Management of Acute Aortic Dissection Associated With Malperfusion Syndrome: About 4 Cases and Review of the Literature

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Abstract

Original Research Article

Background: Acute aortic dissection is a medico-surgical emergency that is associated with a poor prognosis, especially when complicated by malperfusion syndrome. **Purpose:** The objective of our study was to assess the prognosis and management of aortic dissection complicated by malperfusion syndrome. **Methods:** Retrospective study from December 2017 to October 2019 including 4 patients hospitalized for aortic dissection with malperfusion syndrome operated at the Ibn Sina University Hospital. The diagnosis was made on clinical, biological and CT scans. **Results:** Patients' mean age was $65.75 \pm 11,5$ years with a sex ratio M / F of 1/3. Hypertension was the main risk factor and was found in all patients, only one of our patients was a smoker. The organs affected by malperfusion, one case of renal malperfusion, two (02) cases of lower limb malperfusion, two (02) cases of digestive malperfusion. The management was surgical. Three patients underwent supracoronary tube placement and one patient underwent Bentall's procedure. Aorto-coronary bypass, an axillofemoral bypass and a cross bypass were performed to remove myocardial and lower limbs malperfusions. Mortality at one month was 50%, with one patient dying in-hospital from cardiogenic shock and one patient dying at one month from lower limb gangrene. **Conclusion:** Surgery is still relevant in managing aortic dissection associated with malperfusion. In our study, the survival at one month was 50%, suggesting the necessity to improving our management of aortic dissection when associated with malperfusion.

Keywords: Dissection; aortic; malperfusion; prognosis; management.

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INTRODUCTION

Aortic dissection (AD) is a pathology of the aorta that results in a tear in the intima with an inflow of blood into the wall. The delay of less than two weeks since onset defines the acute nature [1]. Several classifications are used in aortic dissection. According to the Stanford classification, a distinction is made between type A in which the ascending aorta is affected and type B in which only the descending aorta is involved. The main risk factor of AD is high blood pressure [2].

Acute AD can be complicated by a malperfusion syndrome, defined as a complication in an organ system secondary to ischemia and resulting in organ dysfunction, such as the brain, myocardium, kidneys, lower limbs and viscera (3) which increases the severity of this condition [4-6].

Through four (04) cases and data from the literature, we will try to show the impact of

malperfusion on the prognosis and therapeutic management of AD.

PATIENTS AND METHODS

This is a retrospective study covering the period of two (02) years: from December 2017 to October 2019 of four patients operated in the cardiovascular surgery department of "Hopital Ibn Sina" for AD with malperfusion.

We used medical observations to collect clinical, paraclinical and therapeutic data, establishing a prognosis study.

Patients were followed up for one month to assess progress. We summarized the clinical cases of our patients in a table (Table 1).

RESULTS

The mean age is 65.75 years (53-78 years), with a sex ratio of 0.33 (three women and one man) and all our four (04) patients had a history of hypertension.

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The first patient had cerebral malperfusion, the second had visceral and lower limb malperfusion, the third had coronary malperfusion, and the last had digestive, renal and lower limb malperfusion.

The clinical signs that are consistently encountered and are suggestive of malperfusion syndrome are:

- Signs of lower limb is chaemia found in two of our patients: Claudication, gangrene
- Confusional syndrome found in one patient
- Anginal pain found in one patient
- Melena + abdominal pain in one patient
- Anuria

Biology allowed us to find myocardial damage by measuring troponin, renal insufficiency and rhabdomyolysis.

Parameters Patients	Sex	Age	Risk Factors	Clinical signs	CT Angiography	Echocardiography	Intervention	1 month survival
Patient 1	Female	78 years old	High Blood Pressure	-Chest pain -Dyspnea -Confusional syndrome	AD type A extended to the abdominal aorta, TABC, left common carotid artery, Hemopericardium	Appearance of AD with false channel and intimal flap, root of aorta not dilated, Circumferential pericardial effusion of moderate size,Intact aortic valve, LV with preserved systolic function	Placement of a live supra- coronary tube	Alive
Patient 2	Female	53 years old	High Blood Pressure Chronic smoking	Chest pain Claudication of the right lower limb melena	Type A AD extending to the celiac trunk, right renal artery, inferior mesenteric artery with CT evidence of intestinal ischaemia, Pericardial effusion	AD appearance with false channel and intimal flap, undilated aortic root, Circumferential pericardial effusion of moderate size, Intact aortic valve LV with preserved systolic function	Placement of a supra- coronary tube	Alive
Patient 3	Female	60 years old	High blood Pressure	Epigastric pain NYHA stage IV dyspnoea	Type A AD arising from an aneurysmal aorta, Entry portal in the ascending aorta with exit orifice about 5.6cm from the valve. Right and left coronary arteries arising from the true channel without a clearly individualisable intimal flap (at the limit of the examination carried out, artefactual at this level) Respect for the arch and the rest of the aorta	AD beginning at the sino-tubular junction and extending to the ascending aorta, respecting the aortic isthmus and descending aorta Massive aortic insufficiency, Dilated, non-hypertrophic left ventricle, Inferior hypokinesia, ejection fraction impaired to 48%. High left ventricular filling pressures Minimal pericardial effusion	Bentall procedure with right coronary artery bypass surgery	Died

Table 1: Summary table of clinical cases in our series

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Patient 4	72 years old High Blood Pressure	Lower limb ischemia Abdominal pain	Dissection of ascending aorta extended to common iliac arteries, intestinal ischaemia, moderate pericardial effusion	Initial aorta dilated to 43mm with dissection (presence of intimal flap from initial part of aorta) Intact aortic valve Dry pericardium	Right lower limb angioplast y: Axillofem oral bypass with cross- over Placement of a supra- coronary tube	Died
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Table 2: Comparative table of organs with malperfusion between our study and those of Schoell T et al., and Yang

et al.,						
	Studies	Our study	Schoell T et al.,	Yang et al.,		
Organs	_					
affected by malperfusion						
Cerebral		25%	57.1%	5.2%		
Myocardial		25%	3.6%	2.8%		
Digestive		50%	3.6%	16.7%		
Renal		25%		12%		
Lower limb		50%	28.6%	1.3%		



Figure 1: Sagittal section showing type A aortic dissection with extension to the brachiocephalic artery trunk (1: Entry tear, 2: Intimal flap, 3: brachiocephalic artery trunk)



Figure 2: 3D reconstruction showing a type A dissection of the aorta extended to the left common iliac artery treated with a supracoronary tube

DISCUSSION

The incidence of malperfusion in acute AD is 25-30% [7]. The main risk factor is high blood pressure, found in 76.6% of cases [8]; all our 4 patients were hypertensive.

There are two types of pathophysiological mechanisms explaining ischaemia in AD: a static mechanism and a dynamic mechanism [9].

Dynamic mechanism:either secondary to insufficient blood flow through the true lumen which can lead to hypoperfusion when the branched vessel is maintained by the true lumen, or secondary to protrusion of the intimal flap into the ostium of a peripheral vessel compromising perfusion of the latter [9].

Static mechanism:secondary to extension of the dissection to a collateral [9]. Our study included four (04) patients with AD complicated by malperfusion, the malperfusion profile was distributed as follows: Two (02) cases of lower limb malperfusion, two (02) cases of digestive malperfusion, one (01) case of renal malperfusion, one (01) case of cerebral malperfusion, and one (01) case of myocardial malperfusion.

Malperfusion profile is diversified according to the literature. (see table 2 comparing the malperfusion profile of our study with other studies of the literature).

The diagnosis of AD with malperfusion syndrome in our study was made on the basis of clinicobiological signs of ischaemia or necrosis with CT confirmation.

All four of our patients underwent routine thoracic-abdominal-pelvic CT in order to detect any clinico-biological silent malperfusion.

The diagnostic approach is sometimes more thorough in the literature, using invasive methods. In the Yang *et al.*, study on the value of revascularisation prior to surgery in 597 patients, 135 of whom had malperfusion, angiographic evaluation was indicated prior to the therapeutic decision [10].

Selective arteriography is sometimes performed to confirm obstruction, locate the terminal extension of a dissected arterial branch, and distinguish between true and false lumen [10]. Historically, the treatment for type A AD has been emergency open surgery.

IRAD data show that the mortality rate in the absence of surgery with medical treatment only is 24% in 24 hours, 29% in 48 hours, 44% in 7 days, 49% in 14 days [1, 11]. Surgical intervention reduces the mortality rate to 1% in 24 hours, 16% in 7 days, 20% in 14days [1, 11].

Immediate open surgery is indicated to prevent the risk of aortic rupture [1, 11]. However, when malperfusion syndrome is associated, this approach is discussed. Some teams adopt this same attitude which would allow the removal of malperfusion.

Myocardial ischaemia due to extension of the dissection to the coronary ostia is very often corrected by replacement of the ascending aorta and exclusion of the false channel of dissection at segment 0 of the aorta.

Cerebral ischaemia can also be resolved with resolution of neurological deficits, but this depends on the extent of the ischaemia and the time to surgery [12].

On the other hand, more distal ischaemia, notably renal and visceral, persists despite surgical treatment, particularly when it is due to a static mechanism, dynamic malperfusion being most often corrected by surgical treatment with restoration of flow in the true channel.

This is the technique that was adopted for our patients and when malperfusion persisted, the treatment was complemented by revascularisation techniques.

Other teams, such as the Michigan team in the Yang *et al.*, study, propose deferred surgery to revascularise patients early, allowing improvement of organ dysfunction and selection for surgery of patients with the best prognosis. There remains the risk of aortic rupture with 4 and 16% rupture while waiting for surgery.

The immediate risk of mortality from aortic rupture would be lower than that from organ failure secondary to malperfusion.

In the Yang and al. study conducted by the Michigan team, the risk of mortality from organ failure secondary to malperfusion was about 7 times greater than that of aortic rupture even after malperfusion was removed.

In the case of lowerlimb ischaemia, the question also arises as to whether or not lower limb reperfusion should precede aortic surgery.

Delaying reperfusion carries the risk of reperfusion syndrome with renal failure and metabolic acidosis. However, data from the literature show that aortic repair should be preferred;

This attitude, in addition to preventing the risk of rupture, which is fatal, makes it possible to resolve the malperfusion of the lower limb in 60-100% of cases [13].

If malperfusion persists, revascularisation of the ischaemic limb by aorto-femoral bypass is performed. Malperfusion syndrome is an independent factor for mortality in AD.

Data from the GERAADA registry show that mortality varies with malperfusion and with the number of organs affected by malperfusion.

Mortality is 12.6% in the absence of malperfusion, 21.3% if only one organ is affected, 30.9% for two organs and 43.4% for three organs, i.e. an increase of approximately 10% per organ affected [14]. The nature of the organ affected is also a determining factor in the prognosis.

According to IRAD data, the mortality rate was 63.2% in the case of mesenteric malperfusion, especially as this is most often associated with other types of malperfusion [15].

In our study, intra-hospita lmortality concerned one (01) patient, i.e. 25%, and one-month mortality amounted to two (02), i.e. 50%.

CONCLUSION

Aortic dissection with malperfusion syndrome is associated with a very poor immediate prognosis.

Early management conditions the prognosis by minimising the risks of rupture and mortality related to malperfusion.

The development of endovascular techniques should be used to improve the prognosis. Cohort studies are desirable to propose standardised management.

REFERENCES

- 1. Levy, D., Goyal, A., Grigorova, Y., & Le, J. K. (2017). Aortic dissection.
- Hagan, P. G., Nienaber, C. A., Isselbacher, E. M., Bruckman, D., Karavite, D. J., Russman, P. L., ... & Eagle, K. A. (2000). The International Registry of Acute Aortic Dissection (IRAD): new insights into an old disease. *Jama*, 283(7), 897-903.

- 3. Yagdi, T., Atay, Y., Engin, C., Mahmudov, R., Tetik, O., Iyem, H., ... & Buket, S. (2006). Impact of organ malperfusion on mortality and morbidity in acute type A aortic dissections. *Journal of cardiac surgery*, 21(4), 363-369.
- Cambria, R. P., Brewster, D. C., Gertler, J., Moncure, A. C., Gusberg, R., Tilson, M. D., ... & Abbott, W. M. (1988). Vascular complications associated with spontaneous aortic dissection. *Journal of vascular surgery*, 7(2), 199-209.
- Lauterbach, S. R., Cambria, R. P., Brewster, D. C., Gertler, J. P., LaMuraglia, G. M., Isselbacher, E. M., ... & Moncure, A. C. (2001). Contemporary management of aortic branch compromise resulting from acute aortic dissection. *Journal of vascular surgery*, *33*(6), 1185-1192.
- Oderich, G. S., Panneton, J. M., Bower, T. C., Ricotta, J. J., Sundt, T. M., Cha, S., & Gloviczki, P. (2008). Aortic dissection with aortic side branch compromise: impact of malperfusion on patient outcome. *Perspectives in vascular surgery and endovascular therapy*, 20(2), 190-200.
- Crawford, T. C., Beaulieu, R. J., Ehlert, B. A., Ratchford, E. V., & Black III, J. H. (2016). Malperfusion syndromes in aortic dissections. *Vascular Medicine*, 21(3), 264-273.
- Evangelista, A., Isselbacher, E. M., Bossone, E., Gleason, T. G., Eusanio, M. D., Sechtem, U., ... & Eagle, K. A. (2018). Insights from the international registry of acute aortic dissection: a 20-year experience of collaborative clinical research. *Circulation*, 137(17), 1846-1860.
- Williams, D. M., Lee, D. Y., Hamilton, B. H., Marx, M. V., Narasimham, D. L., Kazanjian, S. N., ... & Deeb, G. M. (1997). The dissected aorta: part III. Anatomy and radiologic diagnosis of branchvessel compromise. *Radiology*, 203(1), 37-44.

- Yang, B., Rosati, C. M., Norton, E. L., Kim, K. M., Khaja, M. S., Dasika, N., ... & Williams, D. M. (2018). Endovascular fenestration/stenting first followed by delayed open aortic repair for acute type A aortic dissection with malperfusion syndrome. *Circulation*, 138(19), 2091-2103.
- Mehta, R. H., Suzuki, T., Hagan, P. G., Bossone, E., Gilon, D., Llovet, A., ... & Eagle, K. A. (2002). Predicting death in patients with acute type A aortic dissection. *Circulation*, 105(2), 200-206.
- 12. Morimoto, N., Okada, K., & Okita, Y. (2011). Lack of neurologic improvement after aortic repair for acute type A aortic dissection complicated by cerebral malperfusion: predictors and association with survival. *The Journal of thoracic and cardiovascular surgery*, *142*(6), 1540-1544.
- 13. Preece, R., Srivastava, V., Akowuah, E., & Kendall, S. (2017). Should limb revascularization take priority over dissection repair in type a aortic dissection presenting as isolated acute limb ischaemia. *Interactive cardiovascular and thoracic surgery*, 25(4), 643-646.
- 14. Czerny, M., Schoenhoff, F., Etz, C., Englberger, L., Khaladj, N., Zierer, A., ... & Carrel, T. P. (2015). The impact of pre-operative malperfusion on outcome in acute type A aortic dissection: results from the GERAADA registry. *Journal of the American College of Cardiology*, 65(24), 2628-2635.
- 15. Di Eusanio, M., Trimarchi, S., Patel, H. J., Hutchison, S., Suzuki, T., Peterson, M. D., ... & Fattori, R. (2013). Clinical presentation, management, and short-term outcome of patients with type A acute dissection complicated by mesenteric malperfusion: observations from the Acute International Registry of Aortic of Dissection. The Journal thoracic and cardiovascular surgery, 145(2), 385-390.