

## CASE REPORT

### Clostridial aortitis causing ruptured dissecting aneurysm in a young adult female

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#### Abstract

Ruptured dissecting aortic aneurysm more commonly occur in men in the 40 to 70 age group, and most commonly is associated with atherosclerosis. Uncommon causes are previous heart surgery, connective tissue disorders and aortitis. Despite its rarity, *Clostridium spp* aortitis progresses very rapidly with a mortality rate of approximately 79% in adults, typically occurring within 48 hours of infection. We present a case of sudden death due to clostridial aortitis causing ruptured aortic dissection in an apparently healthy adult female, 7 weeks post-spontaneous vaginal delivery. This case highlights the pathology of aortic dissection and cystic media necrosis as presentations of *clostridium spp* infection in young female adult.

**Keywords:** aortic dissection, cystic media necrosis, *clostridium* infection

#### INTRODUCTION

Ruptured dissecting aortic aneurysm more commonly occur in men in the 40 to 70 age group and most commonly is associated with atherosclerosis. Other uncommon causes are previous heart surgery, connective tissue disorders and aortitis.<sup>1</sup> Infection of the aorta has an overall incidence of between 0.7% and 2.6% with *salmonella spp.* representing the most commonly identified pathogens. A rare cause of aortic aneurysm is *clostridium spp.*, which typically occurs in patients with multiple co morbidities.<sup>2</sup> *Clostridium spp* infection progresses very rapidly with a mortality rate of approximately 79% in adults, typically occurring within 48 hours of infection.<sup>3</sup> In younger individuals, aortic dissection is most commonly associated with inherited connective tissue defects such as Marfan's syndrome and Ehlers –Danlos syndrome. Here we report the unusual case of a 16-year-old lady who delivered a full term baby boy, presenting with aortic dissection due to *clostridium spp* aortitis. She died at 7 weeks post-delivery due to cardiac tamponade.

#### CASE REPORT

A 16-year-old lady was found dead at her house on 21<sup>st</sup> May 2014. She was married and had a

7-week-old baby at the time of her death. The baby was delivered via spontaneous vaginal delivery at a government hospital on 2<sup>nd</sup> April 2014. Her antenatal and intrapartum period was uneventful. She had no co-morbidities or significant family history. All her first degree family members were still alive.

#### Autopsy findings

A post-mortem examination was carried out as ordered by the police. She looked pale and her body mass index was 21. There was no significant mark of injury and no features suggestive of Marfan's syndrome. CT scan showed gas shadow in the pericardium but none in the lungs which suggested the gas shadow was not due to decomposition (Fig. 1).

The aortic valve had 3 cusps and was not dilated. There was aortic dissection spanning the entire ascending thoracic aorta. A 4 cm transverse intimal tear was present just above the aortic sinus in the dilated ascending aorta (Fig. 2), causing dissection (Fig. 3), rupture into the pericardial sac and a 400 ml hemopericardium including blood clots (Fig. 4). There was no aortic atherosclerosis. No malignancy was detected. The uterus was grayish in colour and firm in consistency.

Microscopical sections showed acute aortitis

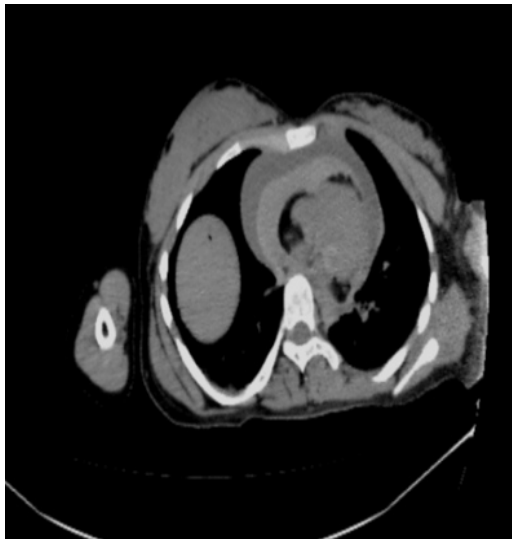


FIG. 1: CT scan demonstrating the presence of gas shadows in the pericardium

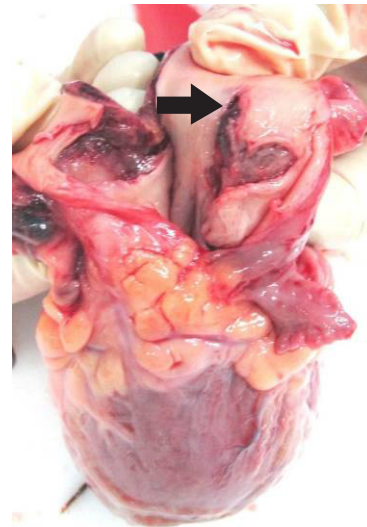


FIG. 2 : Transverse intimal tear of the aorta (arrow)



FIG. 3: Aortic dissection; note the plane of dissection

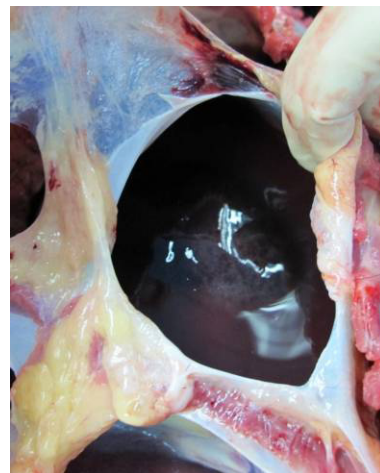


FIG. 4: Fatal cardiac tamponade. The pericardial sac is filled with 400ml of liquid and clotted blood

that involved the media and adventitia (Fig. 5). There was also basophilic degeneration of the aortic wall consistent with cystic media necrosis (Fig. 6). Histology of the uterus revealed marked inflammation of the myometrium (Fig. 7) with presence of gram positive rods.

Postmortem anaerobic blood culture grew *clostridium spp* which was consistent with gram stain findings from histopathological examination.

## DISCUSSION

Aortic dissection occurs when blood enters the aortic wall through an intimal tear and dissects

along the laminar planes of the media usually between the middle and outer thirds and can extend proximally as well as distally spanning the entire length of the aorta. The blood may rupture back into the main aortic lumen to form a 'double-barreled aorta' or as is often the case, rupture causing massive hemorrhage or hemopericardium. In fact it is only second to ruptured myocardial infarction as a cause for cardiac tamponade in cases of sudden death.<sup>4</sup> Although hypertension is the most common predisposing factor for aortic dissection, vasculitis is also not an uncommon factor. Vasculitis due to infection is rare but often fatal.

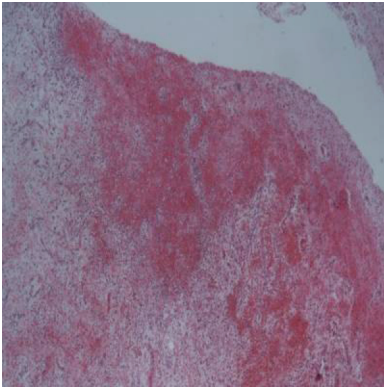


FIG. 5: Histology of the ascending aorta showing marked inflammation ( H & E X100 )

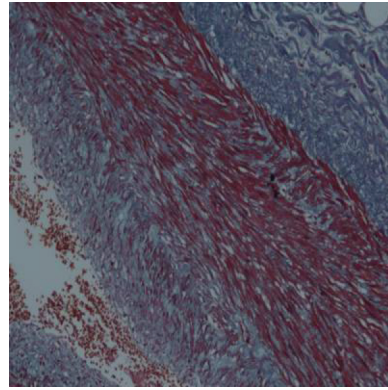


FIG. 6: Section from ascending aorta showing smooth muscle fragmentation with fine cystic lesions in the media (Masson Trichrome X 100 )

Infection of the aorta has an overall incidence between 0.7% and 2.6% with *salmonella spp.* representing the most commonly identified pathogen. A rare cause of aortic aneurysm is *clostridium spp.*, which typically occurs in patients with multiple co-morbidities. *Clostridium spp.* infection progresses very rapidly with a mortality rate of approximately 79% in adults, typically occurring within 48 hours of infection.<sup>2</sup>

The genus *Clostridium* consists of relatively large, gram-positive, rod-shaped bacteria in the Phylum *Firmicutes* (*Clostridia* is actually a Class in the Phylum). All species form endospores and have a strictly fermentative type of metabolism. *Clostridia* are ubiquitous and are found in the soil, marine sediment, decaying vegetation and intestinal tract of humans, other vertebrate and insects. Human infections with *clostridia* can result from endogenous or exogenous infection. Most *clostridia* will not grow under aerobic conditions and vegetative cells are killed by exposure to O<sub>2</sub>, but their spores are able to

survive long periods of exposure to air. *Clostridia* produce a wide variety of extracellular enzymes to degrade large biological molecules (e.g. proteins, lipids, collagen, cellulose, etc.) in the environment into fermentable components. In anaerobic clostridial infections, these enzymes play a role in invasion and pathology.<sup>5</sup> The pathogenesis of a mycotic aneurysm is by one of four mechanisms: (1) direct extension of a local infectious process, (2) trauma with contamination, (3) septic emboli to bifurcation points of smaller vessels or vasa vasorum of larger vessels and (4) haematogenous seeding from a remote source of bacteremia. This last type often involves an abnormal vessel and usually involves the thoracic or abdominal aorta.<sup>6</sup> In our case, the source of bacteremia was likely from the infected myometrium, seeding on an abnormal aorta as evidenced by cystic media necrosis (CMN) on histopathological examination.

CMN is a disorder of large arteries, in particular the aorta, characterized by an accumulation of basophilic ground substance in the media with cyst-like lesions. Gsell and Erdheim proposed the concept of idiopathic CMN attracting considerable attention, which was recognized as related to aortic aneurysm, dissection, rupture and Marfan's syndrome.<sup>7</sup> In the media, degenerative disruptions of collagen, elastin and smooth muscles may result in weakening of the arterial wall.<sup>8</sup> CMN is not the cause but is a common pathological finding, probably as a result of a primary disorder, like fibrillin deficiency in some, or advanced apoptosis in others. CMN is a common finding in elastic arterial specimens and the difference between normal and abnormal is the amount of ground substance in the media layer. Also

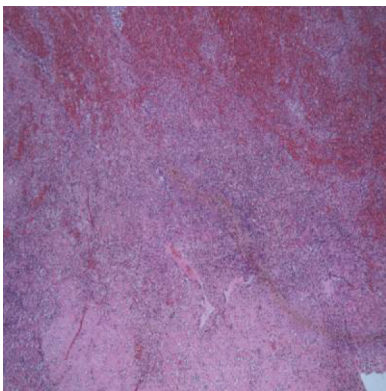


FIG. 7: Section from uterus showing marked inflammation (H&E X100)

the presence of abnormal amounts of elastic fragmentation and muscle cell apoptosis or disruption is common in these “abnormal” specimens, as described by Carlson *et al.*<sup>9</sup> The pathogenesis of these aortopathies has now been considered more a consequence of the aortic wall weakness. In particular, in Marfan’s syndrome, fibrillin-1 deficiency can be more related to aortic wall medial fragility, which is histologically evidenced by CMN, elastic fragmentation and smooth muscle cell apoptosis or necrosis.<sup>10</sup> The weakness of the aorta favours bacterial invasion and subsequent inflammation. The natural history of infectious aortitis is that of rapid aneurysmal transformation occurring in as little as 1 to 3 weeks with subsequent rupture and death.

In our case, the deceased was of aboriginal origin and most probably had traditional treatment during the postnatal period which favours clostridium infection. It has been reported that plants including roots have been used orally and topically by aborigines during the postpartum period.<sup>11,12</sup> The weakness of the aorta as evidenced by media necrosis favours *Clostridial* infection. Gross, histopathological examination, CT findings of gas shadows in the pericardium and subsequent blood culture supported the diagnosis.

The overall mortality rate is 64% with a 6 month mortality of 100% in those who do not undergo surgical intervention.<sup>13</sup> This case highlights the essential features of infectious aortitis caused by *clostridium spp* and the associated need for a high index of suspicion for rapid diagnosis and management. Also this case is peculiar because *clostridial* infection of the uterus usually cause sepsis and shock but in our case it caused aortitis, complicated with ruptured dissecting aneurysm, resulting in cardiac tamponade and death.

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