PSEUDOANEURYSM OF THE DISTAL AORTIC ARCH IN BEHÇET'S DISEASE

A Case Report

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Behçet's disease, manifested as a triform of relapsing iritis, stomatitis and genital ulceration, is now recognized as a systemic vasculitis that affects both veins and arteries. However, aneurysms of the aortic arch associated with Behçet's disease are extremely rare. We report on a successfully resected pseudoaneurysm of the distal aortic arch in a patient with Behçet's disease. The aneurysm was evaluated preoperatively by computed tomography (CT), magnetic resonance imaging (MRI) and intravenous digital subtraction angiography (DSA). We found that these non-invasive methods were quite useful for diagnosing the aneurysm in the aortic arch. Aneurysms enlarge rapidly when coupled with infection and are prone to rupture, thus requiring extensive surgical repair. However, since false aneurysm formations often occur at the site of anastomosis, postoperative follow-up should be monitored very closely.

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Behçet's disease, originally described in 1937, is characterized by uveitis and iridocyclitis, aphthous stomatitis and genital ulceration. The syndrome is most often observed among inhabitants of the Eastern Mediterranean region and Japan; usually in young adults 18—40 years of age and is twice as common among males. It is now clear that Behçet's disease is a systemic vasculitis affecting both arteries and veins, with lesions ranging from arterial occlusions and aneurysms to superficial thrombophlebitis and occlusion of the superior and inferior venae cavae. Most vascular deaths from this disease are related to a rupture of the aneurysms.

We report on a successfully resected pseudoaneurysm of the distal aortic arch in a patient with Behçet's disease. We also present a review of the literature on aneurysms occurring in cases of Behçet's disease.

CASE REPORT

A 25-year-old man had suffered subarachnoid hemorrhage, sterile meningitis and cerebellar hemorrhage 7 years prior to this report. Four-vessel cerebral angiography taken at that time showed no evidence of aneurysm or arterial-venous malformations. Aphthous stomatitis and genital ulceration had been noted 6 years previously and the diagnosis of Behçet's disease was made. He was treated with prednisone (20 mg/day) for a short period at the Department of...
Aneurysm of the Aortic Arch in Behçet's Disease

Fig. 1. (a) Chest roentgenogram (February 1, 1991) shows a slightly enlarged aortic arch. (b) Chest roentgenogram taken on admission (March 20, 1991) shows a very high-rising aortic arch (arrow).

Fig. 2. CT conducted after contrast medium infusion shows a mass (↑) adjacent to the aortic arch involving the left subclavian artery (↑↓). Lumen of the mass as well as the aortic lumen were filled with contrast medium.

Fig. 3. Lateral magnetic resonance scan of the chest shows a mass along with a thrombus (arrow) overlying the aortic arch.

Fig. 4. DSA examination of the aorta (left anterior oblique position at 30°) shows an aneurysm in the distal aortic arch.

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He experienced persistent fever and left back pain beginning in mid-January 1991. However, there was no history of any thoracic trauma. A chest roentgenogram taken on February 1 (Fig. 1-a) showed a slightly enlarged aortic arch. The fever and left shoulder pain continued despite oral administrations of antibiotics. Another chest roentgenogram taken on March 20 (Fig. 1-b) showed a very high-rising aortic arch. He was admitted to the Department of Internal Medicine for further evaluation.

Physical examination revealed blood pressure of 130/90 mmHg in the right arm and 128/90 mmHg in the left arm; no differences could be found between the upper and lower extremities. His temperature was 37.8 °C and pulse rate, 84. The head, eyes, ears, nose and throat were normal. The lungs were clear. Heart rhythm was regular, and no murmur could be detected. Examination of the abdomen showed no aneurysm; liver and spleen could not be palpated. The genitalia were normal. Laboratory tests showed that hematocrit was 42.2% and white-cell count was 9,600, with 66% neutrophils, 29% lymphocytes, and 5% monocytes. Platelet
Fig. 5. Gross findings at operation. (a) $5 \times 5 \times 6$ cm mass (fist size) was found protruding from the aortic arch and involving the left subclavian artery. (b) A patch of woven Dacron was sutured to the excised area in the aortic arch. The proximal portion of the left subclavian artery was reconstructed using a Gore-tex graft.

Fig. 6. (a) Photomicrograph of the resected aneurysm shows thickening of intima with thrombi, marked inflammatory cellular infiltration in the media and proliferation of vasa vasorum (HE×40). (b) In an Elastica-van Gieson stained specimen, elastic fibers were found to occasionally disappear in the media (×100).

count was 365,000, and erythrocyte sedimentation rate was 110 mm/h. C-reactive protein was high at 6.0 mg/dl (normal: <0.4 mg/dl). The Wasserman reaction was negative. Blood chemical analysis failed to indicate anything abnormal.

Computed tomography (CT) of the mediastinum (Fig. 2) showed a mass situated adjacent to the aortic arch. The lumen of the mass as well as the aortic lumen were filled with contrast material which had been infused intravenously for CT study. Magnetic resonance imaging (MRI) was conducted with a Toshiba MRT-50A operated at 0.5 T using a cardiac spin-echo pulse sequence. Images were obtained at TR=718 ms and TE=20 ms. In the sagittal plane (Fig. 3), the mass along with a thrombus was seen located above the aortic arch. Intravenous digital subtraction angiography (DSA) of the aorta (Fig. 4) revealed a saccular aneurysm of the distal aortic arch. The patient was treated with prednisone 40 mg/day and ceftazidime.

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Symptoms of fever and left shoulder pain subsequently subsided. He was discharged from Ichinomiya City Hospital on April 4 and referred to the Department of Thoracic Surgery at Komaki City Hospital. Surgery was first scheduled for May 14, but an emergency operation had to be performed on May 11 due to severe chest pain, coughing and delirium, suggesting rapid progressive enlargement of the aneurysm. Operative findings revealed a 5×5×6 cm mass (fist size) protruding from the distal aortic arch adjacent to the orifice of the left subclavian artery and compressing the trachea. It involved the left subclavian artery (Fig. 5-a) and communicated with the aortic lumen at the site of the distal arch. A hole (1.8 cm in diameter) in the aortic wall was present at the neck of the aneurysm. The aneurysm was identified as a pseudoaneurysm which had arisen from a rupture in the aortic wall. The aneurysm containing the left subclavian artery was resected from the aorta, and the aortic wall near the resected site was excised over a wide area. A patch of woven Dacron was sutured to the excised area, and the aortic arch was supported with Teflon felt. The proximal portion of the left subclavian artery was reconstructed using an 8 mm Gore-Tex graft (Fig. 5-b). The aneurysm was opened where both the new and old thrombi could be identified.

Histological findings of the resected aneurysm near the aortic arch revealed thickening of the intima along with thrombi, marked inflammatory cellular infiltration in the media and proliferation of the vasa vasorum (Fig. 6-a). In an Elasticavan Gieson stained specimen, elastic fibers were found to occasionally disappear in the media (Fig. 6-b). These pathological findings suggested that the aneurysm was in fact associated with Behçet's disease.

Postoperative CT showed no aneurysm formations at the site of the distal aortic arch. The patient was discharged at the end of June 1991 and has continued to be under periodic observation.

DISCUSSION

Behçet’s disease is a multisystemic illness which may be accompanied by arthritis (up to 60%), with intestinal (up to 30%), and/or nervous system involvement (15%–30%). The neurologic manifestation of Behçet's disease include sterile meningoencephalitis, brain stem lesions, cranial and peripheral nerve palsies, and cerebellar and long tract sings. Sterile meningitis, subarachnoid and cerebellar hemorrhage occurring 7 years prior to the formation of the aneurysm in this case may have been associated with Behçet's disease.

Cardiovascular involvement in Behçet’s disease occurs in 7–29% of all cases. A review of the literature showed that, of 94 arterial lesions reported, 65% (61 vessels) were aneurysms and 35% (33 vessels) were occlusions. The most common site for an aneurysm to occur was in the abdominal aorta, followed by the femoral and pulmonary arteries. Aneurysms of the thoracic aorta are relatively rare with 14 cases reported in Japan including our case: 7 males and 7 females at a mean age of 47 (range 25–58 years). It took approximately 11.5 years (range 4–24) for aneurysms to form following the initial diagnosis of Behçet’s disease. There were 2 cases of dissecting aneurysms, 8 pseudo-aneurysms and 4 true aneurysms. Eight of these 14 cases did not undergo any operation. Four patients died of a ruptured aneurysm, and one died of gastrointestinal bleeding. Surgery was conducted in 6 patients, who all survived. From the literature, we also found that aneurysms of the abdominal aorta or peripheral arteries tended to occur more frequently in males. In addition, aortic arch aneurysms are extremely rare, with only 3 reported cases in Japan including the present case.

Aneurysms associated with Behçet's disease usually occur relatively early in life and are clinically life threatening due to the high risk of rupture. The pathogenesis for arterial aneurysms in cases with Behçet’s disease may be obliterator endarteritis of the vasa vasorum with consequent dilatation and aneurysm formation, or perforation and pseudoaneurysm formation. Corticosteroid therapy, immuno-suppression, and occasionally fibrinolytic therapy have been carried out palliatively with limited success.

Mundth et al. reported the frequency of ruptured aneurysms at 20% in patients with
no inflammation and 54% in cases with infection. After the diagnosis has been confirmed, an aggressive surgical approach is considered necessary to relieve aneurysms associated with this disease. During surgery, however, radical resection of the affected areas is not always carried out since the aneurysm often adheres tightly to involved veins, and surrounding perivascular tissue, making the operation difficult. Postoperative infection of the graft and hemorrhage from the anastomosis may also sometimes lead to new aneurysms which develop at the anastomotic site. Great care must thus be taken before, during, and after surgery. In our case, aneurysm of the aortic arch enlarged rapidly over a period of 2 months coupled with fever and back pain. The symptoms and inflammatory reaction subsided after therapy with steroids and antibiotics. However, surgical treatment was eventually undertaken because chest roentgenography showed progressive enlargement of the aneurysm. Total arch replacement may be recommended for aortic arch aneurysms in cases with Behçet’s disease to reduce the risk of postoperative suture aneurysm formation. In our case, however, we decided to resect the aneurysm and suture a patch of woven Dacron to the excised area and to support the aortic arch with Teflon felt since the patient was critically ill, requiring an emergency operation. Moreover, we anticipated this case would experience cerebral ischemia during reconstruction of the cerebral and cervical vessels and have poor surgical results of the total arch replacement due to earlier subarachnoid and cerebellar hemorrhage which had been followed by a V-P shunt.

In diagnosing aneurysms, axial transverse CT with an infusion of contrast material may help to clarify any abnormal mediastinal configuration or mass. In our case, CT of the mass before and after infusion of contrast material was the same as CT visualization of the aortic arch. MRI has proved to be of great value for elucidating various disorders of the aortic arch and thoracic aorta since it allows for direct multiplanar imaging. Sagittal projection is particularly useful for demonstrating aneurysms of the thoracic aorta, aortic wall and mural thrombus. Intravenous DSA can clearly demonstrate aortic anatomy and is as safe as conventional angiography. These methods are considered quite useful in cases with Behçet’s disease because aneurysm formation after arteriography may occur at the site of the arterial puncture. CT or MRI may also be of great value for diagnosing aneurysms at central locations which are ordinarily inaccessible during physical examination in the early stage and may also be used as a follow-up to check for graft patency and for possible new aneurysm formations at the anastomotic sites.

Physicians should always be aware of the diverse clinical manifestations of Behçet’s disease, especially the vascular complications. An early diagnosis and major surgery are necessary to successfully relieve aneurysms in the treatment of this disease.

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