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Eosinophilic Arteritis with Coronary Aneurysms and Stenoses
Paul B. Poommipanit, MD, Mark Lensky, MD, Jonathan Tobis, MD

A 43-year-old Chinese woman with eosinophilia and a swollen neck was found to have extensive aneurysmal dilatation of both carotid arteries. Although there were no anginal symptoms, cardiac investigation revealed an abnormal radionuclide perfusion study. Cardiac catheterization showed large coronary aneurysms with complete occlusion of the left circumflex (LCx) and right coronary arteries (RCA), and 90% stenosis of the mid-left anterior descending (LAD) artery with an ejection fraction (EF) of 25%. Discussion of this unusual form of eosinophilic arteritis and its treatment are presented.

Case Report. A 43-year-old Chinese woman, with a medical history of hypertension and severe patchy alopecia for two years, complained of painless “fullness of the neck” that developed six months prior to her hospitalization. A magnetic resonance imaging (MRI) study revealed aneurysmal carotid arteries with mural thrombi (Figure 1). The patient was started on anti-hypertensive medications as well as coumadin. A few days later, she developed sudden onset of right arm and leg weakness and was found to have a left parietal-occipital infarct by MRI. The patient’s white count was elevated at 11,000 with 65% eosinophilia. She was given high-dose steroids, and the eosinophilia resolved.

The patient did not report any anginal symptoms, but because of the carotid involvement as well as symptoms of dyspnea on exertion and an elevated B-type naturetic peptide (BNP) of 1,140 pg/ml, an adenosine Myoview™ (Amersham Health, Waukesha, Wisconsin) study was performed. This showed large areas of ischemia and infarction in the anterior and inferolateral walls. Coronary angiography revealed diffuse aneurysmal dilatation of all three coronary arteries, associated with complete occlusion of the left circumflex artery (LCx), right coronary artery (RCA), and 90% stenosis of the mid-left anterior descending artery (LAD)(Figures 2A and B).

The EF by ventriculography was 25% and demonstrated global hypokinesis. Right ventricle biopsy obtained two weeks after the institution of steroid therapy revealed no evidence of inflammatory infiltrate or eosinophilic myocarditis. A chromium cobalt 3.0 x 18 mm stent (Vision, Guidant Corp., Indianapolis, Indiana) was positioned across the LAD stenosis and was deployed at a pressure of 16 atm (Figure 3). Because the LAD represented the patient’s sole remaining coronary circulation, the procedure was performed with intra-aortic balloon pump (IABP) support. Neither the abdominal aorta nor the renal arteries were affected by the aneurysms.

She was continued on high-dose corticosteroid therapy and subsequently received one dose of IV cyclophosphamide. After discharge from the hospital, she returned to her usual daily activities, with some residual weakness in the right arm and leg, but was clinically stable nine months later.

Discussion

1. Vasculitis (Table 1). There are several vasculitides that affect large- and medium-sized arteries which share features with this case. Takayasu’s arteritis affects the aorta and its branches, which include the carotid and coronary arteries.1 Our patient did not have involvement of the aorta or the subclavian branches. Takayasu’s arteritis is also not associated with eosinophilia.2 Giant cell arteritis involves the large and medium vessels,
but most often involves the cranial branches of the aorta, and is also not associated with peripheral eosinophilia. Churg-Strauss arteritis, the vasculitis most commonly associated with peripheral eosinophilia, affects medium- and small-sized vessels and normally is associated with allergic rhinitis, sinusitis or asthma. There are case reports of Churg-Strauss coronary arteritis, but none involving both the coronary and carotid arteries. Our patient did not have typical symptoms of asthma, rhinitis or sinusitis, although the alopecia was felt to represent an auto-immune phenomenon. Polyarteritis nodosa involves the medium and small muscular arteries and occasionally leads to coronary artery disease and aneurysms, but does not involve the carotid arteries and is not associated with peripheral eosinophilia. The most notable vasculitis to affect the coronary arteries is Kawasaki’s disease. Kawasaki’s disease, however, occurs predominantly in children, presenting with fever lasting several days. It is commonly associated with “strawberry” tongue, palmar erythema, rash, or cervical lymphadenopathy. The vasculitic syndrome affecting this patient may represent an eosinophilic variant of Takayasu’s arteritis versus Churg Strauss arteritis with involvement of medium and large vessels, however it is difficult to specify an exact category beyond an “eosinophilic vasculitis.”

2. **Coronary angioplasty in aneurysmally dilated and stenotic arteries.** Our case report describes successful stenting of a narrow lesion in the sole remaining coronary vessel that was severely affected by aneurysmal dilatation and inflammation secondary to eosinophilia. The case was also unusual because of the lack of symptoms of angina or infarction at presentation to suggest any involvement of the coronary arteries. The management was complicated by the absence of distal targets for possible bypass grafting of the RCA or LCx, and the high risk associated with open heart surgery in this setting. The coronary angioplasty was technically difficult due to the presence of extensive aneurysmal dilatation of the LAD. The risk of the procedure was high because the LAD was the sole remaining source of myocardial blood supply. There have been a number of case reports documenting treatment of coronary artery aneurysms with stent implantation, although alternative therapies have been surgical excision or coronary artery bypass grafting.

3. **Vasculitis involving the coronary arteries.** The mechanism behind the changes that produce coronary artery stenosis in vasculitis involves immunologically mediated inflammation with intimal thickening from the accumulation of fibrous tissue and proliferation of smooth muscle cells. There is also pathologic evidence that stimulated eosinophils are directly toxic to myocardial cells and arterial wall components. Aneurysmal dilatation is associated with destruction of the media, as observed in the intravascular ultrasound image (Figure 3). This report is unusual because it describes a nonspecific eosinophilic vasculitis associated with severe coronary and carotid artery aneurysm and stenosis formation in which a sole remaining LAD stenosis was successfully treated with percutaneous revascularization.
<table>
<thead>
<tr>
<th>Classification</th>
<th>Clinical Features</th>
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<tbody>
<tr>
<td>Takayasu's arteritis</td>
<td>Usually affects the aorta and its branches, which include the carotids and coronary arteries.</td>
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<tr>
<td>Giant cell arteritis (Temporal artery)</td>
<td>Involve the large and medium vessels, but most often involves the carotid branches of the aorta.</td>
</tr>
<tr>
<td>Polyarteritis nodosa</td>
<td>Typically involves the small and medium sized arteries and occasionally leads to coronary artery disease and aneurysms.</td>
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<tr>
<td>Kawasaki's arteritis</td>
<td>Involved the large, medium and small arteries including the coronary arteries, but usually occurs in children.</td>
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<tr>
<td>Churg-Strauss arteritis</td>
<td>Most commonly associated with eosinophils and involves the medium and small sized arteries.</td>
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<tr>
<td>Wegener's granulomatosis</td>
<td>Involve the medium and small arteries and typically involve the upper and lower airways as well as glomerulonephritis of the kidneys.</td>
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**References:**


