Arterial Aneurysms in Wegener's Granulomatosis: Case Report and Literature Review

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Objective: Arterial aneurysms are characteristic of medium-size vessel vasculitis but are a very unusual feature of Wegener's granulomatosis (WG). We describe a typical WG case, complicated by arterial aneurysms and review previously reported cases.

Methods: Medline database search of cases published between January 1978 and July 2006, in English, reporting arterial aneurysms complicating WG.

Results: Five years after diagnosis, a 29-year-old man with typical WG developed macro- and microaneurysms located on branches of the hepatic and renal arteries during a disease relapse. The main symptoms were abdominal pain, vomiting, and altered general status. He was successfully treated by coil embolization in combination with prednisone, intravenous mycophenolate mofetil, and high-dose immunoglobulins. Twelve additional cases of WG complicated by arterial aneurysms are reported in the English literature. This represents a life-threatening complication since rupture occurred in half of the patients.

Conclusions: Although small-vessel injury predominates in WG, inflammation of medium-size arteries may occur and lead to aneurysm formation. Abdominal angiography should be recommended when unexplained abdominal pain occurs during a WG flare.

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Wegener's granulomatosis (WG) is a small-vessel vasculitis involving primarily the respiratory tract and kidneys. Histological examination reveals necrotizing vasculitis affecting mostly small-size vessels (eg, capillaries, venules, and arterioles). It may also affect larger arteries, and thus, the vascular distribution overlaps with that of the medium- and large-size vessel vasculitides (1). Chronic inflammation can lead to arterial aneurysm formation, a characteristic of medium-size vessel vasculitis, but a very unusual feature of WG. This case report presents a patient with typical WG who developed several aneurysms in branches of the hepatic and renal arteries during a relapse.

METHODS

A literature survey from January 1978 to July 2006 was performed using a MEDLINE (PubMed), searching for cases with coexisting WG and aneurysm. Only cases published in English with sufficient clinical data for analysis were reviewed.

CASE REPORT

An Algerian man, born in 1973, was admitted to our department of internal medicine in January 2003, because of weight loss, vomiting, and epigastric pain. He had been diagnosed with WG in May 1997 on the basis of nasal ulcerations with epistaxis, nasal perforation and saddle-nose deformity, sinusitis, orchitis, polyarthralgias, myalgias, subcutaneous nodules, polyneuropathy, and acute renal insufficiency related to pauci-immune crescentic glomerulonephritis. Histological examination of skeletal muscle biopsy showed medium-size artery vasculitis. Cytoplasmic antineutrophilic cytoplasmic antibody

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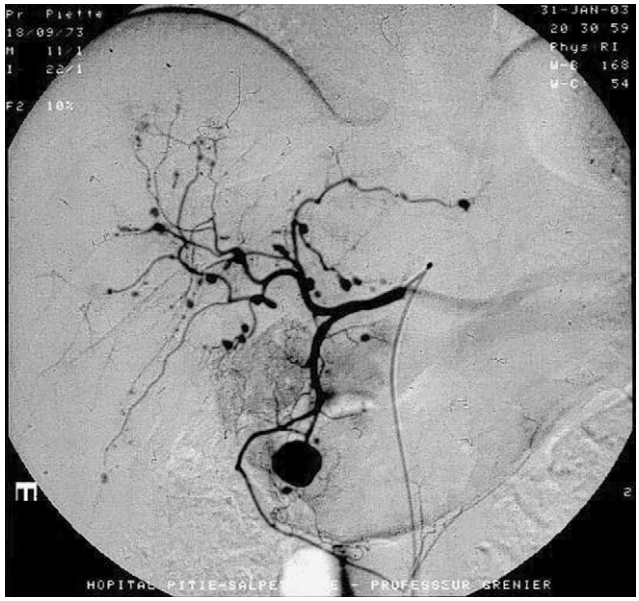

Figure 1 Common hepatic artery angiography showed a 2-cm-diameter aneurysm located on the superior pancreatic-duodenal artery and multiple microaneurysms of the hepatic branches.

(ANCA) with antiproteinase-3 (anti-PR-3) specificity was present. Serologic tests for hepatitis B, C, and human immunodeficiency virus (HIV) were negative. Remission was obtained with 1 mg/kg daily prednisone, monthly intravenous 1.2 g cyclophosphamide pulses, replaced in September 1999, after the ninth pulse, by azathioprine 150 mg daily. At that time, daily prednisone dose was 20 mg, and ANCA testing was negative.

In September 2002, while the patient had been receiving prednisone 10 mg/d, azathioprine 150 mg/d, and trimethoprim-sulfamethoxazole, he developed rhinitis, nodular lesions of the elbows and ear lobe, myalgias, hematuria, and proteinuria (1.2 g/24 hour). Histological examination of skin nodules showed perivascular giant-cell granuloma and necrotizing vasculitis involving small arteries. Anti-PR3 antibody titer was 35 IU/mL ($N < 10$). The patient received prednisone 35 mg daily (patient declined a 1 mg/kg/d dosage) and 3 methylprednisolone pulses, along with monthly 1.4 g intravenous cyclophosphamide pulses.

In January 2003, while on a prednisone dose of 27.5 mg daily, he complained of persistent epigastric pain, vomiting, a 15-kg weight loss during the previous 3 months, and rhinitis. His abdomen was moderately tender in the epigastric quadrant, without rigidity. Nodular skin lesions located on the elbows and ears persisted, and lower limb purpura and left big toe ischemia appeared. Laboratory tests showed the following: leukocyte count, 20,000/mm^3; hemoglobin, 9.9 g/dL; C-reactive protein, 157 mg/L; serum albumin, 24 g/L; normal serum lipase, microscopic hematuria, proteinuria 2.3 g/24 hour; serum creatinine, 127 μmol/L; and anti-PR3 ANCA antibodies, 15 IU/mL. Sinus computed tomography (CT) scan showed left maxillary sinusitis and chest CT scan was normal. Upper digestive fibroscopy revealed oesophagitis and abdominal echography showed a $2 \times 2 \times 3$ cm aneurysm near the pancreas head. Abdominal CT scan confirmed the presence of a 2-cm-diameter aneurysm located on the superior pancreatic-duodenal artery and multiple microaneurysms in the hepatic and renal arteries branches (Fig. 1), parietal thickening of the small bowel, and splenic infarction.

The patient was then treated by coil embolization of the largest aneurysm and methylprednisolone pulses (1g

Table 1 Large- and Medium-Size Vessels Aneurysms in Wegener's Granulomatosis: Literature Review

References	Gender Age (y)	Criteria for Diagnosis of WG	Duration Between Aneurysm and WG Diagnosis	Involved Artery
Sieber (3)	M/59	Clinical picture, NGV, cANCA +	9 months before	Aorta
Blockmans (4)	M/42	Clinical picture, NGV, PR3-cANCA + PICG	Concomitant	Aorta
Carels (5)	M/63	Clinical picture, NGV, MPO-ANCA +	Concomitant	Aorta
Shirit (6)	M/58	Clinical picture, pulmonary capillaritis, MPO-ANCA +	Concomitant	Subclavian
Senf (7)	M/35	Clinical picture, NGV, PR3-cANCA +	1 month after	Hepatic, renal, splanchnic
Backer (8)	M/24	Clinical picture, NGV	Concomitant	Renal
Moutsopoulos (9)	M/30	Clinical picture, NGV	4 months after	Renal
	F/53	Clinical picture	1 month after	Renal
Famularo (10)	M/67	Clinical picture, NGV, PR3-cANCA + PICG	1 month after	Pancreatic-duodenal
Aoki (2)	M/56	Clinical picture, NGV, PR3-cANCA +	Concomitant	Left gastric
Den Bakker (11)	M/55	Clinical picture, NGV, PR3-cANCA + PICG	15 days after	Hepatic
Takei (12)	M/34	Clinical picture, PR3-cANCA + PICG	1 year before	Intracranial
Present study	M/29	Clinical picture, NGV, PR3-cANCA + PICG	5 years after	Pancreatic-duodenal, hepatic, renal

M, male; F, female; WG, Wegener's granulomatosis; NGV, biopsy proven necrotizing granulomatous vasculitis; PICG, biopsy-proven pauci-immune crescentic glomerulonephritis; PR3-cANCA, cytoplasmic antineutrophil cytoplasmic antibody with anti-proteinase 3; MPO-ANCA, myeloperoxidase-ANCA; CY, cyclophosphamide; NS, not stated, IVIg, intravenous immunoglobulin; MMF, mycophenolate mofetil.