

Sudden death due to aortic dissection in pregnancy: case report

Francesco Ventura*, Maria Celeste Landolfi, Federica Portunato, Francesco De Stefano

Abstract: Pregnancy is a well-known risk factor for aortic dissection and half of the cases reported in women under 40 years of age have been estimated to occur during pregnancy, usually during the third trimester. The authors present a case of unexpected maternal death due to acute aortic dissection in a woman at the 41st week of gestation with silent clinical history. In the case reported the patient had no previous medical history when she delivered her first full-term baby, neither connective congenital abnormalities nor other morbid states that could potentially cause cardiovascular accidents. The dissecting aneurysm was due to pregnancy.

In this case, early diagnosis would have allowed early intervention which would have prevented the death of the patient, although she did not present a striking clinical situation. Under these circumstances, it is clearly of the utmost importance not to underestimate seemingly trivial symptoms that may conceal unexpected fatal clinical situations.

Keywords: Forensic pathology, sudden death, aortic dissection, pregnancy-related death, medical liability

Sudden death in young people always requires systematic forensic investigation [1]. Thus, the unexpected death of a young woman at the 41st week of gestation, without any possible explanation, always deserves close attention.

Pregnancy-related maternal death is the death of a woman resulting from, or related to, her own pregnancy and/or postpartum condition. Although pregnancy is considered a normal biologic process, it is associated with various physiologic and anatomic changes resulting in an increased risk of death [2].

Recent estimates of maternal mortality ratios vary from more than 1000 per 100,000 live births in some African countries to less than 10 per 100,000 live births in some European countries [3].

The most common types of maternal death include vascular accidents, ranging from 10 to 20%: along with pulmonary embolism, cervical and coronary thrombosis, cerebral haemorrhage, also rare events such as rupture of splenic artery aneurysm. [4- 5]

In forensic literature acute aortic dissection is rarely reported as cause of unexpected and sudden death in pregnancy without genetic connective tissue abnormalities [2].

A case of unexpected death of a young woman in the forty-first week of gestation is presented here.

Case report

A 35-year-old woman at 41 weeks of pregnancy died unexpectedly at home, while sleeping. Clinical history reports that a few days before death the woman complained of chest and back pain and underwent obstetrical consultation five days and two days before death.

* Corresponding author: Francesco Ventura, Department of Legal Medicine, University of Genova, tel + 39-010-3537838, fax +39-010-3537643, e-mail: francesco.ventura@unige.it, Department of Legal Medicine, University of Genova, via De' Toni 12, 16132 Genova, Italy

The gynaecologist considered both the course of pregnancy and the pain presentation physiological. The clinical and instrumental observations were unremarkable.

As medical malpractice was suspected, a thorough forensic autopsy was performed. Mother: abdomen was normally sized as expected in pregnancy at the 41st week (Figure 1). The pericardial sac contained 100 ml of liquid blood and 225 grams of clotted blood (Figure 2).

The haemopericardium was due to a full-thickness micro-rupture of the internal layer of the aorta (Figure 3). The aortic arch wall showed a dissecting aneurysm of rear wall (sized approximately cm 6 x 5), with other small areas of non-full-thickness dissection. Samples were collected for histological, immunohistochemical and toxicological examination. The histologic study was performed using formalin-fixed paraffin embedded tissue sectioned at 4 µm and stained with haematoxylin-eosin, Masson's Trichromic, Gomori's Trichromic and Weigert's elastic stain (Figure 4). The microscopy confirmed that aortic arch dissection was acute because of the presence of red blood cells and leukocytes in the pericardial blood clot.

- Fetus: female, 3060 gr, 50 cm long, morphologically compatible with the gestational age.

Pleural and sub-epicardial petechiae were observed. No pathological and morphological alterations were found. The placenta had a morphology compatible with the third trimester of pregnancy. The histologic examinations showed no pathological signs.

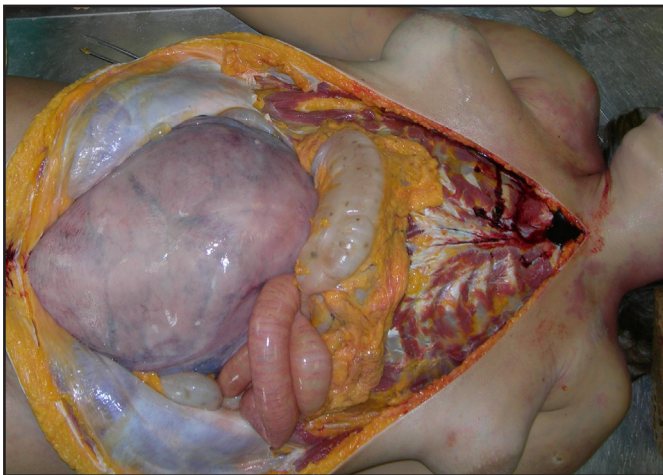


Figure 1. Maternal abdomen. The maternal abdomen normally sized as expected in pregnancy at the 41st week.

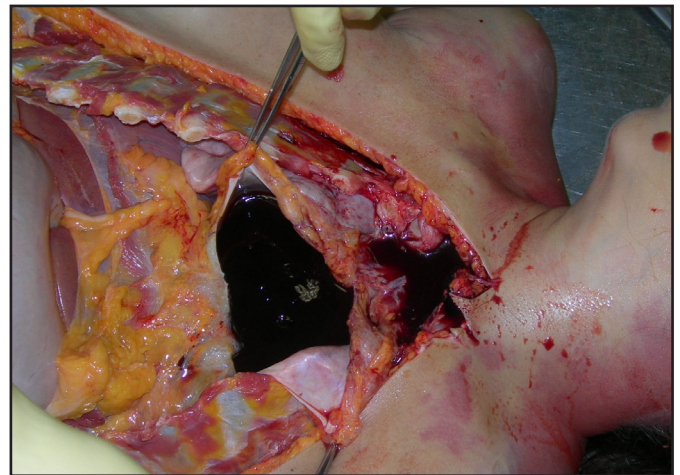


Figure 2. Haemopericardium. The pericardial sac contained 100 ml of liquid blood and 225 grams of clotted blood.

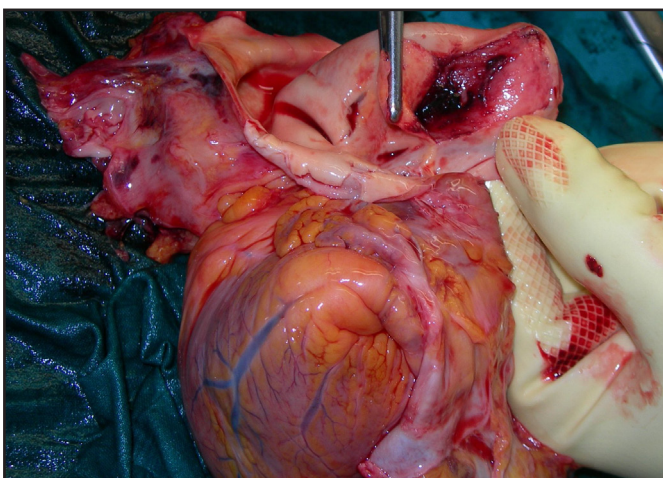


Figure 3. Dissecting aneurysm. Micro-rupture of the internal layer of the aorta.

The intrauterine fetal death was attributed to acute hypoxia secondary to maternal cardiac arrest.

Discussion

Aortic dissection is a tear in the wall of the vessel causing the flow of blood amongst the layers of the wall of the aorta. Dissection often begins from a micro-rupture of the internal layer of the aortic wall. Through this rupture the column of blood flows at high pressure and forms a false lumen in aorta that usually extends distally and, less commonly, proximally.

Dissection continues in the medial layer and can break into the adventitia or into the intimal layer. Death is often due to aortic rupture, usually within the pericardial cavity or left pleural cavity [6].

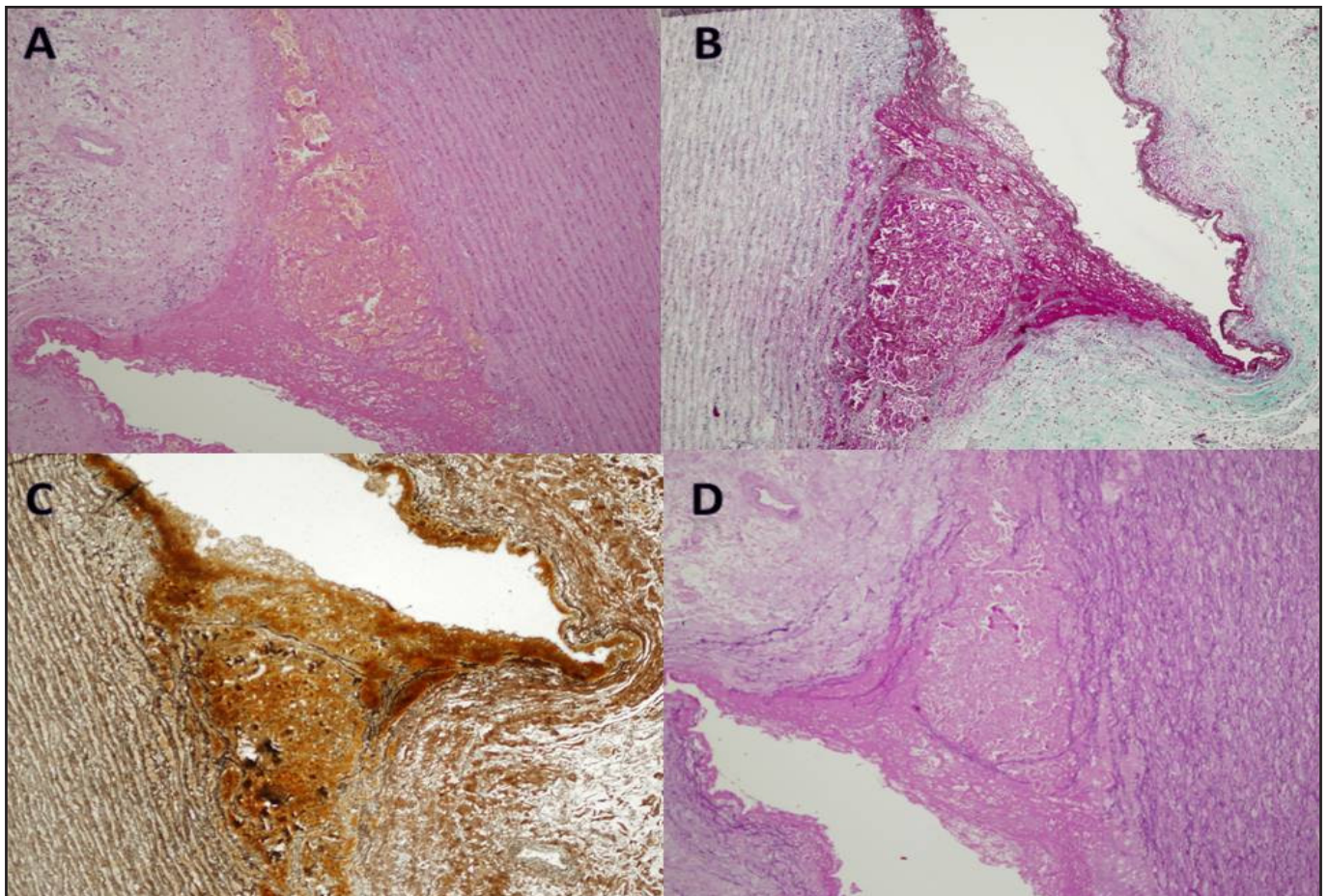


Figure 4. Histological pictures. A. Dissecting aneurysm with haemorrhagic infiltration of internal layer of the aorta. (H&E 10x); B. No evidence of fibrotic tissue (Masson's Trichromic 10x); C. Absence of cystic medionecrosis (*Gomori's* Trichrome Staining 10x); D. No evidence of elastin fragmentation (Weigert's elastic stain 10x).

The De Bakey classification system [7] divides aortic dissections into three types: the first involves the ascending and descending dissections (70% of cases), the second involves only the ascending tract (19%) and the third only the descending tract (11%).

The clinical onset of this disease is often associated with intense chest pain that migrates back along the aorta with the distal progression of the lesion. The location of the pain often indicates which aortic segments are involved: chest pain suggests a proximal aortic involvement, while back and abdominal pain usually indicates involvement of the distal segment. When dissection involves the proximal aorta, an involvement of the aortic valve or coronary artery at the level of ostia is also possible.

The damaged wall can break; if the ascending segment is involved, there may be an accumulation of serohematic fluid in the pericardium, causing cardiac tamponade and rapid cardiovascular arrest [8].

In this case, the amount of blood in the pericardium was certainly sufficient to determine the cardiovascular arrest and death of the woman. The finding of 225 grams of clotted blood and of about 100 ml of liquid blood in the pericardium suggests that the blood loss had occurred at least some hours before. The accumulation of blood in the pericardium is however considered of acute onset (a short time before death), as the histopathological examination showed clotted red blood cells and some leukocytes. The pericardial tamponade that caused the cardiac arrest was due to the small full-thickness dissection of aortic arch.

Aortic dissection in pregnancy is a rare event [9-14], a complication due to the physiological changes of pregnancy. The first cases were reported in 1944 by Schnitker and Bayer [15] and in 1957 by Pedowitz and Perelli [16].

In women under 40 years of age, about 50% of aortic dissections appear during pregnancy or puerperium, being pregnancy considered as an independent risk factor [17].

During pregnancy the high incidence of cardiovascular diseases does occur in the last months. At the onset of these problems, pregnancy-induced haemodynamic changes (increase in cardiac output and heart rate, with an increase in left ventricular mass) play an important role. Particular studies about aortic medial layer show morphological and biochemical changes, such as fragmentation of reticulin fibers, decreased acid mucopolysaccharide and collagen synthesis [18-21].

Other studies [22-24], showed that this pathology occurs more frequently in pregnant women with Marfan Syndrome, an inherited autosomal dominant syndrome with complete penetrance involving connectival alterations. The prevalence of this syndrome is 1 in 5000 Europeans; about 25% of presentations are sporadic cases. Marfan syndrome involves the cardiovascular system, with high incidence of aneurysms, aortic dissection and mitral valve prolapse. Eyes, bones and nervous system, lungs, skin and ligaments are also involved [25- 26].

Other diseases frequently associated with dissecting aortic aneurysm may be Ehlers-Danlos syndrome type IV [27] and Loeys-Dietz syndrome [28- 29].

Some studies also show a correlation between the onset of an aortic dissection and hypertension or other systemic diseases, like cardiovascular malformations, or bicuspid aortic valve [30- 31].

In the absence of other concomitant cardiovascular diseases, the onset of a soft back pain at the end of pregnancy may lead the physician to justify such pain as a result of an altered posture, a common situation in pregnancy.

The lack of positive clinical data for aortic dissection in this case complicated the definition of medical liability profiles related to obstetrical consultations in the days before death.

It would have been difficult to determine the time needed to carry out a differential diagnosis of chest pain and then to reach the correct diagnosis of aortic dissecting aneurysm, which would have required the implementation of non-routine instrumental examinations, as a chest pain associated with dyspnea can be a symptom of various pathological conditions, including dissecting thoracic aortic aneurysm.

In the first instance, therefore, the routine tests performed in cases of chest pain associated with dyspnea would be an ECG, blood tests (in particular, cardiac enzymes) or possibly a chest radiography.

The patient then should undergo more specific and invasive instrumental examinations, with some limitations arising from the advanced state of pregnancy.

As suggested by literature, transesophageal thoracic echocardiography could help gynecologists in diagnosing aortic dissection.

It was clear that suspecting that the chest pain was attributable to a cardio-vascular disease, the correct diagnosis would not have been immediate, but the patient might have been kept under observation in hospital where, in a timely manner, she could have undergone any investigations (including non-routine ones) that might enable the detection of aortic aneurysm and possibly (if dissection of the aneurysm was already there) started surgery without delay.

In the hospital where the woman was hospitalized, she would certainly have spent some hours and possible emergency surgery could have been performed within 12-24 hours after the time of the gynecological examination.

In the event that the patient had underestimated her symptoms while reporting them to the doctors, then, the treating physicians would not have had any reason to conduct an investigation for the differential diagnosis of chest pain. Particularly, it can be stressed that the onset of back pain or neck pain is a frequent situation at the end of pregnancy, while a dissecting aneurysm is a very rare complication and this, in retrospect, was the real cause of chest pain.

From a therapeutic point of view, in almost all cases of acute dissection involving the proximal aorta a surgical treatment is recommended. In the case of disease occurring during pregnancy, the literature suggests a surgery following a caesarean section after 30 weeks of gestation [32].

If the rare complication that led to the death of the patient had been suspected earlier, the treatment indicated, therefore, would have been surgical, consisting in the extraction of the mature fetus (41 weeks

of gestation), and subsequent repair of the aortic laceration in the patient. Under these conditions, literature data indicate varying rates of maternal and fetal mortality in the 20-30% range for the fetus and 2-6% for women. The statistics relative to maternal mortality rate in the presence of untreated aortic dissection argue that it increases by 1% to 3% per hour after the onset of the disease, and is approximately 25% within the first 24 hours, 70% at one week and 80% two weeks after dissection [33].

This case has provided interesting elements for a discussion regarding the hypothesis of medical malpractice, that was particularly difficult to investigate because of the different statements about the symptomatology manifested by the patient during the routine gynecological checkups. The medico-legal evaluation of medical liability according to the Italian Penal Code had to take into account the death rates shown above while considering the demonstration of a causal relationship between medical wrongdoing and death.

It was possible to state that, with a right diagnosis of aortic aneurysm made earlier, the woman might have undergone a caesarean section together with aneurysm surgery.

Regardless of when the surgical intervention would have taken place, the caesarean delivery would have probably given birth to a healthy newborn. A surgical intervention on the aorta, on the other hand, would have surely given the patient some more chances of survival, that however would have been lower as the time of a full-thickness aneurysm rupture approached.

Thus, on the basis of the aforementioned statistical considerations and taking into account that the aortic dissection started five days before death (i.e., the time of the first manifestation of the symptoms – when the back pain started), the patient would have been likely to survive, while a surgical intervention just one or two days before death, as the aortic dissection went further, would have given her less chances of survival with a high risk of death during the intervention and in the postoperative time span.

Conclusions

In the case reported the patient had no previous medical history when she delivered her first full-term baby, neither connective congenital abnormalities nor other morbid states that could potentially cause cardiovascular accidents. The dissecting aneurysm was due to pregnancy.

In this case, early diagnosis would have allowed early intervention which would have prevented the death of the patient, although she did not present a striking clinical situation.

Under these circumstances, it is clearly of the utmost importance not to underestimate seemingly trivial symptoms that may conceal unexpected fatal clinical situations.

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