Aortic Aneurysms in Patients with Takayasu’s Arteritis: CT Evaluation

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OBJECTIVE. The objective of this study was to evaluate the incidence, development, and outcome of aortic aneurysm on CT in a group of patients with Takayasu’s arteritis.

MATERIALS AND METHODS. We reviewed the files of 31 patients with Takayasu’s arteritis between January 1990 and March 1999. All patients were followed up for more than 6 months with CT, and the mean follow-up period was 52.9 months (median, 36.0 months). In all patients, initial CT was performed within 6 months of diagnosis of Takayasu’s arteritis. The study group comprised 24 female patients and seven male patients; their ages at the first CT examination ranged from 8 to 72 years (mean, 42.6 ± 16.5 years).

RESULTS. Seventeen aortic aneurysms were found in 14 (45.2%) of the 31 patients. Patients with severe calcification of the aorta showed significantly lower incidence of aneurysm formation than those without severe calcification of the aorta (p < 0.05). Of the 17 aneurysms, three were not present at the time of initial CT and appeared during the follow-up period. Nine of 17 aneurysms increased in size during the follow-up period. Three of the nine aneurysms rapidly increased in size and ruptured during the follow-up period. In all three of these aneurysms, aortic wall thickening was identified on CT. The remaining six aneurysms slowly increased in size but did not rupture.

CONCLUSION. Aortic aneurysm associated with Takayasu’s arteritis is not rare. The aorta with little calcification has a greater possibility of aneurysm formation in patients with Takayasu’s arteritis. Aortic aneurysms with wall thickening can have fatal consequences.

Takayasu’s arteritis is a primary arteritis of unknown etiology that commonly affects the aorta and its major branches as well as the pulmonary artery [1–4]. Although more common in Asia, the disease has been found worldwide, usually affecting young women [1–5]. The disease involves the vessel walls and results in luminal abnormalities including stenosis, occlusion, and aneurysm formation [1–5].

The long-term prognosis for patients with Takayasu’s arteritis is relatively good [6, 7]. However, an aneurysm that forms can progress to a fatal outcome such as heart failure due to aortic valve regurgitation and aneurysm rupture, which is the most frequent fatal course in Takayasu’s arteritis [6, 7].

We reviewed cases of aortic aneurysm due to Takayasu’s arteritis and evaluated the CT appearances of the aorta using follow-up results. The purpose of this study was to evaluate aortic aneurysm formation and the fate of aortic aneurysms in patients with Takayasu’s arteritis.

Materials and Methods
Patients
The ethics committee at Omura Municipal Hospital approved this study. We reviewed the files of 31 patients with Takayasu’s arteritis between January 1990 and March 1999. All 31 patients were followed up for more than 6 months with CT. The mean follow-up period was 52.9 months (median, 36.0 months). The diagnosis of Takayasu’s arteritis was based on guidelines established by the Aortitis Syndrome Research Committee of Japan and included clinical signs and symptoms, laboratory findings, and angiographic features [1, 8, 9]. In all patients, initial CT was performed within 6 months from diagnosis of Takayasu’s arteritis. The study group comprised 24 female patients and seven male patients, with their ages at the first CT examination ranging from 8 to 72 years (mean, 42.6 ± 16.5 years). None of the patients had Marfan’s syndrome. Thirteen patients had hypertension; in two of them, the hypertension was renovascular. In two patients with aortic valve regurgitation, surgery was performed during the follow-up period. Three patients had undergone steroid therapy for more than 1 year. One had undergone surgical...
revascularization (abdominal aorta to left common carotid artery bypass, abdominal aorta to left vertebral artery bypass, and left vertebral artery to left axillary artery bypass) for the occlusion of both the subclavian artery and left common carotid artery. Hypertension was defined as 140 mm Hg or greater brachial systolic, 90 mm Hg or greater brachial diastolic, or both; or 160 mm Hg or greater popliteal systolic, 90 mm Hg or greater popliteal diastolic, or both at the time of initial CT [6]. In all nine patients with aortic wall thickening, steroid therapy was initiated at diagnosis of Takayasu's arteritis.

**Imaging Methods**

The initial and follow-up imaging studies were performed with unenhanced and enhanced CT in all patients. For enhanced CT, a bolus of 100 mL of non-ionic contrast material was injected. CT was performed on either a 9800 scanner (General Electric Medical Systems, Milwaukee, WI) or a Somatom Plus 32S (Siemens, Erlangen, Germany). Axial images were generated with 5-mm-thick sections and 5-mm-thick intervals from the top of the aortic arch to the abdominal aorta. CT follow-up was planned at 4-month intervals (20 patients) or 6-month intervals (11 patients); CT scans were obtained sooner if warranted by clinical symptoms. A total of 424 CT studies were performed (mean, 31.3 studies per patient).

**Analysis of Images**

CT images were evaluated by two experienced cardiovascular radiologists according to the following features. The final decisions about the findings were reached by consensus.

* Aortic wall thickening.—Aortic wall thickening was defined as the wall being more than 1 mm thick with enhancement on a contrast-enhanced CT image when the previous study had revealed that the aortic wall was either less than 1 mm thick or imperceptible in healthy adults [3,10].

* Calcification of the aortic wall.—Calcification of the aortic wall was classified as none, moderate, and severe. Severe calcification referred to calcification that was completely circumferential and involved thickness of the aortic wall [10].

* Diameter of the aorta.—CT measurements were obtained by measuring several diameters with a direct-reading caliper from hard copy images and correcting for appropriate scale [11]. The largest short-axis diameter of the outer contour of the affected aorta was measured. In saccular aneurysm, the diameter from the outer contour of the affected aorta to the outer contour of the aneurysm was measured. In the aortic arch, the diameter perpendicular to the curvature was measured.

An aneurysm was defined as a local increase in the diameter of the aorta (increase in diameter of >50% compared with the normal adjacent segment) and was categorized as either fusiform or saccular on the final CT examination [12]. A fusiform aneurysm was defined as circumferential enlargement of the aorta, and a saccular aneurysm was defined as enlargement predominantly involving a portion of the aorta [12].

**Growth rate of the aneurysm.**—The mean difference in the aortic size between the initial and final examinations was measured at approximately the same aortic portion where the aneurysm had increased in size. The growth rate of the aneurysm was calculated in patients who were followed up for more than 6 months from the initial examination. The growth rate of the aneurysm was obtained by dividing the difference in the diameter between the initial (D1) and the final (D2) measurements by the time interval (T) between the two measurements [11]: growth rate = (D2 – D1)/T.

**Statistical Analysis**

The data are expressed as means plus or minus the standard deviation. Comparison of the means between the groups was performed with the Mann-Whitney U test. Statistical comparisons of characteristics were made with Fisher’s exact test because the expected number of cells was less than five. Analysis was performed with STAT-VIEW software for Macintosh (Abacus Concepts, Berkeley, CA). A p value of less than 0.05 was considered significant.

**Results**

**Comparison of Patients With and Without Aortic Aneurysms**

The CT findings and clinical outcomes of 31 patients with Takayasu's arteritis are summarized in Table 1 and Figure 1.

<table>
<thead>
<tr>
<th>Location of Aortic Aneurysm</th>
<th>CT Findings</th>
<th>No. of Patients with Hypertension</th>
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<tbody>
<tr>
<td></td>
<td>Wall Thickening</td>
<td>Severe Calcification</td>
</tr>
<tr>
<td>Ascending aorta (n = 2)</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Aortic arch (n = 3)</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Descending aorta (n = 1)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ascending aorta to descending aorta (n = 1)</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Aortic arch to descending aorta (n = 2)</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Abdominal aorta (n = 8)</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Total (n = 17)</td>
<td>7</td>
<td>3</td>
</tr>
</tbody>
</table>

| Outcome of Aneurysms in Patients with Takayasu’s Arteritis |

Seventeen aortic aneurysms in 14 patients included one saccular aneurysm and 16 fusiform aneurysms. Of the 17 aneurysms, three were not present at the time of the initial CT study and appeared during the follow-up period. During the follow-up period nine of the 17 aneurysms increased in size and the remaining eight aneurysms did not increase in size. Of the nine aneurysms that increased in size during the follow-up period, five were located in the thoracic aorta and four in the abdominal aorta (mean increase in aortic diameter, 2.33 ± 1.94 cm). The mean growth rate of aneurysms was 0.04 cm/year.

Three aneurysms rapidly increased in size and ruptured (mean increase in aortic diameter, 4.67 ± 1.53 cm). Of these three aneurysms, two were located in the thoracic aorta and one in the abdominal aorta. The mean growth rate of these three aneurysms was 1.16 cm/year. In all three aneurysms, aortic
CT of Aortic Aneurysms in Takayasu's Arteritis

Fig. 1.—Schematic diagram shows results of 31 patients with Takayasu’s arteritis.

Fig. 2.—8-year-old girl with Takayasu’s arteritis. A, Initial contrast-enhanced CT image shows fusiform aneurysm and wall thickening of descending thoracic aorta (arrows). This finding suggests active inflammatory process. B and C, Contrast-enhanced CT images obtained 2 months (B) and 4 months (C) after A show that fusiform aneurysm has rapidly increased in size and wall thickness, in spite of early steroid treatment.
wall thickening was identified on CT (Figs. 2 and 3), and neither severe calcification nor hypertension was detected. Steroid therapy was not effective in these patients.

Six of nine aneurysms slowly increased in size but did not rupture (mean increase in aortic diameter, 1.67 ± 0.41 cm) (Fig. 4). Three were located in the thoracic aorta and three, in the abdominal aorta. The mean growth rate was 0.03 cm/year. No severe calcification of the aortic wall was identified in any of the six aneurysms. In four of the six aneurysms, aortic wall thickening was identified during the follow-up period. Of the six patients (mean age, 47.5 ± 18.1 years) with these six aortic aneurysms, one was male and five were female; all had hypertension.

Eight of the 17 aneurysms did not increase in size (Fig. 5). Four were located in the thoracic aorta and four, in the abdominal aorta. During the follow-up period severe calcification of the aortic wall was identified in two of these eight aneurysms, and aortic wall thickening was identified in three. In the eight patients with these eight aortic aneurysms (mean age, 41.9 ± 11.8 years), one was male, seven were female, and two had hypertension. Comparing the six aneurysms that increased slowly in size with the eight aneurysms that did not increase in size, there was a significant difference in the presence of hypertension (p < 0.01). However, no significant difference was detected between the two groups in terms of severe calcification of the aortic wall, location (thoracic aortic aneurysm versus abdominal aortic aneurysm), aortic wall thickening, age, and initial diameter of the aorta (p > 0.05).

Fig. 3.—45-year-old woman with Takayasu's arteritis.
A, Initial contrast-enhanced CT image shows wall thickening of aortic arch wall (arrows). This finding suggests active inflammatory process. B-D, Contrast-enhanced CT images obtained 12 months (B and C) and 15 months (D) after A show that aortic wall thickening (arrows, B and C) and saccular aneurysm have rapidly increased.
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Fig. 4.—56-year-old woman with Takayasu's arteritis and hypertension.
A. Initial contrast-enhanced CT image shows fusiform aneurysm in abdominal aorta.
B. Contrast-enhanced CT image obtained 3 years after A shows that fusiform aneurysm has increased in size.
C. Unenhanced CT image obtained on same day as B shows little calcification is present in aortic wall.

Fig. 5.—56-year-old woman with Takayasu's arteritis.
A. Unenhanced CT image shows fusiform aneurysm with severe calcification in abdominal aorta.
B. Unenhanced CT image obtained 5 years after A shows size of aorta has not changed. Calcification of aortic wall has become much heavier. Calcification indicates chronic arteritis.
Discussion

Takayasu’s arteritis is a primary arteritis of unknown etiology involving the vessel walls that results in luminal abnormalities including stenosis, occlusion, and aneurysm formation [1–5]. Although stenotic lesions are identified most frequently, aneurysm formation is the most frequent fatal complication in Takayasu’s arteritis [6, 7]. To our knowledge, only a few reports have focused on aortic aneurysms in patients with Takayasu’s arteritis [13–15]. The purpose of this study was to evaluate aortic aneurysms on CT in patients with Takayasu’s arteritis.

For the diagnosis of Takayasu’s arteritis, conventional angiography has been regarded as necessary. However, a recent report has shown that CT can clearly delineate aortic mural changes that are difficult to detect on conventional angiography including wall thickening, calcification, and mural thrombus [3].

Several reports have described aneurysm formation as being rarely associated with Takayasu’s arteritis. The incidence of aneurysm formation in previous reports about patients with Takayasu’s arteritis varies from 0% to 87.5% [13, 14]. In the present study, aortic aneurysms were identified in 45.2% of the patients. This result suggests that aneurysm formation is not rare in patients with this disease. However, this finding may have some bias because patients were not selected randomly and because the definition of aneurysm varies. More studies are needed to clarify this issue.

Pathologic studies of Takayasu’s arteritis have revealed marked thickening of the media, marked disruption of the elastic fibers, pronounced fibrotic thickening of the adventitia, and intimal thickening, all of which are causes of stenotic lesions [1, 16]. Previous studies described aneurysm formation as possibly caused by the degeneration and weakening of the media [1, 15]. In the present study, there was a significant difference between patients with and those without aortic aneurysms in the presence of severe calcification of the aortic wall. This finding suggests that aneurysm formation can occur in patients without severely scarred changes in the aortic wall because dystrophic calcification represents deposition of calcium in the scarred media and intima [1, 17]. Therefore, the aorta with no or little calcification has a greater possibility of aneurysm formation in patients with Takayasu’s arteritis.

Kumar et al. [14] reported that the aneurysmal form of Takayasu’s arteritis was associated with a higher incidence of aortic valve regurgitation, which may result from enlargement of the ascending aorta and an elevated erythrocyte sedimentation rate. In the acute (early) phase of Takayasu’s arteritis, abnormal laboratory findings are often identified, such as persistent elevation of erythrocyte sedimentation rate and a positive C-reactive protein test [1, 18]. Although laboratory findings were not evaluated in detail because our study was based on CT findings, aortic wall thickening is often characteristic on CT during the acute phase of Takayasu’s arteritis [1–3]. In the present study, there was no significant difference between patients with and those without aortic aneurysms in the presence of aortic wall thickening. One possible reason for this finding could be that steroid therapy was promptly initiated during the acute phase.

In the present study, three of 17 aneurysms rapidly increased in size and ruptured. In all these aneurysms, aortic wall thickening was identified. By showing arterial wall changes, cross-sectional imaging techniques such as CT play an important role in the early diagnosis of Takayasu’s arteritis [1–3]. Although the clinical and radiologic features of the acute (early) phase of Takayasu’s arteritis differ from those of the late (occlusive or pulseless) phase, the two phases of the disease are not clearly different because of the insidious onset of the disease and the relapse of active arteritis in some patients. In some patients with acute phase Takayasu’s arteritis, the significant feature is aortic wall thickening. Histologic examination of aortic wall thickening yields findings of florid inflammation in the media and adventitia [1, 10, 19]. According to our results, aortic wall thickening is not necessarily a trigger of aneurysm formation if proper steroid therapy is administered. However, close follow-up examination is needed in aneurysms with aortic wall thickening because this type of aneurysm can be fatal.

In the present study, six of 17 aneurysms slowly increased in size but did not rupture. All six aneurysms were fusiform and similar in appearance to atherosclerotic aneurysms. Comparing the six aneurysms that slowly increased in size with the eight aneurysms that did not increase, we did not find a significant difference in the presence of hypertension (p < 0.01). A previous report described the growth of atherosclerotic aortic aneurysms as being related to the location in the aorta, the initial diameter of the aneurysm, and the patient’s age and blood pressure [11]. The growth of atherosclerotic aneurysms is more closely related to the location, and, in addition, thoracic aortic aneurysms are more easily dilated than abdominal aortic aneurysms [11]. In the present study, no significant difference in terms of location was detected. Our results suggest the growth of aortic aneurysms in Takayasu’s arteritis is more closely related to blood pressure than to location. Histologic differences between the thoracic and abdominal aortas have been reported [10, 20]. The thoracic aortic media contains vasa vasorum thinner than that in the abdominal portion and contains more lamellar units. This finding suggests that aortic wall scars are more severe in the thoracic aorta than in the abdominal aorta. Scars can limit further enlargement of thoracic aortic aneurysms in Takayasu’s arteritis. Hirose et al. [11] reported that the growth rate of atherosclerotic aortic aneurysms was 0.34 cm/year, whereas the present study showed the growth rate of six aneurysms in patients with Takayasu’s arteritis was 0.03 cm/year, which is slower than that of the previous study. This result suggests that aortic wall scars are more severe in patients with Takayasu’s arteritis than in those with atherosclerosis. Therefore, aneurysms associated with Takayasu’s arteritis increase in size more slowly than atherosclerotic aortic aneurysms.

In conclusion, aortic aneurysms associated with Takayasu’s arteritis are not rare. The aorta with little calcification has greater possibility of aneurysm formation in Takayasu’s arteritis.

Some aortic aneurysms can rapidly increase in size and rupture. In particular, aortic aneurysms with aortic wall thickening should be closely followed up. In patients with aortic aneurysm without aortic wall thickening, long-term follow-up examination is also needed because the aneurysms may increase in size as a late complication.

References

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