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Case Report

Rupture of Ascending Thoracic Aortic Aneurysm in Postpartum: 2 Cases Report

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Thoracic aortic aneurysms are less common than abdominal aortic aneurysms, however they are life-threatening and usually asymptomatic until acute complications occur. The majority of thoracic aorta aneurysm are associated with medial degeneration rather than atherosclerosis and the fusiform aortic aneurysm is common. Considering that it usually occurs during the sixth and seventh decades of life, its occurrence in a peripartum woman is unusual. Aortic dissection or thoracic aortic aneurysm with aortic insufficiency during pregnancy or peripartum has been reported, however, to our knowledge, the case of ascending thoracic aortic aneurysm in peripartum women, with saccular type without aortic valve involvement but not diffuse dilatation, has not been reported. Herein, we presented two autopsy cases of ascending thoracic aortic aneurysm during postpartum.

Key Words : Thoracic aorta, Aneurysm, Peripartum

Introduction

Rupture of aortic aneurysm is a life-threatening condition. And thoracic aortic aneurysms are usually asymptomatic and not easily diagnosed unless an acute catastrophic complication develops.[1)] Because it is rare in young adults, especially in females during peripartum, considering that it occurs most commonly in the sixth and seventh decade of life and males are predominant,[2, 3)] the fatal sudden and unexpected death following rupture of thoracic aortic aneurysms may bring about a medicolegal dispute.

Thoracic aortic aneurysm is typically fusiform and the saccular type is very rare except aneurysms of infectious or traumatic types.[4)] The saccular type of aneurysm in ascending aorta in females during peripartum has not been known to our knowledge,

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although aortic dissections or aortic aneurysm with bicuspid aortic valve or aortic valvular insufficiency during pregnancy or peripartum, has been known.[5, 6] Therefore we present two cases of rupture of ascending thoracic aneurysm during postpartum, with review of literature.

Case Description

Case 1

A healthy 39-year-old woman who received regular antenatal check-up, came to a local clinic for delivery. She was a nullipara and did not have any specific findings during antenatal follow-up. Caesarean section was performed, because fetal distress was strongly suspected due to cephalo-pelvic disproportion (CPD) while fetal monitoring. The baby was born as a healthy girl and Apgar scores at 1 and 5 minutes were all 9 points. After delivery, her condition was getting better. But, on the 4th day after delivery, she complained of headache on both sides of temporal region, and pitting edema on both of her lower legs. Her vital signs showed an elevated blood pressure of 150/110 mmHg, the pulsation rate of 56/minute, and the respiration rate of 20/minute. After two hours, her vital sign showed no change. On neurological examination, no specific signs were identified. Electrocardiogram also showed no abnormalities. The blood test of CK-MB and troponin-I were all within normal limits. Nifedipine, furosemide and acetaminophen were administrated to control her unstable state. About 8 hours later, she was found to be unconscious and transferred to the university hospital. At arrival, her mental status was drowsy, the blood pressure was 80/40 mmHg, blood sugar was 86 ㎎/㎗, and O2 saturation was 90%. Despite cardiopulmonary resuscitation, she did not respond and expired. It was the 6th day after delivery.

There was no injury on external examination. On internal examination, the pericardial sac was filled with blood and clots (about 500 ㎖) but the weight of the heart was normal (350 g). A saccular type of aneurysm (4.0×4.0×2.0 ㎝) was identified at the ascending aorta just superior to coronary sinus. A small perforation (0.1×0.1 ㎝) was identified in the aneurysmal wall (Fig. 1). The coronary arteries and myocardium showed no pathologic findings. The descending aorta, abdominal aorta and other great vessels also revealed no specific findings. Other internal organs showed no significant findings except the uterus and adnexa showing typical postpartum findings and multiple intramural leiomyomas.

On histologic examination, the aneurysmal wall of ascending thoracic aorta showed thinned aortic wall of

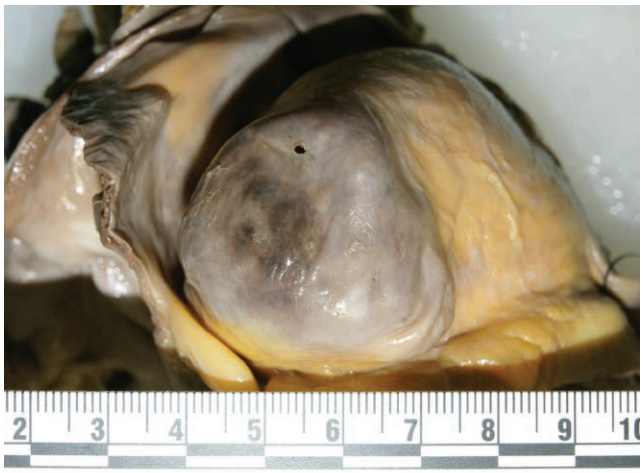

Fig. 1. The saccular type aneurysm with rupture is observed on the ascending aorta and aortic wall cut longitudinally showing true aneurysm (Case 1).

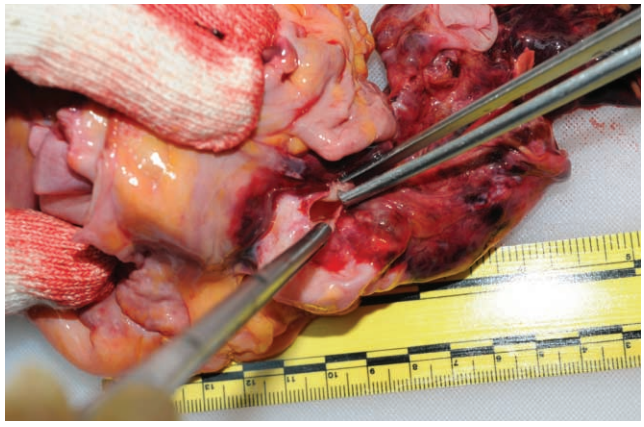

Fig. 2. The ruptured aneurysm is identified on the ascending aorta, with localized mild atherosclerotic changes (Case 2).

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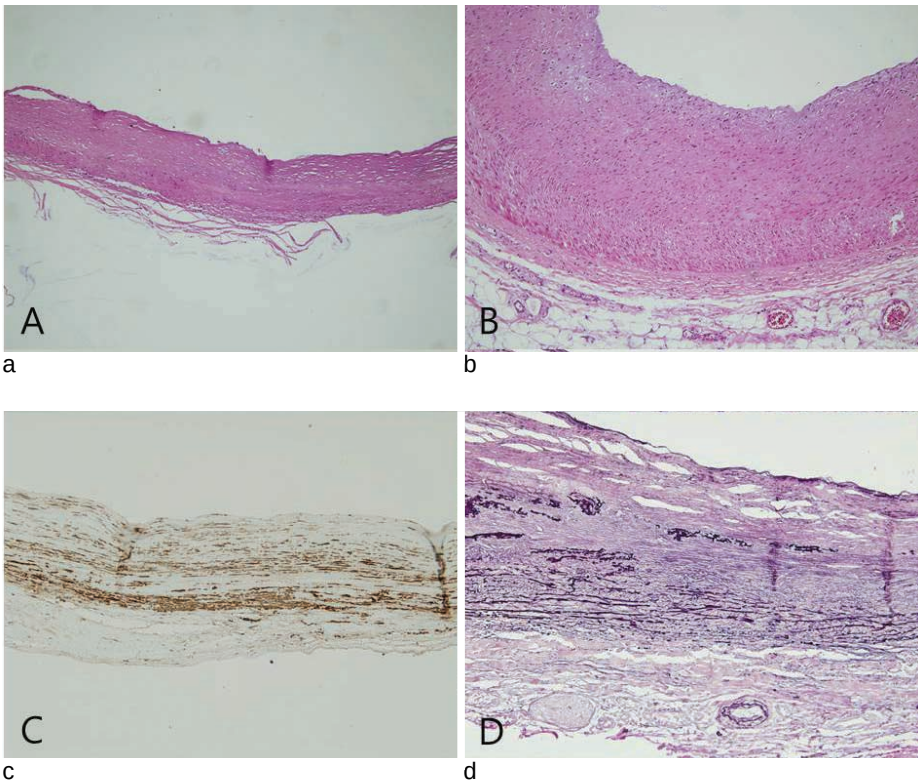
all 3 layers (Fig. 3a & 3b). The aortic wall adjacent to the perforation site revealed hemorrhage with mild inflammation. It showed no evidence of dissection, and cystic medial necrosis, which is characterized by myxoid degeneration of media with deposition of gelatinous or mucoid substance. And there was also mild atherosclerosis. The immunohistochemical staining for smooth muscle actin was positive for myofibroblast in media, supporting that it was true aneurysm (Fig. 3c). Disrupted elastic laminae were observed on the special staining for elastic fiber (Fig. 3d). Toxicological test was negative.

Case 2

The deceased was a 35 years old woman who had two healthy children. She was also pregnant and gave a birth to a third baby. During antenatal period and delivery, she had no problem and was discharged. Early in the morning, on the 13th day from delivery, she complained of unexpected left chest pain and dyspnea while she nursed a baby, and became unconscious for a moment. She was transferred to a hospital. On physical examination, the characteristics of her pain were neck tightness or squeezing of the chest and neck. On echocardiogram, moderate to severe pericardial effusion, collapse of right ventricle during diastole, and normal heart chamber with good global left ventricular function were observed. The blood test showed that hemoglobin was 11.9 g/*dl*. Although she recovered her consciousness, she was admitted to the hospital under the impression of postpartum pericarditis and received supportive treatment with careful observation. During admission, her pain improved and because there was no other symptom or sign, she was to be discharged. While she talked to her friends about her discharge, she unexpectedly lost her consciousness again, and expired in spite of cardiopulmonary resuscitation (CPR). Bloody pericardial fluid was drained from the emergency pericardiocentesis, and the blood test showed hemoglobin level of 5.8 g/*dl*.

On external examination, conjunctivae and oral mucosae were pale. There was no injury except



a b c d

Fig. 3. The wall of the aneurysm is extremely thin (a: H & E, × 100) compared with normal aortic wall (b: H & E, × 100), but is composed of all 3 layer of vessel wall. Immunohistochemical staining for smooth muscle actin is positive for myofibroblast in media, which support the aneurysm is a true aneurysm (c: SMA, × 200). Special stain for elastic fiber shows disrupted elastic laminae (d: elastin, × 200).

needle marks due to pericardiocentesis and intravenous injection. On internal examination, there were widespread hemorrhages; hemothorax in both sides of chest cavity (right about 600 *ml*, left about 700 *ml*), hemopericardium (about 100 *ml*), extending to posterior mediastinum and retroesophageal space. The saccular type of aneurysm with perforation was identified on the posterior wall of the ascending aorta which was just above the aortic valve (Fig. 2). On the histologic examination, the aneurysmal wall showed thinned aortic wall (true aneurysm) with hemorrhage and the adjacent aortic wall showed mild atherosclerosis and focal bluish areas suggesting accumulations of proteoglycans. But there was no evidence of dissection or cystic medial necrosis in the aorta. The coronary arteries and sinoatrial & atrioventricular nodes were normal. There was not fibrosis or necrosis on myocardium. Local fractures of bilateral ribs, and localized superficial laceration of liver with peritoneal hemorrhage (about 400 *ml*) were identified, which were assumed to be a secondary findings by CPR. Internal organs were also pale. The liver showed moderate fatty change but other organs had no specific findings. Toxicological test was negative.

Discussion

Aneurysm development denotes that there are some underlying medial changes which cannot prop up the high systolic blood pressure. There are three pathological processes of medial changes: Atherosclerosis, noninflammatory medial 'degeneration' of the media, and inflammation of aorta.[7] Atherosclerosis is related to 1% of ascending thoracic aortic aneurysm[4] and it is less common than abdominal aorta,[1] although the relative proportion of each type turns on the patient population, methods of classification and inclusion criteria.[8] In our cases, the aneurysms also showed only focal and mild atherosclerosis, which is not associated with the perforation site. Typically, thoracic aneurysms occur most commonly in the sixth and seventh decade of life, predominantly in males.[3, 4] However, the deceased in our cases were females in their 30s without specific symptoms or clinical features of aortic aneurysm.

Noninflammatory medial 'degeneration' of the aortic media is presumed to be the major culprit of most thoracic aortic aneurysm as well as dissections.[7] Noninflammatory medial 'degeneration' is known as medial necrosis but it is a misnomer. It is characterized by disruption and loss of elastic fibers and increased deposition of proteoglygans, and there are typically areas of loss of smooth muscle cells in the media.[1] In one of our cases, there were focal bluish areas suggesting deposition of proteoglycans, but not typical findings of cystic medial degeneration. Nevertheless, such histological changes may occur to a certain degree in any aorta with aging dilatation and even in cases of thoracic aneurysms, the media appears histologically normal as it is in one of our cases.[4, 7] The marked medial degeneration is known to be found in patients with hereditary connective tissue diseases such as Marfan's disease. But such severe changes can be also found in aortic root dilatation, and even in a postinflammatory change in aortitis.[4-7] The significance of these histological findings should be further studied, even though the value of a diagnostic clue seems to be controversial.

For some portion of aortic aneurysm, genetic factors such as connective tissue diseases can contribute to its occurrence.[4, 7, 8] Our cases did not show any specific symptoms or characteristic features implying the possibility of connective tissue diseases. Their hearts did not show any anatomical abnormality. There was no valvular involvement by aortic aneurysm, and bicuspid valve or mitral valve prolapse is not observed, either. And in their family histories, there were no specific past histories.

'Noninflammatory medial degeneration' was initially coined in order to be extricated from infectious aortic disease. The biochemical pathways and proteins involved with medial degeneration have not been clearly delineated, even though some

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literature supports the presence of inflammatory process.[1, 9, 10] A subset of matrix metalloproteinases (MMP-2, MMP-9) in the media of thoracic aortic aneurysms, which is known to have elastolytic activity, is related to medial degeneration. For accumulation of proteoglycans in the aortic media, no studies have determined the exact mechanism or pathophysiology. Loss of smooth muscle cells appeared to be related to an initial adaptive response to minimize increased wall stress resulting from vascular dilatation.[1]

Aneurysm (or true aneurysm) is defined as a permanent localized dilatation of an artery, having at least a 50% increase in diameter compared to the expected normal diameter of the artery in question.[1] Among the types of thoracic aneurysms, the fusiform is most common, and only infectious aneurysms and posttraumatic pseudoaneurysms are typically saccular.[4] The saccular type which came from noninflammatory medial degeneration is rare, although a case with a saccular aneurysm of ascending aorta was introduced as a representative example.[4] Ectasia is defined as arterial dilation less than 150% of normal arterial diameter,[1] which might be considered as fusiform. It should be differentiated from annuloaortic ectasia which involves aortic valve, resulting in aortic valve dilation and aortic insufficiency. And the term is used only in describing thoracic aneurysm which is formed by noninflammatory medial degeneration. Because our cases were all saccular types of aneurysm with no involvement of aortic valve, they are not annuloaortic ectasia but also have distinct and unusual features compared to conventional cases. To the best of our knowledge, the saccular type of aneurysm in ascending aorta in postpartum females has not been reported. Both hemodynamic and hormonal physiologic changes in pregnancy is assumed to play some roles in the underlying process, but it is not clearly determined why and how the aneurysm occurred.

Thoracic aortic aneurysm has no specific clinical features and it can be ignored or misdiagnosed, even by specialists, and may lead to a medicolegal dispute. In our cases, the clinicians could have not done the initial diagnosis appropriately. Because the deceased's symptoms were nonspecific, their ages are unusual for aortic diseases, and the clinical course was aggravated very rapidly. Finally, they might have had an impression of the possibility of aortic diseases but it was too late to perform proper medical treatment. Therefore, we present these rare and unusual cases of ascending thoracic aortic aneurysms in females during peripartum.

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