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CASE REPORT

PATHOLOGY/BIOLOGY

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Descending Thoracic Aortic Aneurysm Rupture During Postpartum Period

ABSTRACT: Aortic aneurysm refers to the pathological dilatation of the normal aortic lumen involving one or several segments. Thoracic aortic aneurysms are much less common than aneurysms of the abdominal aorta. Descending thoracic aortic aneurysm leading to dissection and spontaneous rupture is a potentially catastrophic illness. Although rare, dissection and rupture of a preexisting aortic aneurysm have been reported during pregnancy and early puerperium. To the best of our knowledge, such cases among young pregnant women are rarely reported in literature. Herein, an autopsy case of spontaneous rupture of a clinically undiagnosed descending thoracic aortic aneurysm during early puerperium in a young woman is presented along with the review of relevant literature. The victim was found dead on her hospital bed on the seventh day of puerperium. Autopsy with ancillary investigations revealed that the young woman died because of hemothorax from a ruptured dissecting descending thoracic aortic aneurysm secondary to chronic aortitis.

KEYWORDS: forensic science, forensic pathology, autopsy, aortic aneurysm rupture, pregnancy, maternal, postpartum

Dissecting thoracic aortic aneurysm leading to spontaneous rupture is an uncommon and potentially catastrophic illness. The overall incidence of thoracic aortic aneurysm is estimated to be around six per 100,000 patient-years (1). The incidence of aortic dissection ranges from 0.5 to 2.95/100,000 per year (2–4). Rupture of an aortic aneurysm represents a life-threatening condition. Most patients die within 6 h after rupture (5). Acute aortic dissection and rupture are rare in the young and usually associated with trauma, Marfan's syndrome, or pregnancy (6–8). Although dilatations and aneurysms have been reported in late phases of aortitis, rupture of dissecting aneurysm of the descending thoracic aorta during puerperium, the period from the delivery of the placenta through the first 6 weeks after the delivery, has not been reported in the young. Herein, we report an autopsy case of rupture of a clinically silent aortic aneurysm of the descending thoracic aorta during puerperium along with review of relevant literature.

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Case Report

An apparently healthy 26-year-old woman was recovering well in a hospital after a full-term lower segmental Caesarean section. She died suddenly on the seventh day postdelivery. Unexpected sudden death in a healthy asymptomatic primigravida compounded with the inability of the treating doctor to provide a reasonable explanation about the cause of death arouse the suspicion of negligence in the minds of the bereaved family which necessitated a medico-legal autopsy at the nearby teaching referral hospital.

Review of the hospital records of the deceased revealed that she had regular antenatal checkups. Lower segmental Caesarean section was necessitated by the premature rupture of the membranes before the onset of labor pains. Both operative and postoperative periods were uneventful till she died suddenly. The victim did not have any history of serious illness, operations, or hospitalization as per the information obtained from her mother and husband.

A forensic autopsy was performed 24 h after death. The victim appeared to be well developed and nourished. A partially healed transversely placed Caesarean section mark was evident over the lower part of the abdomen. Injection marks were present over the front of both the elbows. There was evidence of postdelivery changes in the mammary glands and in the form of vaginal discharge. No evidence of trauma was discernible externally. Marfanoid features in the form of elongated body habitus or pigmented maculae were not seen.

The left thoracic cavity contained 2500 mL of partially coagulated blood. Removal of partially coagulated blood and on searching for any bleeder revealed a saccular aneurysm on the left lateral side of the descending thoracic aorta just below the isthmus corresponding to the body of the fifth thoracic vertebrae. It measured 3.5 in length, 3.5 in width, and 3 cm in depth. A perforating tear

measuring 0.6 × 0.4 cm was present on markedly thin wall of the aneurysm-causing hemothorax. Margin of the perforation showed a minimal separation between intima and media with associated thinning of the aortic wall. The intima of the saccular aneurysm had multiple, hard, glistening, raised yellowish areas with a grating feel on sectioning at places (Fig. 1). The circumference of the aorta at the level of the coronary ostia was 4.9 cm, at ascending aorta 3.5 cm, at isthmus 3 cm, and 4 cm at the level of celiac trunk. Macroscopically, the vessel wall involvement was limited to the proximal end of the descending thoracic aorta. Neither the aortic sinus, arch of aorta, remaining part of descending thoracic aorta, nor abdominal aorta was dilated, and overall, the aorta and its main branches were free of atherosclerosis and other abnormalities. The heart weighed 200 g and had unremarkable musculature, walls, and coronary arteries. The lungs, kidneys, and liver were pale on cut section. Uterus had evidence of lower segmental Caesarean section.

Histologic examination of the descending thoracic aortic aneurysm shows thinned aortic wall with myxoid degeneration characterized by replacement of the connective tissue of the aortic wall by gelatinous or mucoid substance, destruction of elastic fibers, large areas of calcification, intimal chronic inflammatory cell infiltrates, adventitial chronic inflammation, and luminal thrombus suggesting it to be a ruptured dissecting descending thoracic aortic aneurysm secondary to chronic aortitis (Figs 2 and 3). These changes were limited to the saccular aneurysm, and other portions of the aorta and its main branches were essentially intact.

Postmortem toxicological screening of blood and urine did not detect alcohol, common illicit, or prescribed drugs or pesticides. Treponema pallidum hemagglutination assay and venereal disease research laboratory test of the blood were negative. Antibodies against cell nuclei and cytoplasm components (ANA Global Test) assay were negative.

The death was attributed to hemothorax due to ruptured dissecting descending thoracic aortic aneurysm secondary to chronic aortitis.

Discussion

The aorta is a complex vascular structure with many different functions varying along its course. The thoracic aorta provides compliance with elastic recoil to maintain blood pressure and antegrade blood flow throughout diastole. The more distal abdominal aorta functions mainly as a conduit. The varying functions are reflected in the histologic structure of the aorta. The elastin/collagen ratio is highest in the thoracic part and decreases distally. With age, the aortic wall structure changes. Elastin fragmentation, fibrosis, and media necrosis occur in the aorta as signs of aging (9). Furthermore, various diseases alter aortic structure and function and may cause obstruction or dilatation of the aorta (10,11). Both obstruction and dilatation may be circumscript, segmental, or spread throughout the entire aorta (12).

The term aortic aneurysm refers to the pathological dilatation of the normal aortic lumen involving one or several segments. Aneurysms are usually described in terms of their location, size, morphological appearance, and origin. The morphology of an aortic aneurysm is typically fusiform, which is the more common shape, or saccular. A fusiform aneurysm has a fairly uniform shape with symmetrical dilatation that involves the full circumference of the aortic wall. Saccular aneurysms, on the other hand, have more localized dilatation that appears as an outpouching of only a portion of the aortic wall (13).

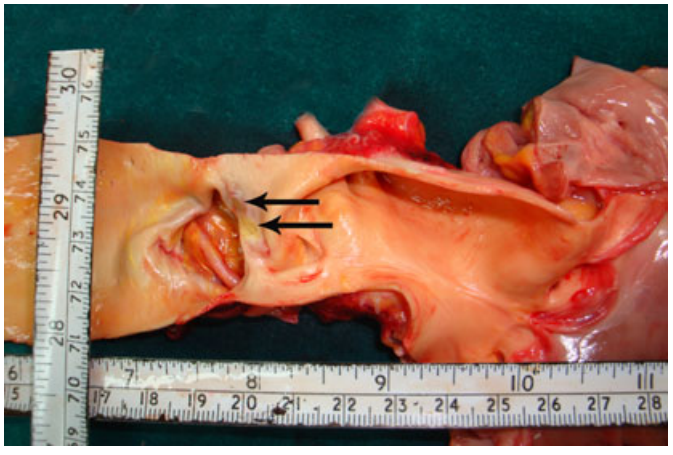

FIG. 1—*Cut open aorta showing the yellowish-white calcified lesions (arrow) over its margin and sack of the aneurysm.*

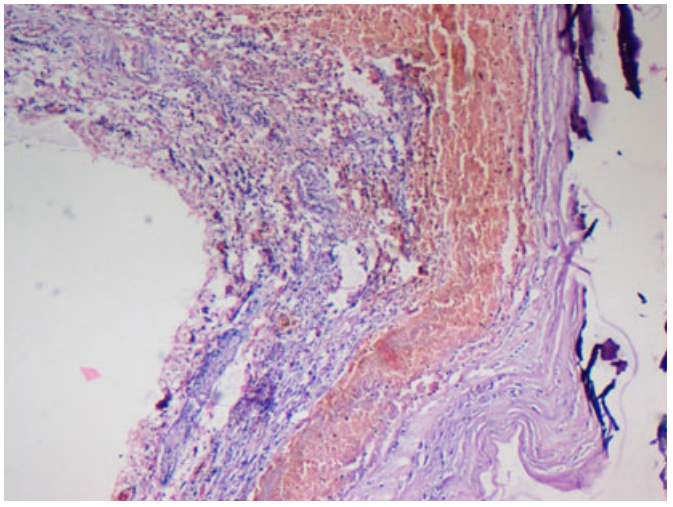

FIG. 2—*Wall of the aorta showing fibro-intimal hyperplasia with calcification; extensive hemorrhage in the media is suggestive of dissection and dense infiltrates of chronic inflammatory cells in the adventitia (Hematoxylin and Eosin 200×).*

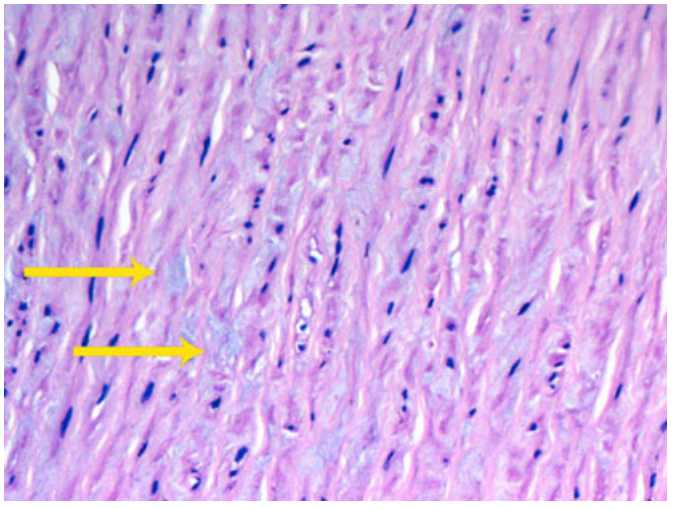

FIG. 3—*Myxoid degeneration of the aortic media characterized by separation of smooth muscle fibers by amorphous bluish myxoid material (arrow) (Hematoxylin and Eosin 400×).*

Thoracic aortic aneurysms are much less common than aneurysms of the abdominal aorta (13). The overall incidence of thoracic aortic aneurysm is estimated to be around six per 100,000 patient-years, with men being affected two to four times more often than women. The location of the aneurysms is most often the ascending aorta (60%), followed by aneurysms of the descending aorta (30%), whereas arch aneurysms and thoraco-abdominal aneurysms occur less frequently. This anatomical distinction is important because the etiology, natural history, and treatment of thoracic aneurysm differ for each of these segments. There are several heritable disorders that affect the thoracic aorta, predisposing patients to both aneurysm formation and aortic dissection, including Marfan syndrome, bicuspid aortic valve, Ehlers–Danlos syndrome, and familial forms of aortic dissection, aneurysm, or annuloaortic ectasia (1,13,14). The leading cause of mortality from this type of aneurysm is aortic rupture, accounting for 60% of deaths (15). Thoracic aortic aneurysms often present with no symptoms and are thus typically detected on routine physical examination or on evaluations for another problem. However, severe chest or back pain is frequently reported when rupture of the aneurysm occurs (16).

Dissecting aneurysms (17) differ from other aortic aneurysms in that the lumen of the vessel is usually not enlarged by the dissection but remains normal in size or is made smaller by the hematoma. The basic cause of dissection of the aorta is a defective media that can be congenital or acquired. Men are affected more frequently than women and are usually over the age of 50. After 60, the number of affected women is proportionately greater; 20–25% of reported cases are below the age of 40, and a considerable number of these are pregnant women (17). Cystic medial degeneration is the chief predisposing factor in aortic dissection. Therefore, any disease process or other condition that undermines the integrity of the elastic or muscular components of the media predisposes the aorta to dissection. Cystic medial degeneration is an intrinsic feature of several hereditary defects of connective tissue, most notably Marfan and Ehlers–Danlos syndromes, and is also common among patients with bicuspid aortic valve. The degenerative changes involve the collagenous and elastic tissues, muscle, and ground substance in varying degrees (13,17).

Schnitker and Bayer (18) found that half of the 49 cases of fatal dissection in women under 40 were in pregnant women, typically in the third trimester and also occasionally in the early postpartum period. During pregnancy, important maternal cardiovascular changes occur, such as an increase in blood volume, heart rate, stroke volume, cardiac output, left ventricular wall mass, and end-diastolic dimensions, which starts as early as the fifth week (19). In addition, hormonal changes occur that lead to histologic changes in the aorta. Fragmentation of the reticulum fibers, a diminished amount of acid mucopolysaccharides, and loss of the normal configuration of elastic fibers have been observed in the aortic wall of pregnant patients (20). Thus, both hemodynamic and hormonal mechanisms have been suggested to play an important role in the increased susceptibility to dissection in women during pregnancy and puerperium (20–25).

Aortitis is defined as an inflammation of the aortic wall with or without disruption of elastic fibers, aortic wall necrosis, or fibrosis (26). The classification of aortitis broadly includes underlying rheumatologic (noninfectious diseases) and infectious diseases along with isolated aortitis. The noninfectious diseases include Takayasu disease, Giant cell arteritis, systemic lupus erythematosus, rheumatoid arthritis, the HLA-B27–associated Spondyloarthropathies, antineutrophil cytoplasmic antibody–associated vasculitis, Behçet disease, Cogan syndrome, and sarcoidosis. Although most cases of aortitis are noninfectious in nature, the possibility of an infectious cause must always be considered because treatment strategies for infectious and noninfectious aortitis diverge widely. A number of organisms have been associated with infectious aortitis, most commonly the Salmonella and Staphylococcal species, *Streptococcus pneumonia*, *Mycobacterium tuberculosis* and *Treponema pallidum* (27). Aortitis can even be caused by viral infections like influenza A (H1N1, H3N2), B1, coxsackie A10, A16, B2, or herpes simplex (28).

In the present case, the decedent is a young non-Marfan, who underwent a lower segmental Caesarean section without any operative or postoperative complications. She was found dead on her hospital bed on the seventh day of puerperium. Her serological studies for exogenous pathogens and antibody assay did not yield any positive findings. Subsequent complete forensic autopsy along with ancillary investigations revealed that the victim died because of hemothorax from ruptured dissecting descending thoracic aortic aneurysm secondary to chronic aortitis. The thoracic aortic aneurysm could be a preexistent process as evidenced by the calcification. The compounding hormonal and hemodynamic physiological changes related to pregnancy and puerperium in the already weakened descending thoracic aorta resulted in early aneurysm rupture and dissection in her puerperium. Medial destruction of aortic aneurysms is sometimes caused by an inflammatory response within the media and adventitia, namely aortitis, which could have been a possibility in the present case as evidenced by microscopic features of chronic aortitis. In this case, the exact etiology of aortitis could not be ascertained owing to the nonspecific histologic findings and negative serological results for exogenous pathogens.

Aortic dissection during pregnancy and puerperium is reported rarely in literature. Besides, this autopsy case is reported for peculiarities, such as preexistent but asymptomatic aortitis, an early rupture of saccular descending thoracic aneurysm, and dissection during postpartum period. Pregnancy-induced stresses in diseased arterial wall could hasten the underlying pathology leading to catastrophically serious but rare event of early rupture. Rapid recognition of the acute presentation and appropriate management will improve the chances of survival in such victims. Identifying risk factors during ante- and postnatal care may assist physicians in preventing this disaster during late pregnancy and early postpartum period.

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