Extracranial carotid aneurysm in Takayasu’s arteritis

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Extracranial carotid aneurysm caused by Takayasu’s arteritis is extremely rare. We have experienced six cases of extracranial carotid aneurysm among 106 cases of Takayasu’s arteritis that were treated surgically in the past 50 years. We herein review these cases and discuss the surgical indications and postoperative course of this rare disease. We report original observations about extracranial carotid aneurysm in Takayasu’s arteritis. (J Vasc Surg 2001;34:739-42.)

Takayasu’s arteritis is an arteritis of unknown etiology, resulting in stenotic or dilative lesions of the aorta and its major branches. However, there has been no description of the details of extracranial carotid aneurysm in Takayasu’s arteritis in the English medical literature. We report a case of right common carotid aneurysm due to Takayasu’s arteritis in a young woman and review five cases we have experienced in our hospital from 1953 to 1999.

CASE REPORT
A previously healthy 22-year-old woman was admitted to another hospital with a 3-month history of a pulsating mass in her right anterior cervical region, together with low-grade fever and general fatigue (Table, case 6). She had no history of hypertension, trauma, surgery, or irradiation in her neck, or any infection except for genital herpes. Blood tests showed erythrocyte sedimentation rate (ESR) of 120 mm/h and C-reactive protein (CRP) of 13.0 μg/mL. Blood culture was negative for microorganisms. Administration of prednisolone was started at 60 mg/d under the diagnosis of common carotid aneurysm due to nonspecific inflammation. Five days later, she was transferred to our hospital for further treatment.

On admission, her height was 159 cm, her weight was 53 kg, and her general condition was good. A physical examination revealed no abnormal finding except for a pulsatile mass in the right anterior cervical region. Body temperature was 36.1°C, blood pressure was 95/50 mm Hg in both arms, and pulse rate was 66 beats per minutes. Neurologic and ophthalmologic examination showed no abnormality.

Laboratory data showed diminished inflammatory reaction due to steroid therapy: an ESR of 19 mm/h and a CRP of 0.4 μg/mL. The results of routine hematologic, blood chemical, and coagulation tests were all normal except for an elevated white blood cell count of 14,200 per cubic millimeter. Test results for autoimmune antibodies and infectious diseases including syphilis, human immunodeficiency virus, and hepatitis B and C were all negative. A skin test was positive for tuberculosis; however, chest roentgenogram and sputum culture revealed no active infection. A reaction to needle puncture, which is often seen in patients with Bechet’s disease, was absent.

A duplex scan and a computed tomography (CT) scan of the neck showed a 24 × 40-mm fusiform aneurysm with a thick mural thrombus in the right common carotid artery. The arterial wall just proximal to the aneurysm was thickened. A CT scan of the abdomen showed thickening of the wall of the supraceliac aorta. Digital subtraction angiography showed a fusiform dilatation of the right common carotid artery and complete occlusion of the right external carotid artery with normal images of other parts of the artery including the left common carotid artery (Fig 1). The diagnosis of Takayasu’s arteritis was made on the basis of the presence of systemic inflammatory reaction and the anatomical locations of the affected arteries, the common carotid artery and supraceliac aorta, in a young woman.

Two weeks after admission, surgical treatment with the patient under steroid coverage was performed to prevent the risk of rupture. Adventitial fibrosis was observed just proximal and distal to the aneurysm. The aneurysm, together with the affected carotid artery with thickened wall, which was marked preoperatively on ultrasonography, was resected. Reconstruction was performed with an autologous saphenous vein graft. She was discharged without major complications.

On microscopic examination, there was a partial defect of the lamina elastica of the media. The intima showed hypertrophic change. The media had prominent fibrosis and disruption of the elastic lamina. The adventitia also had moderate fibrosis, and a few inflammatory infiltrates were seen extending from the adventitia to the media. No giant cells were seen (Fig 2). The findings in this case corresponded to the fibrotic type of Takayasu’s arteritis.1

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Competition of interest: nil.

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Published online Aug 13, 2001.


0741-5214/2001/$35.00 + 0 24/4/116808
From 1953 to 1999, we have experienced six cases of extracranial carotid aneurysm due to Takayasu’s arteritis including this case among 106 surgically treated cases of Takayasu’s arteritis (Table). The patients comprised three men and three women. The average age at diagnosis was 28 years, ranging from 21 to 40 years. The chief complaint was a neck mass in all six patients. Active systemic inflammation, which was diagnosed on the basis of a positive CRP or an elevated ESR of more than 20 mm/h before operation, was present in five of the six patients, and three of them underwent surgery while under corticosteroid coverage. Other arterial lesions were found in all six patients: occlusive lesions in four patients in the descending aorta, abdominal aorta, carotid, celiac, and radial artery, and aneurysmal lesions in five patients in the ascending aorta, descending aorta, brachiocephalic, and subclavian artery. In one of the five patients with aneurysmal lesions, a new aneurysm developed in the coronary artery 8 years later, which was successfully repaired. There were two cases of rupture. In one case of rupture (case 4), a neck mass with a diameter of 20 mm had been the only manifestation, without an elevation of ESR or CRP at the time of diagnosis. Eight months later, however, the aneurysm, the diameter of which had reached 30 mm at that time, suddenly started enlarging and ruptured 2 weeks later. In the other case of rupture (case 5), a sudden swelling of the right neck occurred a month after the development of slight fever and mild back pain. ESR and CRP were markedly elevated, and the diameter of the artery was 30 mm. During the long-term follow-up period, anastomotic aneurysms were found 6, 16, and 20 years later in case 2 and 3 years later in case 3. The anastomotic aneurysms in case 2 were all successfully treated with surgery. However, in case 3, we abandoned repair of the aneurysms because of technical surgical difficulty in 1962, and the patient died of ruptured descending aortic aneurysm.
Extracranial carotid aneurysm is a rare disease. The common causes are atherosclerosis, trauma, and previous surgery, followed by infection, previous neck irradiation, and fibromuscular dysplasia.3-6 There are also fewer frequently reported causes such as Marfan syndrome, Böchet’s disease, and Takayasu’s arteritis. Differential diagnosis is made according to each patient’s clinical history, symptoms, and distribution of the affected arteries. In this case, both the involvement of the supraceliac aorta and the elimination of other possible causes in a young woman led to the diagnosis of Takayasu’s arteritis. Nasu1 classified the histopathologic pictures of arterial lesions in Takayasu’s arteritis into three types: granulomatous inflammation, diffuse productive inflammation, and fibrosis. Although no giant cells were seen, the histologic findings of this case corresponded to fibrotic type and supported the clinical diagnosis.

The reported incidence of aneurysmal lesions in Takayasu’s arteritis varies from 4.9% to 31.9%.2,7-10 The most common site of aneurysm is the aorta, followed by the subclavian, brachiocephalic, and common carotid artery, and the incidence of extracranial carotid aneurysm varies from 1.8% to 3.9%.8-10 In our series, all of the aneurysms affected the right carotid artery and none the left side. In other reports, aneurysms of the branches of the aortic arch tended to occur in the right carotid or right subclavian arteries in Takayasu’s arteritis.9,10

**Clinical information of six cases of extracranial carotid aneurysm due to Takayasu’s arteritis**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Diameter (mm)</th>
<th>Site</th>
<th>ESR (mm/h)</th>
<th>CRP</th>
<th>Duration of symptoms before operation (mo)</th>
<th>Other vascular lesions</th>
<th>Steroid therapy before surgery</th>
<th>Surgical treatment for carotid aneurysm</th>
<th>Long-term postoperative course</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>26</td>
<td>F</td>
<td>20</td>
<td>R</td>
<td>16</td>
<td>Not known</td>
<td>10</td>
<td>Diffuse dilatation of ascending aorta, stenosis of left carotid artery, occlusion of left radial artery</td>
<td>None</td>
<td>Periadventitial sclerosis therapy with dicetylphosphate*</td>
<td>Died of heart failure at age 50.</td>
</tr>
<tr>
<td>2</td>
<td>40</td>
<td>F</td>
<td>30</td>
<td>R</td>
<td>40 (—)†</td>
<td>84</td>
<td>Diffuse dilatation of ascending aorta and left subclavian occlusion</td>
<td>None</td>
<td>Resection and reconstruction with Teflon graft</td>
<td>Successful reoperations for anastomotic aneurysms 6, 16, and 20 years later. Died of heart failure at age 65. Diagnosed of anastomotic aneurysm 3 y later that was not repaired. Died of ruptured aneurysm of descending aorta at age 27. Died during surgery.</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>21</td>
<td>F</td>
<td>15</td>
<td>R</td>
<td>120 6+</td>
<td>12</td>
<td>Aneurysms of descending aorta and brachiocephalic artery, occlusion of celiac artery and stenosis of abdominal aorta</td>
<td>Prednisolone 1 mg/kg per d for 2 wk</td>
<td>Resection and reconstruction with Dacron graft</td>
<td>Diagnosis and operation for coronary aneurysm 8 y after operation. Alive &amp; well for 16 years after initial operation. Alive &amp; well for 1 y after operation.</td>
<td></td>
</tr>
<tr>
<td>4†</td>
<td>33</td>
<td>M</td>
<td>20</td>
<td>R</td>
<td>23 (—)†</td>
<td>19</td>
<td>Left subclavian aneurysm and coarctation of aorta</td>
<td>None</td>
<td>Ligation of uncontrollable bleeding at emergency operation</td>
<td>Died of uncontrollable bleeding at emergency operation. Died of left heart failure at age 50.</td>
<td>Died of heart failure at age 50.</td>
</tr>
<tr>
<td>5‡</td>
<td>23</td>
<td>M</td>
<td>30</td>
<td>R</td>
<td>150 6+</td>
<td>1</td>
<td>Dilatation of descending aorta</td>
<td>Prednisolone 0.5 mg/kg per d for 10 d</td>
<td>Resection and reconstruction with autologous saphenous vein graft</td>
<td>Died of heart failure at age 50.</td>
<td>Died of left heart failure at age 50.</td>
</tr>
<tr>
<td>6§</td>
<td>22</td>
<td>F</td>
<td>25</td>
<td>R</td>
<td>120 13.0 µg/mL</td>
<td>3</td>
<td>Thickening of wall of supraceliac aorta</td>
<td>Prednisolone 1 mg/kg per d for 3 wk</td>
<td>Resection and reconstruction with autologous saphenous vein graft</td>
<td>Died of ruptured aneurysm of left subclavian artery</td>
<td>Died of left heart failure at age 50.</td>
</tr>
</tbody>
</table>

† Negative.
‡ Case of rupture.
§ Present case.
R, Right.

**DISCUSSION**

Extracranial carotid aneurysm is a rare disease. The common causes are atherosclerosis, trauma, and previous surgery, followed by infection, previous neck irradiation, and fibromuscular dysplasia.3-6 There are also fewer frequently reported causes such as Marfan syndrome, Böchet’s disease, and Takayasu’s arteritis. Differential diagnosis is made according to each patient’s clinical history, symptoms, and distribution of the affected arteries. In this case, both the involvement of the supraceliac aorta and the elimination of other possible causes in a young woman led to the diagnosis of Takayasu’s arteritis. Nasu1 classified the histopathologic pictures of arterial lesions in Takayasu’s arteritis into three types: granulomatous inflammation, diffuse productive inflammation, and fibrosis. Although no giant cells were seen, the histologic findings of this case corresponded to fibrotic type and supported the clinical diagnosis.

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that some hemodynamic mechanisms contribute to the formation of these aneurysms.

There is no established indication for surgery. Our experience indicates that all extracranial carotid aneurysms in Takayasu’s arteritis have a risk of rupture, even in the noninflammatory stage. Surgical treatment is recommended for almost all extracranial carotid aneurysms in Takayasu’s arteritis. No clear relationship could be observed between the activity of systemic inflammation or steroid administration and the incidence of early postoperative complications or anastomotic aneurysm formation in the late period, and an early operation with the patient under steroid coverage should not be avoided even in the acute inflammatory stage. Anastomotic aneurysms occurred at the anastomosis with a prosthetic graft. Although the precise mechanisms are not known, these experiences alert us to the importance of an all-autologous-vein policy for carotid reconstruction in Takayasu’s arteritis. Other aneurysms, including anastomotic aneurysms, can appear even after resection of the primary aneurysm, and periodic evaluation of grafts and other arteries is important in the postoperative follow-up.

REFERENCES


Submitted Jan 8, 2001; accepted Apr 4, 2001.