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Disseminated intravascular coagulation associated with aortic aneurysm: Case report

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Priscila Buosi Rodrigues Rigolin[1], Jussara de Almeida Bruno[2], Matheus de Souza Camargo[3], Gabriel Henrique Ferreira Alves[4], Armando Carromeu Dias Pioch[5], Ana Vitoria Nunes Assis[6], Vitor Hugo Russi Mendes[7], Luana Junqueira Santos[8], Ana Victoria Carvalho Domingues[9], Louize Franci Mendonça[10], Marcelo de Jesus Garcia Júnior[11] and João Pedro Jacometo Lima[12]

ABSTRACT

Aortic aneurysms, without distinction between thoracic and abdominal, are dilations or disturbances of the functional morphological structure of the homonymous artery, which can have critical repercussions if not addressed early. More commonly, aneurysms grow at a slow pace, going unnoticed for years by patients, who often discover the disease when it is already in a very advanced stage, or when the aneurysm ruptures or leads to the dissection of the aortic layers, which translates into an acute clinical presentation that is very painful and very severe for the patient. In cases where aortic rupture or dissection occurs, mortality reaches 20% even in those patients who get emergency care 1,2.

Keywords: Aortic aneurysms, Disseminated intravascular coagulation (DIC), Aortic dissection.

[1] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[2] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[3] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[4] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[5] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[6] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[7] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[8] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[9] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[10] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[11] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente
[12] Medical students at the Universidade do Oeste Paulista - UNOESTE campus in Presidente Prudente

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INTRODUCTION

Aortic aneurysms, without distinction between thoracic and abdominal, are dilations or disturbances of the functional morphological structure of the homonymous artery, which can have critical repercussions if not addressed early. More commonly, aneurysms grow at a slow pace, going unnoticed for years by patients, who often discover the disease when it is already in a very advanced stage, or when the aneurysm ruptures or leads to the dissection of the aortic layers, which translates into an acute clinical presentation that is very painful and very severe for the patient. In cases where aortic rupture or dissection occurs, mortality reaches 20% even in those patients who get emergency care [1,2].

These deformations of the aortic wall are a very prevalent disease; in absolute terms, aneurysms are not more common than atheromatous formations, resulting from dyslipidemias, which can even lead to the formation of aneurysms. The prevalence is around 8 per 100,000 inhabitants [3]. Usually, these alterations have a multifactorial origin, being associated with a history of poorly controlled systemic arterial hypertension, iatrogenic and, less commonly, genetic dysfunctions such as Marfan and Ehler-Danlos syndromes, which alter the collagen matrix, impacting the structure of the artery wall [2].

The correction of these structural disorders is usually done by vascular reconstruction through prostheses, which can be biological or artificial. There is also the option of endovascular repairs, which are less invasive and can be a safer approach, however, each case must be evaluated individually. Ideally, patients should be diagnosed early and undergo intervention electively; The mortality rate of elective procedures for aneurysm repair is substantially lower than the mortality rate of cases that become acute with ruptures or dissections, and patients who undergo surgery fully recover their life expectancy after a few years [1,3].

The available options for aneurysm repair are known to be safe and approved by the scientific community, and long-term complications that may arise as a result of the procedure are rare. In the long term, one of the complications that can arise is the filling of a false aortic lumen, forming a new aneurysm in the region where the first one was corrected. There may be a rupture of the intimal layer, promoting blood influx through the media layer, forming a damming of blood outside the true lumen of the vessel, or even a leak through the anastomosis between the prosthesis and the patient's arterial tissue, which can also open the way for blood to accumulate between the layers of the aorta [4].

Another very uncommon complication, but which can have important repercussions, is disseminated intravascular coagulation (DIC). This hematological alteration is most often

associated with neoplastic processes, trauma, liver failure, or septicemia. In general, this clinical condition is the result of a series of events, which begins with the exacerbated activation of coagulation factors, predisposing to the formation of thrombi that can circulate through important arteries and veins, causing ischemia. Subsequently, due to consumption, coagulation factors become scarce, and a condition tends to hemorrhages. In rarer situations, DIC can be secondary to chronic diseases, such as those of the cardiovascular system, which develop slowly and gradually, such as aortic aneurysms5.

Normally, the cases of DIC found in the general population are those resulting from systemic infectious processes - or sepsis, and trauma, where there is a significant loss of volume, which significantly reduces the amount of plasma and alters coagulation factors in a short period of time. In these cases, thrombi form at several points, usually the most important infectious foci or regions where there is exposure to tissue factors; These thrombi travel the circulatory system and impact bifurcation regions or peripheral areas where the arteries have their internal diameter reduced. This mechanism can generate multiple organ failure due to the ischemic process that sets in [5].

When it comes to disseminated intravascular coagulation secondary to aortic aneurysm, there is a much rarer condition with different mechanisms. Lopes *et al*, 2019, suggest that what exists is a slowing and change in the direction of aortic blood flow in the aneurysmal region, which in itself may be enough to activate the coagulation cascade, generating a chronic condition of hypercoagulability, which, if acute, becomes a DIC, which consequently ends up consuming the coagulation factors and leaning towards a hemorrhagic state that can be equally severe [5].

The objective of this scientific text is to contribute to the scientific literature through the review of a clinical case in which a patient who was under investigation for an upper gastrointestinal hemorrhage and who suddenly presented with chest pain of strong intensity, radiating to the mandible. He had already undergone surgery to correct a previous dissecting aneurysm, and when submitted to laboratory and imaging tests, a new aneurysm formation was found close to where the first one had occurred, and concomitantly, the patient's hematological condition showed that he had an important hematological dyscrasia in progress, which was later discovered to be associated with the mural thrombus resulting from the aortic aneurysm.

METHODS

This is a clinical case report, based on an investigation of medical records containing medical evolutions, drug prescriptions, and laboratory and imaging tests, which were later compared with current scientific articles available in the PubMed and Lilacs databases.

CASE REPORT

A 59-year-old patient was admitted to investigate persistent epistaxis. During her hospital stay, she suddenly experienced chest pain in moderate intensity. The patient was immediately referred to the emergency room, where she was monitored and tests were initiated to identify the source of the pain. The electrocardiogram showed no changes that would explain the condition.

After consulting the cardiac surgeons who performed the first surgery at the same hospital, it was recommended to reduce heart rate and blood pressure. The cardiac surgery team, considering the clinical and tomographic findings, classified the aneurysm as stable, and the patient remained in the coronary unit (CCU). A chest CT scan was performed, which revealed a saccular aneurysmal dilatation in the ascending aorta, measuring 7.3 x 3.4 cm in the longest axial axes, with a parietal thrombus up to 2.1 cm thick and sparse atheromatosis.

At the same time, the patient's laboratory tests showed significant hematological changes, especially in the coagulation cascade, in addition to an anemia already expected due to the hemorrhagic condition. Although the platelet count was within the normal range, the prothrombin time (PT), activated partial thromboplastin time (APTT), and INR were severely increased, with the INR reaching 8.7.

The hematology team, after detecting some hepatic alterations, concluded that the decrease in coagulation factors was not caused by the liver, but rather by their continuous consumption, possibly due to the parietal thrombus. With this, transfusion support with plasma and red blood cells was initiated. While the hematological parameters were being corrected, the surgical team discussed and planned the approach to the case, considered unprecedented by those involved due to the proximity of the compromised region to the area of rapfia of the bovine pericardium implanted 12 years earlier, which made the procedure more delicate and complex.

There were no new cardiovascular complications, but the coagulation cascade remained altered. Laboratory follow-up showed that the patient's coagulation parameters did not respond satisfactorily to transfusion support in the first days. It took three weeks for the values to approach normality, allowing a safer surgical approach, although still outside the ideal parameters.

Dyscrasia was expected to resolve after removal of the aneurysm and mural thrombus. During surgery, the aneurysm was found in the distal portion of the ascending aorta, before the brachiocephalic trunk, extending to the right over the vena cava and right pleural protrusion. When the aorta was opened, the aneurysm and thrombus were clearly visible in the suture region between the aorta and the Dacron tube from the previous surgery.

It was decided to perform a direct suture with a bovine pericardial patch, forming a new aortic tube sutured in the upper and lower portions of the aneurysm. The procedure was uneventful, and the patient was kept in the intensive care unit (ICU) for 4 days, evolving without complications. Later, she was transferred to the cardiology ward. As expected, the hematological condition improved after surgery. After 9 days in the ward, the patient, who evolved well throughout the period, was discharged and followed up on an outpatient basis, completely recovering her quality of life.

RESULTS

Through the review of the medical records and the literature review, it was possible to verify that this was a rare case, with an incidence rate of 0.5 to 4% among patients with aortic aneurysms. By having all the care well documented, it was possible to report this clinical case with a certain wealth of pertinent details, which generated very satisfactory results for the research team.

DISCUSSION AND CONCLUSION

In this clinical case report, a succession of events is evidenced that, combined, result in an unusual and somewhat curious presentation. The coagulopathy presented by the patient had rare characteristics, such as bleeding and alteration of the coagulation cascade, in the order of 0.5 to 4% among all cases of aneurysms with parietal thrombi6. More frequently, isolated elevation of fibrin degradation products, especially D-dimer, occurs in up to 40% of cases, according to the current literature [5,6].

The presence of the parietal thrombus, which was extensive, was certainly one of the decisive factors for the onset of blood dyscrasia presented by the patient. The turbulence caused by the presence of the same in a region of high blood flow in a high-pressure regime certainly led to blood turbulence, which alone may have been enough to initiate the coagulation cascade continuously, culminating in the depletion of the factors of the cascade itself [5]. This was hypothesized by the hematology team in the interconsultation that was requested.

At first, before upper digestive endoscopy (UGI) was performed, the possibility of a communicating fistula between the aneurysm and the esophagus was speculated, which would explain the patient's melenic evacuation during the period in which he was hospitalized before presenting precordial pain, as well as hematemesis. The UGIE result, however, showed that the lesion surrounded in the esophagus was the hemorrhagic focus, which helped to support the diagnostic hypothesis of DIC caused by the parietal thrombus, a result of the aneurysm.

Although this phenomenon should have been affecting the patient for a longer time than he had been hospitalized, and it is impossible to measure a period, it is compatible with the nature of aneurysms that the condition was completely asymptomatic; Guo *et al*, 2018 and Davis *et al*, 2019, agree that aortic aneurysms can go completely unnoticed by most patients until there is a dissection or other hemodynamic instability resulting from it, which usually results in chest pain, which results in seeking urgent medical care.

The nature of this aneurysm, in itself, is already somewhat unusual. Aneurysms in regions with surgical corrections of previous aneurysms are not frequent. Hernadez-Vaquero *et al*, 2020, conducted a prospective study with 738 patients who underwent aortic aneurysm repair; of these, only 6 developed new aneurysms and 5 ended in a new dissection, requiring a new surgical approach. The data reinforce that surgical correction for disorders of this nature is effective and safe, and there are rare occasions when it will be necessary to reoperate on the patient.

The association of the commemorative presented made it necessary for several professionals from different areas of activity to take joint action, which optimized the results. We can conclude that, although it was facing an infrequent situation, and that, according to the surgeons of the case, all of whom were experienced, it was unprecedented for them, the team did well and knew how to make good use of the resources that were available. This allowed an adequate approach, allowing the best surgical approach to be performed, within a time window where the hematological parameters were the most appropriate possible, given the limitations imposed by the patient's own condition.

CONFLICT OF INTEREST

The authors declare that there is no potential conflict of interest that could interfere with the impartiality of this scientific work.

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