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Aortic Dissection with Atypical Manifestations During Puerperium: A Case Report

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Abstract

Aortic dissection (AD) is a life-threatening condition that can be challenging to detect due to its subtle symptoms. In pregnant women and those in the postpartum period, AD ranks as the third highest cause of maternal mortality from heart-related issues globally.

Aims: To present a case of aortic dissection in a postpartum patient without conventional risk factors.

Methodology: A 35-year-old woman who had given birth via caesarean section just 7 days prior arrived at Hospital San Felix in La Dorada, Caldas, Colombia with sudden loss of consciousness and seizures. These symptoms led to unstable blood pressure and heart function, and during surgery, doctors discovered a tear in her aorta which ultimately proved fatal.

Results: The initial management was appropriate based on clinical manifestations, but a potentially fatal condition like aortic dissection was underdiagnosed due to atypical presentations.

Scientific Novelty: The presentation of aortic dissection can vary greatly, particularly in pregnant women due to physiological changes.

Conclusion: Aortic dissection is common in pregnant women and can manifest atypically, underscoring the need for increased awareness of different symptoms and signs, as well as enhanced monitoring in this population.

Keywords: Aortic dissection; Atypical manifestations; Postpartum; Physiological changes.

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Introduction

Aortic dissection (AD) is a disease with a high mortality rate and is associated with low clinical suspicion and difficulty in diagnosis. AD involves the separation of the middle layer of the thoracic wall longitudinally and circumferentially, forming two aortic channels [1, 17-24]. Within the study period of this pathology, two classifications have been established (DeBakey and Stanford), with the latter currently being favoured for its precision in evaluating the size of the lesion and symptoms based on the affected segment. The Stanford classification was divided on the basis of on whether it affects the ascending aorta (type A), the most frequent and with higher mortality, or only the descending aorta (type B) [2].

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AD is a serious condition that requires prompt and early detection and diagnosis due to its mortality rates. Its presentation is more common in older males, but it is also associated with multiple risk factors such as an aortic root diameter >40 mm, previous AD, acute myocardial infarction, family history of AD or sudden death, genetic disorders such as Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos syndrome, Turner syndrome, anatomical abnormalities such as bicuspid aortic valve and aortic coarctation, lifestyle factors such as heavy smoking, obesity, drug use such as cocaine, and during pregnancy, risk factors such as gestational diabetes and hypertensive disorders have been identified.

During pregnancy and the postpartum period, it has been identified as the third leading cause of maternal cardiovascular death worldwide [3], with a high impact on maternal-foetal well-being. This is associated with multiple physiological changes in hemodynamic, hormonal, and vascular behaviour that occur during pregnancy, extending into the postpartum state and physiological postpartum adaptation [4], that is going to be explained further. Studies have concluded that it accounts for approximately 15% of maternal deaths in the United States [5]. Pregnant women have been shown to have a 20-fold increase in the risk of developing AD [6] AD than non-pregnant women, with type B AD being the most common, with a rupture rate of 1.39/100,000 women per year in the United States [3]. This risk increases as gestation progresses and persists after delivery, with the postpartum period being the most common stage for diagnosing this pathology. Updated research from 1988 to 2012 [6] [7], revealed that only 27 cases of postpartum AD were diagnosed. Additionally, a study in the Netherlands [8] found that around half of maternal cardiac-related deaths (3 per 100,000 births) were linked to AD. Many of these cases resulted in fatal outcomes due to a lack of timely diagnosis, particularly if they occurred during the postpartum period or after the mother had been discharged from the hospital.

Research Methodology

General Background

For this research, an observational study was conducted, involving a feedback and an analysis of the various assessments as well as procedures performed by the different specialties involved in the patient management. Additionally, an investigation was conducted concerning the prevalence statistics of this pathology in the population of pregnant women, as well as the mortality statistics worldwide.

Case Report

The case involved a 35-year-old woman who recently gave birth via c-section. The surgery was necessary because the baby was positioned sideways and larger than average for the stage of pregnancy. Additionally, the woman had been diagnosed with syphilis during her pregnancy and was treated for it on the day of delivery. The patient was currently in the early stages of recovery. The woman was brought to the emergency department due to a clinical state of unknown duration, consisting of generalised clonic movements and sphincter relaxation. At the time of the initial assessment, physical examination revealed a blood pressure of 102/61 mmHg, the heart rate of 100 bpm, disorientation with a Glasgow Coma Scale score of 13/15 points, quadriparesis, and no external injuries or other abnormalities. Given these findings, the initial focus was on postpartum seizure or eclampsia, with the latter being less likely because of lower blood pressure readings. Diagnostic tests were requested to help with the case and eliminate other possible diagnoses.

Subsequently, she was assessed by an emergency medicine specialist, who managed to establish contact with the patient, who reported moderate-intensity oppressive chest pain without radiation or associated symptoms. On physical examination, the patient presented with hypotension (58/32 mmHg), tachycardia (100 bpm), tachypnoea (22 rpm), desaturation (88%), bilateral jugular engorgement, without abnormalities on cardiopulmonary auscultation, no abdominal tenderness, prolonged capillary refill time (3 seconds), oriented, and with recovery of strength in all four limbs. Among the tests performed upon admission, normal glucose levels were found, electrocardiogram showed no evidence of ischemia, brain CT scan revealed no lesions, mild leucocytosis was observed in the complete blood count, renal and hepatic function tests were normal, and partial urine analysis showed no abnormalities. Due to these findings, further studies were requested, including a chest CT scan, which revealed pericardial effusion (Images 1,2,3) and active tuberculosis in the posterior segment of the left lower lobe and all segments of the right upper lobe. Additionally, an ultrasound with the FOCUS protocol was performed, demonstrating pericardial effusion with compressive effect on cardiac cavities. After reviewing the recent findings, it was determined that the loss of consciousness was likely triggered by a syncopal episode linked to hypoxemia resulting from mechanical obstruction within the heart, in addition to presenting a clinical picture compatible with cardiac tamponade with obstructive shock due to pericardial effusion. Thus, medical management was initiated with fluid therapy and vasoactive agents leading to clinical improvement. Subsequently, the woman was assessed by general surgery, where a slight increase in blood pressure (76/56 mmHg) was noted. However, because a persistent hemodynamic instability, the patient was urgently scheduled for left thoracotomy with pericardial window placement.

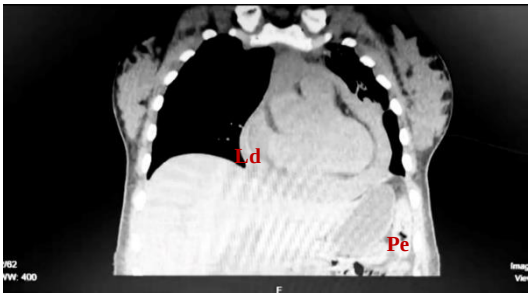

Image 1. Coronal section of a plain chest CT scan.

Left pleural effusion (Pe), Leftward deviation of pulmonary hilum and cardiophrenic angle (Ld)

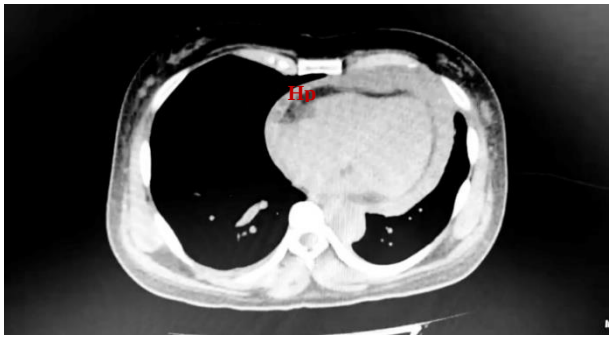

Image 2. Transverse section of a plain chest CT scan.

Hemopericardium (Hp)

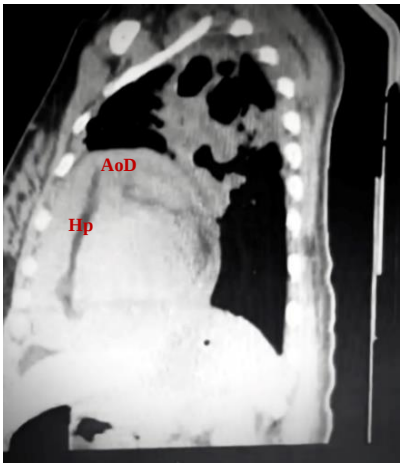

Image 3. Sagittal section of a plain chest CT scan

Hemopericardium (HP), Aortic root dilatation (AoD)

During the procedure, evidence of hemopericardium of approximately 500 cc was observed, clots in the ascending aorta with Type A aortic dissection (AD), and massive bleeding of approximately 3000 cc. As a result, a clamping of the thoracic aorta was performed, with no evidence of the bleeding site and no possibility of correction due to a bloodless heart. The patient experienced cardiac arrest, leading to the start of immediate cardiac massage without success. This ultimately led to the patient's death from haemorrhagic hypovolemic shock.

During the clinical autopsy, signs of generalised cyanosis were evident, the pericardium contained a clot, and the heart showed no structural abnormalities. The ascending aorta exhibited a thin muscular layer of less than 1 mm thickness, extending into the descending aorta. The cause of death was determined to be the rupture of an aortic dissection.

Discussion

The authors pf the paper faced a complex clinical case where, despite providing an initial approach and appropriate follow-up according to the patient's manifestations, tuberculosis findings were noted in diagnostic tests, without respiratory symptoms being present. The patient was underdiagnosed with a life-threatening condition such as aortic dissection (AD) and did not receive timely management because of its atypical manifestations, such as oppressive chest pain, hypotension associated with cardiac tamponade, syncope, and neurological manifestations. In addition, physiological changes in pregnancy involved in the pathophysiological mechanism were overlooked, often due to limited studies and literature on this pathology in pregnant patients, and a lack of diagnostic imaging, such as contrast-enhanced CT scans, which are required for accurate diagnosis. Among the differential diagnoses, a possible case of syphilitic aortitis was considered because of a positive treponemal test on the day of gestation, but this manifestation of syphilis is in the tertiary stage of infection, which corresponds to over a year of infection, inconsistent with negative screening throughout pregnancy except on the day of delivery.

As seen in our case, AD can present in various forms with atypical manifestations, depending on the affected portion of the aorta. It commonly manifests as penetrating, intense, sudden pain, and less commonly as oppressive pain, in different locations depending on the affected level, and is associated with dyspnoea and hemodynamic collapse [9]. If the ascending aorta is affected, there is usually retrosternal pain radiating to the neck, arms, or jaw, whereas if the descending aorta is affected, pain starts in the interscapular region or abdomen. In addition to pain, symptoms such as cerebral ischemia, mesenteric ischemia, and findings on physical examination, such as arterial hypertension, although it may be normal on occasion, or diastolic murmur due to aortic valve involvement, can be found. However, findings of an atypical presentation[10] can include nonspecific symptoms such as isolated abdominal pain, migratory pain, syncope, altered consciousness, congestive heart failure, arterial hypotension, cardiogenic shock, focal neurological deficit, transient ischemic attack, or fever of unknown origin. Specifically, during pregnancy or the postpartum period, they can be underdiagnosed due to some normal manifestations in pregnant women.

During pregnancy, there is an increased incidence of AD due to physiological changes and hemodynamic changes [11] such as increased systolic volume and heart rate leading to increased cardiac output, causing decreased systemic vascular resistance, increased left ventricular muscle mass, and an enlarged aortic diameter, which reaches its peak dilation during the third trimester. Hormonal changes, such as increased levels of progesterone and oestrogens, produce structural changes in the aortic wall, such as reduced mucopolysaccharides and disorganization of elastic fibres with fragmentation of reticulin, in addition to the release of matrix metalloproteinase, leading to degradation of the extravascular structural support [4]. All these changes lead to the weakening of the aortic wall, resulting in histological changes such as fragmentation of reticular fibres, disorganization of elastic fibres, hypertrophy, and hyperplasia of smooth muscle cells, increasing the risk of aneurysmal dilatation or dissection. During delivery and postpartum, other physiological changes associated with AD have been found due to increased vascular stressors such as Valsalva manoeuvres, return of volume to the systemic circulation after delivery, uterine contraction, and the use of oxytocin. Therefore, in many cases of AD reported worldwide during pregnancy or the postpartum period, risk factors mentioned previously, other than pregnancy and its physiological changes on the patient’s hemodynamic status, are not identified.

Given the high mortality rate, it is crucial to achieve an accurate diagnosis and perform differential diagnoses, such as acute coronary syndrome, pericarditis, heart failure, neurological disorders, and aortic insufficiency. The gold standard for diagnosis is contrast-enhanced CT, which can also help identify any irregularities in the various branches of the aortic artery. A more accessible imaging, such as chest X-ray, may show widening of the mediastinum, tracheal deviation, aortic frame dilation, or pleural effusion, although it has low sensitivity. In addition to achieving early detection, knowledge about follow-up and management during pregnancy is required. In patients with identified preconceptional risk factors, MRI followed by echocardiographic follow-up is recommended. The American College of Cardiology (ACC) and the American Heart Association (AHA) recommend monthly follow-up of the diameter and growth of the ascending aorta through echocardiography, whereas the European Society of Cardiology (ESC) recommends every 4-12 weeks during pregnancy and 6 months postpartum, including foetal ultrasound follow-up due to the risk detected by the use of beta-blockers during pregnancy, as it is the pharmacological management of choice. Associated risks include foetal growth restriction, small for gestational age, prematurity, and neonatal death [12]. During delivery, no management has been shown to decrease stress on the aortic wall, nor have benefits or risks of vaginal delivery or caesarean section been identified, but recommendations have been made regarding surgical management in the postpartum period. Patients with type A AD should undergo surgical repair with a 3-month course of beta-blockers, while in type B AD without rupture, the pharmacological management is indicated exclusively, except if rupture occurs, which requires immediate surgical management [13-15].

In our Colombia, as well as in many countries around the world, there is not enough monitoring of this condition during pregnancy. An increased monitoring and appropriate screening schemes should be employed, even if patients do not have conventional risk factors, to decrease maternal mortality and improve maternal-foetal well-being, as it is a disease with high prevalence worldwide that unfortunately is not diagnosed in a timely manner.

A condition such as aortic dissection (AD) during pregnancy is understudied, with little follow-up and no existing screening despite our country having a high prevalence of cardiovascular diseases. International societies such as the American Heart Association (AHA) and the European Society of Cardiology (ESC) recommend monitoring, but it has not been given the relevance it deserves. Henceforth, national and territorial health organizations should review and adopt new screening and follow-up protocols [16], where patients diagnosed with AD are integrated to provide them with counselling and recommendations about their condition, which could be implemented in our country.

Conclusions

In Colombia, there is a lack of clear statistics regarding the frequency of aortic dissection, particularly in pregnant women. Globally, aortic dissection is estimated to have a prevalence ranging from 0.2% to 0.8%, and population-based studies suggest an annual incidence of 2.5 to 7.2 cases per 100,000 individuals, with men being affected more frequently than women at a ratio of 2:1. This lack of data poses a significant risk due to the absence of adequate monitoring for this potentially fatal condition.

Here, the maternal-foetal well-being remains a public health problem, especially in rural hospitals where necessary resources are lacking to provide adequate care and optimal management for various pregnancy-related pathologies. Henceforth, national and territorial health organisations should review and adopt new screening and follow-up protocols, covering all possible diseases, to prevent fatal outcomes for the maternal-foetal dyad. An adequate education for women at high risk, families, healthcare personnel, signs and symptoms, including atypical manifestations and differential diagnoses, should be provided to prevent fatal outcomes and the underdiagnosis of this pathology, as most cases are diagnosed post-mortem.

Additionally, ensuring that complementary studies and tests are more accessible and cost-effective for various institutions across the country is crucial for enhancing prognosis, early detection, and reducing mortality rates in our patients.

Declarations

Author Contributions

Both authors participated in the conceptualisation, creation of the document, as well as the analysis of the clinical case and search for scientific references on the described pathology.

Data Availability Statement

The information and data in this article were available upon request due to restrictions such as privacy or ethics, as it concerns information from the patient's medical history, which is a legal and private document.

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Institutional Review Board Statement

The study was approved by the Ethics Committee of the E.S.E. Hospital San Félix on December 16, 2023

Informed Consent Statement

For the realization of the study, informed consent was requested from the family of the deceased patient.

Conflicts of Interest

No conflict of interests was reported regarding the publication of this manuscript.

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