BRIEF COMMUNICATIONS

OCCULT ACTIVE GIANT CELL AORTITIS NECESSITATING SURGICAL REPAIR

Leslie Dubin Kerr, MD,^a Ya Ju Chang, MD,^a Harry Spiera, MD,^a and John T. Fallon, MD, PhD,^b *New York, NY*

Although giant cell arteritis (GCA) affecting the aorta has been well described, its incidence as a complication of temporal arteritis has been considered infrequent.¹ Aortic dissection and aneurysm rupture were noted as rare, catastrophic sequelae occurring in patients with a previously established diagnosis of temporal arteritis² who had symptoms referable to the cranial arteries or upper extremity claudication.

In contrast, we describe 19 patients who presented, over a 4-year period, with thoracic aneurysms, abdominal aneurysms, or both, caused by active giant cell aortitis documented histopathologically. Only one patient had a history of temporal arteritis before aortic repair. The others lacked vascular symptoms.

Our experience suggests that active aortitis caused by occult GCA occurs more commonly than has been previously appreciated and may represent an important clinical variant of this disease. Efforts to improve recognition of giant cell aortitis to define optimal treatment strategies are warranted.

Patients and methods. A total of 1069 cases of aortic aneurysm repair or replacement performed between 1994 and 1998 were reviewed, in which 19 cases of active giant cell aortitis confirmed histopathologically were identified. All tissues reviewed were obtained at the time of surgery and were routinely processed for hematoxylin and eosin staining, as well as Gram staining, acid-fast staining, and fungal staining. The pathologic diagnosis of GCA was based on the finding of focal areas of medial necrosis surrounded by a granulomatous-like inflammatory reaction. Occasional multinucleated giant cells were present at the periphery of the necrotic areas. The necrotic areas were devoid of cells and contained com-

pacted elastic fibers. The intima and adventitia were not usually involved (Fig 1).

Results

Patient characteristics (Table I). Of the 19 patients, 12 were women, and 6 were men. The mean age at the time of surgical repair was 73.2 years (range, 41-85 years).

Clinical symptoms and clinical course. The most common clinical symptoms were dyspnea on exertion (11/19) and angina (6/19).

Symptoms referable to the cranial arteries, such as scalp tenderness, jaw claudication, extremity claudication, vision loss, or temporal artery tenderness were not mentioned. The mean duration of onset of symptoms was approximately 2.2 years.

The most common physical finding consisted of a new murmur (14/19), with aortic insufficiency confirmed in 9 of 19 patients by echocardiography, cardiac catheterization, or both. Hypertension was present in 6 of 19 patients during the perioperative period. Sedimentation rates were not obtained in any patient. All aneurysms were classified preoperatively as atherosclerotic.

At the time of surgery, 17 of 19 had thoracic aortic aneurysms involving the ascending portion of the aorta. Two of these patients had acute dissection. Six of 19 had aneurysms of both the thoracic and abdominal aortas, and 2 of 19 had aneurysms of the abdominal aorta alone. Of the 9 aortic valves replaced, none had valvulitis. All patients had resection of the aneurysm and graft placement. No mention was made of any unusual gross operative findings indicative of giant cell aortitis at the time of surgery. There was no postoperative mortality, and all patients were discharged home without the use of steroids or immunosuppressive agents, because the importance of the histopathology was not appreciated and rheumatologic consultations were not sought.

Follow-up and outcome. The duration of follow-up varied from 1 to 4 years. During this period, there were two sudden deaths. The exact cause of death, however, could not be ascertained in either case. Nine patients are alive; of these, two are symptomatic.

Discussion. These 19 cases of tissue-documented active giant cell aortitis necessitating surgical intervention define a subgroup of patients with GCA whose active aortitis affecting the thoracic aorta is clinically occult, who lack symptoms referable to the cranial arteries, who have no history of temporal arteritis, and whose outcomes may be catastrophic.

Our experience is similar to that of previous authors. Brack and colleagues³ described two clinical patterns of

From the Department of Medicine, Division of Rheumatology,^a and Department of Pathology and Cardiovascular Institute,^b Mount Sinai School of Medicine, New York, NY.

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Address for reprints: Leslie D. Kerr, MD, Box 1244, Division of Rheumatology, Mount Sinai School of Medicine, 1 Gustave L. Levy Place, New York, NY 10029-6574 (E-mail: daniel.calto@mssm.edu).

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Table I. Clinical characteristics of 19 patients with giant cell aortitis

Patient No	Age (y)	Sex	Cardiovascular symptoms	Blood pressure (mm Hg)	New murmur
1	41	F	Angina	106/60	+
2	74	M	CHF, angina	NR	—
3	83	F	CHF, angina, palpitations	150/90	+
4	66	F	None	100/78	+
5	65	F	CHF, angina, palpitations	160/68	+
6	76	M	CHF	130/58	+
7	77	F	CHF	NR	+
8	79	F	CHF, angina	162/100	+
9	80	M	CHF	116/60	+
10	80	F	None	140/70	+
11	81	F	CHF	130/80	+
12	78	M	None	146/70	+
13	85	M	CHF	150/70	+
14	75	F	CHF, angina	140/50	—
15	72	F	None	121/74	—
16	74	M	None	140/90	—
17	63	F	Palpitations	150/80	—
18	68	M	None	102/52	+
19	74	F	CHF	142/98	+

descend, Descending aorta; *F*, fusiform; *NR*, Not recorded; *CHF*, congestive heart failure; *ascend*, ascending aorta; *S*, saccular.

Fig 1. Tissue obtained from patient 1, demonstrating giant cell aortitis. **A**, Active aortitis. *Arrow* identifies a giant cell. (Hematoxylin and eosin stain; original magnification 200 $\times$.) **B**, Giant cell aortitis with a large area of necrosis. (Hematoxylin and eosin stain; original magnification 100 $\times$.)

GCA consisting of cranial arteritis and large-vessel GCA. Their subgroup of patients with large-vessel disease had upper-extremity claudication or cardiovascular symptoms only. Evans and colleagues¹ described a markedly increased incidence of thoracic aortic aneurysms in their cohort of patients with biopsy-proven GCA compared with an age-matched population. Liu and colleagues,⁴ in a review of 24 patients with thoracic aortic dissection caused by giant cell aortitis documented histopathologically, found that 11 of 24 had no prior symptoms of temporal arteritis and received no diagnosis until their catastrophic illness. Lie⁵ described 72 patients with biopsy- or autopsy-documented active giant cell aortitis or extracranial GCA. Twenty-five percent of these patients had no symptoms referable to the cranial arteries. At surgery or autopsy, active giant cell aortitis affecting the aortic arch and

ascending aorta predominated in 39%, which is similar to what we found in our patients.

Although incomplete, our follow-up data are suggestive of a poor outcome subsequent to successful surgical repair. Because GCA is a systemic disease, it is unlikely that aortic repair, although life-saving in the short term, is curative. Unfortunately, despite histopathologic documentation of active aortitis rather than atherosclerosis alone, none of these patients received either steroids or immunosuppressive therapy because the importance of the histopathology documenting active vasculitis was not appreciated, and rheumatologic input was not obtained.

Further study is required to define the optimal immunosuppressive regimen and is dependent on improved recognition of aortitis by the surgeons involved in these patients' care.

<i>Hemoglobin level (mg/dL)</i>	<i>Location of aneurysms and type</i>	<i>Aortic insufficiency/ aortic valve replacement</i>	<i>Acute dissection</i>	<i>Outcome (1-4 y postoperative)</i>
12.3	Descend (F)	—	—	NR
11.9	Arch, ascend, descend (F)	—	+	Alive
13.0	Arch, ascend (F)	—	—	NR
14.0	Ascend (F)	+	—	Alive
12.3	Ascend (F)	+	—	Alive
13.9	Ascend (F)	+	—	Alive
11.7	Ascend (F)	+	—	NR
9.9	Arch, ascend, descend (S)	+	—	NR
10.1	Ascend (F)	—	—	Dead
NR	Ascend (F)	+	—	NR
14.1	Arch, ascend, descend (F)	—	—	Alive
13.8	Ascend (F)	+	+	NR
14.7	Arch, ascend (F)	+	—	Alive
13.4	Ascend (F)	—	—	Alive
10.5	Arch, ascend, descend (F)	—	—	Alive
14.3	Arch, ascend, descend (F)	—	—	NR
13.0	Descend (NR)	—	—	Alive
14.3	Arch, ascend, descend (F)	+	—	Dead
13.0	Arch, ascend (F)	—	—	Alive

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REFERENCES

1. Evans JM, O'Fallon WM, Hunder GG. Increased incidence of aortic aneurysm and dissection in giant cell (temporal) arteritis. *Ann Intern Med* 1995;122:502-7.
2. Evans JM, Bowles CA, Bjornsson J, Mullany CJ, Hunder GG. Thoracic aortic aneurysm and rupture in giant cell arteritis. *Arthritis Rheum* 1994;37:1539-47.
3. Brack A, Martinez-Taboada V, Stanson A, Goronzy JJ, Weyland CM. Disease pattern in cranial and large vessel giant cell arteritis. *Arthritis Rheum* 1999;42:311-7.
4. Liu G, Shupak R, Chiu BKV. Aortic dissection in giant cell arteritis. *Semin Arthritis Rheum* 1995;25:160-71.
5. Lie JT. Aortic and extracranial large vessel giant cell arteritis: a review of 72 cases with histopathologic documentation. *Semin Arthritis Rheum* 1995;24:422-31.