A Case of Isolated Thoracic Aortic Aneurysm as a Manifestation of Undiscovered Giant Cell Arteritis

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Case Report

A Case of Isolated Thoracic Aortic Aneurysm as a Manifestation of Undiscovered Giant Cell Arteritis

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Abstract A 73-year-old woman was referred to our hospital to investigate dilatation of an aortic arch which had been detected by a chest roentgenogram and severe aortic valve regurgitation detected by echocardiography. On admission, a computed tomography scan of the chest showed a large fusiform ascending aortic aneurysm. She had not shown any symptoms such as headache or polymyalgia rheumatica and had no significant coronary atherosclerosis. She underwent aneurysmectomy and reconstruction of the ascending aorta using cardiopulmonary bypass without aortic valve replacement, and pathological examination of the aneurismal wall revealed giant cell arteritis (GCA). Preoperatively, she did not have any temporal pain, and no signs of inflammation were detected serologically. Postoperatively, aortic valve regurgitation improved and she did well. However, three months after the surgery, she died suddenly due to the rupture or dissection of aorta. In the Japanese population, GCA is reportedly a rare cause of aortic aneurysm. However, retrospective studies show that GCA affects the aorta and that thoracic aortic aneurysm is a possible complication of GCA. In cases of the thoracic aortic aneurysms with unknown etiology, there is a possibility that GCA is the cause of the aortic aneurysm.

Introduction

Giant cell arteritis (GCA) is a systemic vasculitis that primary affects extracranial arteries of the head and neck in the elderly people. It may present with signs and symptoms of a systemic disease and secondary to vascular ischemia. Headache, jaw claudication, fever, polymyalgia rheumatica, and an elevated erythrocyte sedimentation rate (ESR) are typical clinical features. The treatment of choice is therapy with corticosteroids, which usually prevent severe complications such as blindness.

The diagnosis of GCA is usually performed by a biopsy of the temporal artery. It is calculated that approximately 1% of the population >50 years of age may be affected. However, there also has been an increase in the number of reports of late complications due to involvement of the aorta, the arteries of the upper and lower limbs, or of other districts. In cardiology, there are few reports of the cardiovascular disease complicated with GCA. However, GCA has the possibility to be
Giant Cell Arteritis and TAA

We report a case of an ascending thoracic aortic aneurysm with GCA in elderly woman.

Case Report

A 73-year-old woman with hypertension for 15 years, in whom dilatation of an aortic arch detected by a chest radiograph and aortic valve regurgitation detected by an echocardiogram had been diagnosed at 3 years previously and both of them had been worsened gradually, was referred to the Department of Cardiology Center of our hospital. She had not shown any symptoms of temporal arteritis such as headache or polymyalgia rheumatica, and a biopsy of temporal artery has not been performed. She had a mild dyspnea on effort and NYHA class was II. Physical examination revealed a blood pressure with the decrease in diastolic blood pressure of 140/40 mmHg in both arms, heart rate of 72 beats per minute and a Levine 3/6 grade aortic insufficiency murmur. The chest radiograph showed a cardiomegaly (cardiothoracic rate was 57.0%) and a dilatation of the arch of the aorta involving the ascending tract (Fig. 1a). An electrocardiogram revealed normal sinus rhythm (68 beats per minute), normal axis and left ventricular hypertrophy without strain pattern (RV5+SV1=42mm). A two-dimensional echocardiogram showed marked dilatation of the root of the aorta (38mm) and severe aortic valve regurgitation (regurgitant jet diameter = 73% of the outflow tract diameter associated with diastolic flow reversal in the abdominal aorta). However, aortic valve leaflets appeared to be quite normal. A computed tomography scan of the chest and aortography confirmed the presence of a large fusiform thoracic ascending aortic aneurysm (the maximum dimension of the aneurysm was 7.3cm), with diffuse dilatation of the innominate artery, the aortic arch and the descending aorta (Fig. 1b). A right heart catheterization revealed normal right atrial pressure 2 mmHg; normal right ventricular pressure 30/5 mmHg; normal mean pulmonary wedge pressure 6 mmHg; normal pulmonary artery pressure 32/6 mmHg. The thermodilution cardiac index was 3.38 ml/min/M2. The left ventriculogram revealed a dilatation of left ventricular (left ventricular end-diastolic volume index 107 ml/M², left ventricular end-systolic volume index 31ml/M²). The left ventricular end-diastolic pressure was 15mmHg. Coronary angiography showed no significant atherosclerosis in the coronary arteries. The laboratory tests included the following: white blood cell count of 5,710 / mm³ (with 50.0% neutrophilia,
37.2% lymphocytes, 4.3% of eosinophils and 0.6% basophils), red blood cell count of 2.85x106/mm³, hemoglobin concentration of 9.3g/dl, hematocrit of 27.2%, platelets count of 19.4x10⁴/mm³, total serum protein concentration of 7.2g/dl, serum albumin concentration of 4.2g/dl, 22 IU/L aspartate aminotransferase, 16 IU/L alanine aminotransferase, 215 IU/L lactate dehydrogenase, 22mg/dl blood urea nitrogen, 1.3mg/dl creatinine, 0.1mg/dl C-reactive protein, 165 IU/L creatine kinase (CK) (with 4% CK-MB). ESR was not determined. The level of brain-natriuretic peptide (BNP) before the surgery was 167mg/dl.

She had the large thoracic ascending aortic aneurysm with severe aortic valve regurgitation, and we considered that aneurysmectomy and/or aortic valve replacement was necessary to prevent the rupture of the aneurysm and heart failure due to aortic valve regurgitation. The patients underwent aneurysmectomy and reconstruction of the ascending aorta using cardiopulmonary bypass without aortic valve replace-

ment, and pathological examination of the aneurysmal wall revealed several epithelioid cell granulomas with giant cells and inflammatory infiltrate mainly in luminal side of the media (Fig. 2a, 2b, 2c). The Immunohistochemistry suggest that CD68 is positive in histocytes (Fig. 2d). These features suggest giant cell arteritis (GCA). Postoperatively, she had no significant complications, and the echocardiogram revealed the reduction of the size of left ventricular (left ventricular dimension diastolic/systolic from 78/52 to 58/36mm) and the improvement of aortic valve regurgitation from severe to mild (regurgitant jet diameter was 12% of the outflow tract). One month after the surgery, BNP was also decreased from 167mg/dl to 122mg/dl. She did well for three months after surgery (NYHA class I), and blood pressure was well controlled. However, three months after the surgery, she was found with the asystole by her family, and transported to the emergency room of our hospital. Electrocardiograph revealed cardiac arrest and there was no

Fig. 2 (a), (b), (c) The aneurysmal wall showing several epithelioid cell granulomas with giant cells and inflammatory infiltrate mainly in luminal side of the media (hematoxylin-eosin), original magnification (a) x20, (b) x64, (c) x128. (d) Immunohistochemistry showing CD68 (+) in histiocytes, original magnification x164.
respiration. Her pupils were dilated and she did no have light-reflex of pupils. Chest X ray, echocardiograph and puncture of the pleural and pericardial effusion revealed massive bloody pleural and pericardial effusion. There was no response for the cardiopulmonary resuscitation, and we confirmed her death.

Discussion

This case showed an ascending thoracic aortic aneurysm with giant cell arteritis in elderly woman. She had not shown any symptoms of temporal arteritis such as headache or polymyalgia rheumatica, so we did not check ESR, and a biopsy of temporal artery was not performed before the operation. The chest radiograph, aortography, echocardiography and CT scan revealed a dilatation of ascending aorta with severe aortic valve regurgitation. She underwent aneurysmectomy and reconstruction of the ascending aorta using cardiopulmonary bypass without aortic valve replacement, and pathological examination of the aneurismal wall revealed giant cell arteritis. For three months after surgery, she did well with well-controlled blood pressure. However, three months later after surgery, she died suddenly due to the rupture or dissection of aorta.

In this case, a Bentall operation was not performed. After the surgery, aortic valve regurgitation was improved, left ventricular size was decreased, and BNP was significantly decreased. Among the patients with ascending aortic aneurysm due to GCA, recent study suggest that 51% of patients had moderate to severe aortic regurgitation, and 81% of patients underwent graft replacement of the ascending aorta, and 11% of patients had a modified Bentall procedure. Whether we must do the Bentall operation for ascending aortic aneurysm due to GCA to prevent the aortic rupture or dissection is necessary for further investigation. Recent study suggest that a Bentall procedure using a composite valve conduit was used to replace the aortic root in patients with sino–tubular junction dilatation and an abnormal aortic valve and that patients with preserved sino–tubular junction dimensions with an abnormal valve had an aortic valve repair or replacement in addition to replacement of the ascending aorta. Moreover, in that study, if the valve was deemed normal with dilatation of the root, a valve-sparing root reconstruction was performed with either resuspension of the valve or reconstruction of the sinuses and reimplantation of the coronary arteries. In this case, we considered that the Bentall operation was not necessary because the aortic valve was normal with dilatation of the root. Between and after the surgery, this patient had no significant complications, such as bleeding, infection or heart failure, and just before her sudden death, she did well. We did not do the treatment with steroid after surgery, because she did not show either symptomatic vasculitis or elevated markers of inflammation. Whether aneurysmal dilatation of the remaining aorta and great vessels can be prevented by an aggressive regimen of steroids is unclear. We have the speculation that she might have the rupture around the suture site between the aorta and graft, or the dissection. The reasons why she had the rupture or dissection suddenly at 3 months after the surgery and whether the procedures and management of the surgery were correct or not could not been determined. However, in the cases of the aortic aneurysm with unknown etiology, we consider that the biopsy of temporal artery may
be useful to determine whether the etiology is GCA or not.

The immunohistochemistry suggest that CD68 was positive in histocytes. Previous study reported that the formation of new capillary blood vessels within the arterial wall affected by GCA is regulated by giant cells and CD68-positive macrophages by inflammatory cells. Although the patient in this case did not have any symptoms of polymyalgia rheumatica, this result of immunohistochemistry suggest that the ascending aortic aneurysm was due to GCA in this case. We must consider that the patients of ascending aortic aneurysm with the low risk factors of atherosclerosis have the possibility of GCA.

Retrospective studies show that GCA affects the aorta and its major branches in 15% of cases. In 75 to 83% of these patients, symptoms referable to large artery involvement usually occur 2 to 7 years after diagnosis, whereas in the remaining 17 to 25%, they are the presenting complaints of the disease. Thoracic and abdominal aortic aneurysms, due to aortitis, and dissection are feared complications of GCA. Compared with persons of the same age and sex, patients with GCA are 17 times more likely to develop a thoracic aortic aneurysm and 2.4 times more likely to develop an abdominal aortic aneurysm. Aortic valve insufficiency is also possible. Recent population based retrospective cohort study reported that older adults with GCA appear to be at increased risk for developing cardiovascular disease. In this case, we could not determine the etiology of the thoracic aorta aneurysm before the aneurectomy, and because she had not shown any symptoms of temporal arteritis such as headache or polymyalgia rheumatica, we did not perform a biopsy of temporal artery.

In the laborotory findings of GCA, ESR is usually higher than 50mm/hour, but a lower ESR is possible. ESR lower than 50mm/hour is present in about 5% of patients with GCA. However, a completely normal ESR (lower than 30mm/hour) seems to be exceptional. Because she had not shown any symptoms of temporal arteritis such as headache or polymyalgia rheumatica, we did not check ESR in this case.

Recent reports suggest that patients with a typical history of temporal arteritis and/or polymyalgia rheumatica should be closely followed-up by cardiologists to ensure the early detection of large-vessel pathol-oogy, which could be responsible for life-threatening complications such as aortic dissection and/or rupture, severe aortic incompetence, and ischemic syndrome of the upper or lower extremities. Thoracic aortic dissection in GCA is associated with markedly increased mortality. Moreover, when cardiologists are faced with an aortic aneurysm in older individuals, particular women, temporal arteritis should be at least suspected and other elements sought in the patient’s clinical history in order to ascertain the nature of the aortopathy. An investigation of the superficial temporal arteries by duplex ultrasonography may also be of value.

The American college of Rheumatology proposed a list of criteria for diagnosis of GCA, including the following five criteria: 1) Age at onset over 50 years, 2)New headache, 3)Temporal arteritis abnormalities, 4) ESR over 50mm/hour, 5) Positive temporal artery biopsy. The presence of three or more criteria had a sensitivity of 97.5% and a specificity of 78.9% in a French study of patients in whom the diagnosis of GCA was confirmed or ruled out by a temporal artery
biopsy\(^3\). In this case, the age was over 50 years old, but she did not have headache and temporal arteritis abnormalities, and was not performed ESR check and temporal artery biopsy. In the cases of the aortic aneurysm with unknown etiology, we have better check ESR and perform temporal artery biopsy to rule out GCA.

In conclusion, we experienced the ascending aortic aneurysm with GCA in elderly woman. She had not shown any symptoms of temporal arteritis, and pathological examination of the aneurysmal wall after aneurysmectomy revealed GCA. In cardiology, GCA as the etiology of aortic aneurysm may be underestimated, and in the cases of the thoracic aortic aneurysm with unknown etiology, there is a possibility that GCA is the cause of aortic aneurysm.

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**References**


15) Richardson MP, Lever AM, Fink AM, Dixon AK and Hazleman BL: Survival after aortic dissection in giant cell arteri-


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手術により巨細胞性動脈炎の関与が明らかとなった胸部大動脈瘤症例

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胸部エックス線写真での大動脈弓拡大と心臓超音波検査での大動脈弁閉鎖不全にて受診した73歳女性。胸部CT検査にて、上行大動脈に径7 cmの大動脈瘤があることが明らかとなった。術前の各種検査において、全身の動脈硬化病変は冠動脈脈含めてほとんど認められていないかった。心臓外科にて人工心肺下で上行大動脈瘤切除・人工血管置換術を施行された（大動脈弁置換術は施行せず）、その際に切除した上行大動脈瘤の組織所見から巨細胞性肉芽腫が認められ、巨細胞動脈炎が基礎疾患であることが明らかとなった。術前には頭痛やリウマチ性多発筋炎の症状・血沈亢進・CRP陽性がなく、またそれらを含めて側頭動脈炎を示唆する所見がなく動脈生検を行っていなかったため、予測は困難であった。術後は大動脈弁閉鎖不全も改善し、術後経過良好で退院となった（しかし、術後3ヶ月目に吻合部破裂によるものと思われる突然死となった）。日本においては、大動脈瘤の原因として巨細胞性動脈炎の報告例は非常に少ない。しかし、これまでの海外での報告例や剖検例での検討では、巨細胞性動脈炎は大動脈瘤を引き起こすことはまれではない。胸部大動脈瘤の患者において、動脈硬化が軽度などで原因が特定できない場合には巨細胞性動脈炎によるものの可能性がある。