Polyarteritis Nodosa-Induced Pancreaticoduodenal Artery Aneurysmal Rupture

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Abstract

Keywords

- ▶ aneurysm
- ▶ arteritis
- ▶ coil embolization
- ▶ C-reactive protein
- ▶ mesenteric artery
- ▶ vasculitis
- ▶ vessel repair

Polyarteritis nodosa (PAN) is a systemic, necrotizing vasculitis of small- and medium-sized arteries typically with multiorgan involvement. Most cases of PAN are idiopathic, although hepatitis B or C virus infections and hairy cell leukemia are important in the pathogenesis of some cases. PAN is characterized as segmental transmural inflammation of muscular arteries. Diagnosis is based on clinical suspicion, a negative immunofluorescence test for antineutrophil cytoplasmic antibodies, and whenever possible, biopsy confirmation. Angiographic images may reveal microaneurysms affecting the renal, hepatic, or mesenteric vasculature. Aneurysmal formation and rupture are important complications that can be fatal. Treatment may warrant immunosuppression with steroids and cyclophosphamide. If left untreated, PAN can be fatal. To our knowledge, we report the second documented case of PAN-induced ruptured inferior pancreaticoduodenal artery aneurysm.

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To our knowledge, we report the second documented case of PAN-induced ruptured inferior pancreaticoduodenal artery aneurysm (PDAA).

Case Report

An 86-year-old man with a history of diabetes, hypertension, aortic valve replacement, anticoagulation with warfarin, and

5 months postendovascular abdominal aortic aneurysm (AAA) repair presented with generalized and severe abdominal pain without nausea, vomiting, diarrhea, or constipation. He denied any previous history or symptomatology concerning for vasculitis, autoimmune, or connective tissue disorder. On examination, he was awake and alert. His abdomen was obese and moderately tender. His blood pressure initially was 120/60 mm Hg, and heart rate was 70 beats/min. Laboratory work-up revealed hemoglobin, 8.6 g/dL; lactate, 8.2 mmol/L; international normalized ratio, 2.8; blood urea nitrogen, 30 mg/dL; and creatinine, 1.1 mg/dL. Computed tomography (CT) of the abdomen showed retroperitoneal hematoma measuring 3.1 cm near the abdominal aorta but not directly communicating with it (▶ Fig. 1). Coronal reconstruction of the CT scan demonstrating the inferior pancreaticoduodenal artery in direct communication with the hematoma (▶ Fig. 2). He suddenly became unstable and he developed cardiopulmonary compromise with systolic blood pressure of 50 mm Hg and acute respiratory failure. Before transfer to our facility, he was managed with endotracheal intubation, fresh frozen plasma, and blood transfusion along with dopamine infusion. Emergent exploration revealed a ruptured inferior PDA with

Fig. 1 Axial computed tomography image of the abdomen showing retroperitoneal hematoma measuring 3.1 cm near the abdominal aorta but not directly communicating with it.

Fig. 2 Coronal abdominal computed tomography scan image demonstrating the inferior pancreaticoduodenal artery in direct communication with the hematoma.

several arteries converging into it, which were clipped. Histological examination revealed a muscular artery with active PAN with secondary aneurysm formation and rupture (→Figs. 3 and 4).

Furthermore, a CT angiogram of the chest, abdomen, and pelvis did not reveal other vascular abnormalities. Review of his CT scan at the time of his AAA endograft repair did not show the presence of visceral aneurysm. Further laboratory work-up revealed elevated erythrocyte sedimentation rate (ESR), 48 (< 21 mm/h); C-reactive protein (CRP), 15.3 (< 0.5 mg/dL); and serum immunoglobulin A, 490 (80–450 mg/dL). However, he had normal C- and P-ANCA as well as normal C3 and C4 levels. Antihepatitis C virus, hepatitis B surface antigen, and antihuman immunodeficiency virus (HIV) 1 and 2 antibodies were negative.

Fig. 3 Pathological specimen of pancreaticoduodenal artery showing a muscular artery with medial fibrinoid degeneration (arrow) and adventitial leukocytic infiltrate, consistent with polyarteritis nodosa.

Fig. 4 Additional area of the pancreaticoduodenal artery with prominent fibrinoid degeneration, medial disruption, and aneurysm formation.

He did well on prednisone 40 mg per day with a very slow taper plan over 6 weeks with surveillance of ESR, CRP, and imaging.

Discussion

Our understanding of vasculitis continues to evolve as demonstrated in the International Chapel Hill Consensus Conference (CHCC2012) revised nomenclature for vasculitides. This revision defines PAN as: “Necrotizing arteritis of medium or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules, and not associated with antineutrophil cytoplasmic antibodies (ANCA).”¹ Our case is consistent with a diagnosis of PAN, which remains a clinical diagnosis requiring exclusion of other similar disease processes. Other factors consistent with this diagnosis in our case include the vessel involved, typical histopathology finding of microaneurysm, and inflammatory cells involved in the absence of ANCA.

PAN causes transmural necrotizing inflammation which can weaken the wall resulting in visceral, renal, and peripheral aneurysms and rupture.²⁻⁶ Gastrointestinal involvement is common and can result in aneurysm, infarction, and bowel necrosis.⁷

The etiology of PAN is not well understood but believed to be immunopathogenetic and heterogeneous. It may be a consequence of hepatitis B virus infection with some evidence of hepatitis B antigen triggering immune complexes, which activate the complement cascade, leading to attraction and activation of polymorphonuclear neutrophils. There is no solid data on why such complexes deposit in the arterial wall. However, one theory would be the cross-reaction between the arterial wall antigens and the antigen that initiated the disease process.

Other possible etiologies for PAN have been reported including HIV, cytomegalovirus, parvovirus B 19, human T-lymphotropic virus type I, and hepatitis C virus.⁸ The end result is usually inflammation, edema, transmural necrosis, stenosis, aneurysmal formation, or subsequent healing.⁹

The diagnosis of PAN can be challenging due to nonspecific symptoms which may include malaise, weight loss, fever, and other constitutional symptoms. Patients may also present with painful peripheral neuropathy, musculoskeletal pain, vascular nephropathy, and cutaneous involvement (especially livedo reticularis). Presentation can be mild or fulminant.¹⁰ The vascular evaluation is based on presenting symptoms and degree of suspicion.

Laboratory testing may reveal nonspecific elevation of inflammatory markers such as ESR, CRP, α 2-globulin, elevated leukocytes, and eosinophils along with chronic disease anemia. In addition, the presence of hepatitis B surface antigenemia may be found. ANCA have rarely been found positive in PAN.⁸ Angiography has long been considered the gold standard for PAN diagnosis. Typical findings include multiple saccular aneurysms, particularly within the liver and kidneys as well as stenosis of the visceral arteries. Aneurysms as small as 3 mm in diameter can be identified by CT scan, an effective noninvasive way to follow-up patients with PAN.¹¹

Steroids have been used to treat PAN since 1951 and have increased survival rates for patients with PAN.¹² Some reports documented regression of associated aneurysms after combined medical treatment with steroids and immunosuppressant drugs.¹³ Better outcomes were shown in one randomized trial with the addition of azathioprine or cyclophosphamide versus steroids alone in those patients showing lack of clinical improvement in the presence of continuously elevated inflammatory markers.

First-line corticosteroid treatment was able to achieve and maintain remission in only about half of patients. Additional immunosuppressive therapy was effective for treating corticosteroid-resistant disease or major relapses and has achieved 5-year survival rates as high as 80%.^{14,15}

PDAA account for only 2% of all splanchnic artery aneurysms.¹⁶ PDAA are either true or pseudoaneurysms. True PDAA are most often caused by atherosclerosis. However, their association with celiac artery compression or occlusion has been well described.^{5,17-25} Pseudoaneurysms are more

common than true aneurysms and are caused by pancreatitis, infection, and less commonly trauma.²⁶⁻²⁹ Visceral aneurysms associated with polyarteritis are rare and can be lethal. Involvement of the pancreaticoduodenal arteries is extremely rare and requires a high index of suspicion in patients presenting in extremis secondary to a ruptured visceral artery aneurysm. Visceral artery aneurysm involvement with PAN tends to involve multiple vessels and segments of vessels, and most commonly affects the renal arteries.⁹ Hepatic artery involvement was found to be more common than the involvement of PDA based on a review of 18 cases of hepatic artery aneurysm.⁶ To our knowledge, there was only one other documented case report of a massive retroperitoneal hemorrhage involving a PDAA in a patient with PAN, which was successfully embolized.⁶

Management of visceral aneurysms includes medical management of the underlying etiology, surgical management including exclusion, excision, or revascularization. Percutaneous approaches such as coil embolization or N-butyl-2-cyanoacrylate (glue) can be used in stable patients and good renal function. Initial surgical management of PAN associated visceral aneurysm rupture was first described in 1968.

There was no evidence of PDAA on the pre-endograft AAA CT scan. Such fast growing aneurysms raise the suspicion for an inflammatory, infectious, or connective tissue disorder. However, there was no history, symptomatology, laboratory, or imaging findings to suggest infection or connective tissue disorder.

Our case represents a unique arterial complication of PAN, which to our knowledge, is the second case report of PDAA caused by PAN. He was managed successfully with steroids and surgical repair. Endovascular approach was not feasible due to the unstable condition of the patient. Furthermore, cyclophosphamide was not considered due to the lack of systemic symptoms. We suggest considering PAN among the differential diagnoses list when managing patients with PDAA.

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