood culture. Urinalysis revealed proteinuria (2+) and hematuria (2+) with hyaline casts.

A hypo-echoic 2.0-cm mass was observed in the pancreatic head on an abdominal ultrasonogram (Figure 1A). The corresponding lesion was an ill-delineated hypodense mass with poor enhancement, as observed by using a CT scan, and it compressed the distal common bile duct (CBD) and pancreatic duct (PD) (Figures 1B-E). The walls of the gallbladder and bile duct were thickened (Figures 1D and E). A chest CT scan showed slight changes, including bronchial dilation and peripheral inflammation with a centrilobular distribution. Angiographic reconstruction by using a CT scan showed normal visceral arteries of the superior mesenteric artery (SMA) and celiac systems. Vascular stenosis or aneurysms were not detectable. Endscopic retrograde cholangiopancretography (ERCP) demonstrated a double duct sign with compression of the distal CBD and tortuous dilation of the PD (Figure 1F). Bile cytology and culture were negative, according to the results obtained after using the sample via naso-biliary drainage.

We could not exclude the possibility of pancreatic cancer as a cause of the patient’s fever, and therefore we performed pylorus-preserving pancreatoduodenectomy. The pancreatic mass was soft on palpation and did not invade the adjacent tissues. Intraoperative ultrasonography revealed an ill-defined pancreatic mass with low echogenicity. The postoperative course was uneventful, and the patient’s fever completely resolved with a reduction of CRP levels.

In the resected pancreas, the focal stenosis in the CBD was approximately 2 cm distal to the ampulla of Vater. There was marked fibrosis adjacent to the intrapancreatic CBD and PD (Figure 2A). The affected small- to medium-sized arteries in the fibrosis were characterized by necrotizing arteritis with subintimal fibrinoid necrosis, disruption of the elastic laminae, perivascular fibrosis, and inflammatory cell infiltration (Figures 2A and B). Vessel occlusion or thrombus was also observed (Figure 2A). Small-caliber vessels such as the arterioles, capillaries, or venules were spared. Granulomatous inflammation and significant eosinophil infiltration were not found. The fibrotic lesion extended longitudinally towards the hepatic hilus along the bile duct. Necrotizing vasculitis was also observed in the walls of the proximal bile duct and gallbladder, but their mucosal layers were well preserved (Figure 2C). The duodenum also showed arterial changes. These vascular changes were compatible with classical PAN.

To confirm systemic vasculitis, a renal needle biopsy was performed. Global sclerosis affected 20% of the glomeruli whereas cellular crescent was observed in 10 % (Figure 2D). Interstitial fibrosis was observed in the tubule-interstitial area. Both active- and healed-stages of vasculitis were observed in the small arteries and capillaries (Figure 2E). Immune complexes were not detectable. These pathological findings were compatible with the renal changes of MPA, according to the CHCC nomenclature[1,2].

Therapy with prednisone and cyclophosphamide was undertaken to induce remission of the systemic vasculitis. The patient has remained asymptomatic 6 months after the operation.

**DISCUSSION**

In the present patient, a pancreatic mass accompanied by fever and cholestasis was observed; surgical removal successfully improved the patient’s clinical symptoms and data. Pathological study demonstrated extensive vascular injury in the pancreas, bile duct, gallbladder, and duodenum. The affected vessels were small- and medium-sized arteries, and arterioles and capillaries were spared, which is a finding consistent with classic PAN[1,2,4]. Initial clinical manifestation of vasculitis in the pancreato-biliary system is uncommon, with only a few reports documenting pancreatitis or cholecystitis[7]. Other forms of pancreatic vasculitis including mass formation are extremely rare. The articles reporting pancreatic mass associated with vasculitis were collected through a literature search with the words “vasculitis,” “pancreas,” “tumor,” or “mass” in their title. Among them, 7 articles providing radiologic and histopathologic descriptions were reviewed (Table 1). Including our case, there were 3 PAN[5,6], 3 GPA[8-10], and 2 localized PAN[11,12]. The former 2 were major vasculitis presenting a tumor-like lesion in the uro-genital system and breast or kidney, respectively[6]. The median age was 62 years (range, 44- 66 years), with a male predominance (5:3 ratio).Three patients were Japanese, 2 were white, and 1 was Jewish. The symptoms were various and nonspecific, including abdominal pain (5 patients), fever (3 patients), otitis media (2 patients), and jaundice (1 patient). All lesions were 2- 3 cm in diameter, and were localized in the head (6 patients), neck (1 patient) and both body and tail of the pancreas (1 patient). The gallbladder was also affected in 2 PAN patients. Among 4 cases analyzed, ANCA was positive in 3 (GPA, 2; PAN, 1). Use of glucocorticoids and cytotoxic agent was effective in all cases, if treated; otherwise, rapid deterioration of necrotizing vasculitis was fatal as shown in case 5. These findings indicate that early introduction of immunosuppressive treatment based on accurate diagnosis is crucial for better outcome.

One of the obstacles in treatment strategy for vasculitis-induced pancreatic mass is the difficulty in diagnosing it. Our review showed that 7 of 8 patients were diagnosed only after surgery or autopsy (Table 1). Besides neoplasm, the pancreatic mass can encompass a variety of diseases, such as an inflammatory pseudotumor (IPT). IPT includes autoimmune pancreatitis, groove pancreatitis, and lipomatosis[13]. As shown in Table 1, regardless of different types of vasculitis, vasculitis-associated masses were hypoechoic and were hypodense with poor encasement on a CT scan, making it difficult for differentiation from pancreatic cancer or IPT. For a focal pancreatic lesion, fine-needle biopsy is widely used with abdominal or endoscopic ultrasonography, and it is useful in autoimmune pancreatitis[14]. However fine-needle biopsy has potential sampling error problems; indeed, ultrasound- or CT-guided needle biopsy failed to be diagnostic for pancreatic GPA (case 2) and PAN (cases 3 and 4). Negative findings do not exclude the possibility of malignancy, and there is a risk of needle tract seeding or dissemination of tumor cells[15]. Thus, the diagnostic procedure is challenging. Some clinicians do away with the preoperative evaluation in patients with operable focal lesions of a clinically and radiologically suspicious malignancy. The common use of ANCA tests in the future would enhance preoperative diagnosis and avoid unnecessary radical operations.

Another interesting finding in this case was the coexistence of different entities of vasculitis, such as PAN in the pancreato-biliary system and MPA in the kidneys. The renal histopathologic findings of small-caliber vessel (arteries and capillaries) vasculitis and positive MPO-ANCA supported the MPA diagnosis[2,3]. PAN and MPA had often been diagnosed together until proposal of the CHCC nomenclature and distinguishing between these 2 entities is not clinically always straightforward[16]. Our case may represent the so-called polyangiitis overlap syndrome which is characterized by systemic vasculitis with features that overlap more than 1 type of vasculitis[17]. Alternatively, it is possibly a coincidence or part of the MAP or PAN spectrum. Renal MAP has been reported to complicate vasculitic disorders that can be attributed to PAN, such as a rupture of branch of the celiac[18] or SMA system[19], and coronary angiitis[20].

In conclusion, we encountered a patient with a pancreatic mass associated with PAN. A literature review revealed that pancreatic masses have been reported in 7 patients with primary vasculitis. Because of its rarity and lack of reliable discrimination from pancreatic cancer, clinicians should be aware of such cases, and that early diagnosis followed by immunosuppressive treatment is mandatory.

**COMMENTS**

***Case characteristics***

A 66-year-old woman presented with a pancreatic mass accompanied by fever.

***Differential diagnosis***

An inflammatory pseudotumor and pancreatic neoplasms including cancer.

***Laboratory diagnosis***

Laboratory examination showed elevated levels of biliary enzymes (alkaline phosphatase and gamma glutamyl transpeptidase), C-reactive protein, and myeloperoxidase-anti nuclear cytoplasmic antibodies.

***Imaging diagnosis***

An abdominal CT revealed an ill- delineated 2.0 cm pancreatic mass with poor enhancement compressing the distal common bile duct (CBD) and pancreatic duct, as well as the thickened walls of the CBD and gallbladder.

***Pathological diagnosis***

The resected pancreas revealed extensive fibrosis associated with necrotizing vasculitis targeting medium-sized vessels spared small-caliber vessels.

***Treatment***

The patient underwent surgical resection, followed by immunosuppression after pathological diagnosis of polyarteritis nodosa.

***Related reports***

A pancreatic mass as an initial manifestation of vasculitis is extremely rare, with only 7 cases reported in the literature.

***Experiences and lessons***

The case emphasizes that vasculitis should be included in differential diagnosis of a pancreatic mass accompanied by fever.

***Peer review***

Although the immunosuppression is the optimal treatment for vasculitis-associated pancreatic tumor, the diagnosis is challenging because of its rarity and lack of discrimination from pancreatic cancer.

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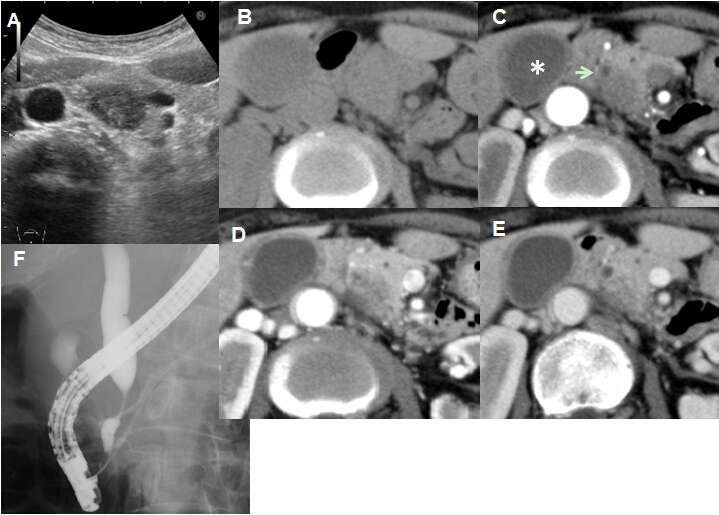
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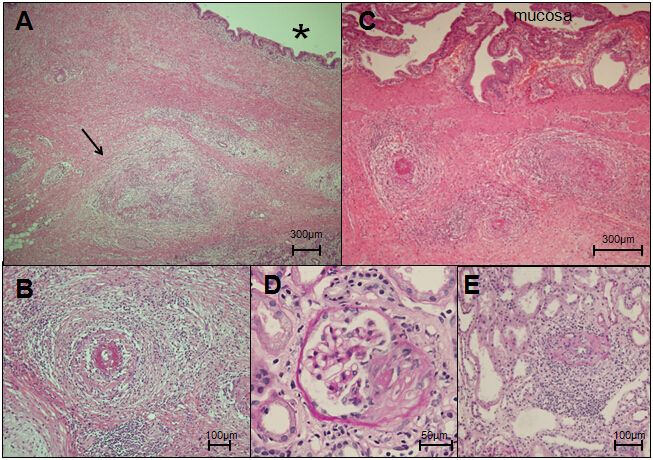
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**Figure 1** **Preoperative images.** A: An ultrasonogram showing a slightly ill-defined hypoechoic mass at the pancreatic head; B- E: Abdominal CT scans. B: The pancreatic mass is slightly hypodense on simple CT; C: Enhanced CT shows a non-enhancing mass (arrow) localized adjacent to the bile duct at the arterial phase; D and E: The mass has sporadic enhancement at later phases. Note that the walls of the gallbladder (asterisk) and bile duct (arrowhead) are thickened; F: A Cholangiogram showing a tapered distal biliary stricture consistent with extrinsic compression by the pancreatic mass.



**Figure 2** **Histological findings of the pancreas, gallbladder, and kidney.** A: Pancreatic fibrotic changes adjacent to the bile duct (asterisk) are evident. An obliterated medium-sized artery (arrow) is accompanied by cellular infiltration and destruction of the wall. The bile duct mucosa is intact (hematoxylin-eosin stain; original magnification, × 40); B: The affected small-sized artery in the pancreatic fibrosis is characterized by necrotizing arteritis with subintimal fibrinoid necrosis and inflammatory cell infiltration (hematoxylin-eosin stain; original magnification, × 100); C: The arteries of the gallbladder are also involved: the mucosa is preserved (hematoxylin-eosin stain; original magnification, × 40); D and E: Renal biopsy showed segmental sclerosis and collapse with fibrocellular crescent (D: Periodic acid –Shiff’s stain, original magnification, × 200) and vasculitis of a small-sized artery (E: Periodic acid –Shiff’s stain, original magnification, × 100 ).



**Table 1 Reported cases of pancreatic tumor associated with vasculitis**

|  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Ref.** | | **Age/sex/**  **Race etc.** | **Final**  **diagnosis** | **Symptoms** | **Sites**  **involved** | **Prior diagnosis** | **Tumor size**  **Imaging findings** | **Diagnostic criteria** | **Outcome** | |
| **Pancreas** | **Patient** |
| Ito *et al*[11] | 44/M/  Japanese | Localized PAN | Epigastralgia | Head | No | ND  ERCP: CBD stenosis | ND | Underwent PD | Discharged |
| O'Neil *et al*[8] | 62/M /  White | GPA | Jaundice  Otitis media  Nasal ulceration | Head  Gallbladder | No | 3 cm  CT: mass  US: hypoechoic  ERCP: CBD stenosis | ANCA (+)  Needle biopsy: non diagnostic  Renal biosy: confirmed | Improved on CYC + CS | Improved on  CYC +CS |
| Damani *et al*[5] | 46/F/  ND | PAN | Right upper abdominal pain | Neck | No | 2 cm  US: hypoechoic  CT: low attenuation, nonenhancing mass | Needle biopsy: non diagnostic  Postoperative histopathology | Cholecystectomy  Distal Px | Died (20 d)  Various complication |
| Kariv *et al*[6] | 65/ M/  Jewish | PAN | Epigastralgia  Weight loss  Low grade fever | Head | No | 3 cm  CT: mass | Needle biopsy: chronic pancreatitis | Underwent PD | Remission  on CS |
| Matsubayashi *et al*[9] | 65/M/  Japanese | GPA | Left abdominal pain  Constipation  Low grade fever  Tympanitis | Body and Tail | S/O GPA | ND  CT: Enlargement of pancreas with sporadic low density lesions | 12PR3-ANCA (+)  Autopsy | No | Died  Hemorrhagic pneumonia  Diffuse necrotizing pancreatitis |
| Tinazzi *et al*[10] | 48/F/  ND | GPA | Mid-epigastic pain | Head | No | 2 cm :  US: Hypoechoic  MRCP: Obstruction of pancreatic duct | Postoperative histopathology | Underwent PD | Improved on  CYC +CS |
| Gonzalez-Gay *et al*[12] | 75/M/  White | Localized PAN | Epigastralgia | Head | No | ND | Postoperative histopathology | Underwent PD | Discharged |
| Our case | 66 /F/  Japanese | PAN  Renal MPA | Otitis media  Fever | Head  Gallbladder  Bile duct  Duodenum | No | 2 cm  US: Hypoechoic  CT: Hypodense  Non-enhancing | MPO-ANCA(+)  Postoperative histopathology | Underwent PD | Improved on  CYC +CS  Discharged |

PAN: Polyartertis nodosa; ERCP: Endoscopic retrograde cholangio-pancreatography; CBD: Common bile duct; PD: Pancreatoduodenectomy; GPA: Granulomatosis with polyangiitis; US: Ultrasonography; ANCA: Anti-neutrophil cytoplasmic antibody; CYC: Cyclophosphamide; CS: Corticosteroids; Px: Pancreatectomy; S/O: Suspect of; PR3: Proteinase 3; MRCP: Magnetic resonance cholangio-pancreatography; MPA: Microscopic polyangiitis; MPO: Myeloperoxidase.