Case study

PAINLESS AORTIC DISSECTION: A RARE AND ATYPICAL CLINICAL PRESENTATION

ABSTRACT:

Aortic dissections (AD) are uncommon but they are highly lethal. It's a vascular emergency that if not diagnosed and treated in a timely manner can result in death. Due to atypical signs and symptoms, diagnosis of type A AD can be easily missed or delayed. We present the case of a 62 year-old female, without any cardiovascular risk factor, presenting to the Emergency Department with digestive symptomatology over 15 days with no chest pain. The patient was clinically stable with normal initial laboratory investigations. Consequently, she was discharged from the Emergency Department. Four days later, she consulted a cardiologist for progressive dyspnea. A thoraco-abdominal CT with contrast was performed confirming the Stanford type A thoracic aortic dissection above the aortic valve, with a thrombosed dilation extending on 61mm. A transthoracic echocardiogram showed a dilation of the thoracic aorta with a type A aortic dissection, and a thrombosed false lumen. Urgent surgery was proposed but unfortunately refused by the patient and its family.

From this case, we learn how it is challenging to diagnose painless AD especially when patient comes with atypical symptoms.

KEYWORDS:

Acute aortic dissection, Atypical and painless presentation, Dyspnea,

INTRODUCTION:

Aortic dissection (AD) is a life-threatening condition associated with significant mortality. **(1)** Symptoms of acute aortic dissection may vary. Most patients complain of an abrupt onset of severe pain in thechest, back, or abdomen **(2)**. Atypical presentations are relatively rare and make the diagnosis difficult, especially when the classic pattern of pain is absent : Painless aortic dissection (PAD). We present an unusual case of acute painless aortic dissection type A, with a progressive shortness of breath and weakness, in a patient without risk factors.

CASE REPORT:

A 61-year-old female with a past medical history of a laminar tricuspid regurgitation (waiting for surgery), without cardiovascular risk factors, presented to the Emergency Department with vomiting, nausea, anorexia, constipation and weakness over 15 days. No associated chest pain, palpitations, or syncope are noted. The patient denies any associated cough, hemoptysis or fever. The clinical examination found a clinically stable patient. The chest X-ray, electrocardiogram, and initial laboratory investigations were all normal.

Consequently, she was discharged from the emergency department. Four days later, she consulted a cardiologist for progressive dyspnea. Vitals signs were: a wide pulse pressure with 107/60 mmHg on the right arm, and 112/70 mmHg on the left one, heart rate of 60 beats/minute, respiratory rate of 18 breaths/minute and temperature of 36.9°C. No signs of heart failure were found. Cardiovascular auscultation was normal. The rest of physical examination was normal as well.

Electrocardiogram showed sinus rate at 80 beats/minute, incomplete left bundle branch block. An acute pulmonary embolism was highly suspected. A thoracoabdominal CT with contrast was performed confirming the Stanford type A thoracic aortic dissection above the aortic valve, with a thrombosed dilation extending on 61mm, excluding pulmonary embolism

(Figure : 1). She underwent transthoracic echocardiography which revealed dilation of the thoracic aorta with a type A aortic dissection, and a thrombosed false lumen, without any aortic valvulopathy. The patient was admitted to the medical intensive care unit for blood pressure control and close monitoring. Vascular surgery was consulted and recommended surgical treatment, that was integrally refused by the patient and its family, due to the high risk of mortality.

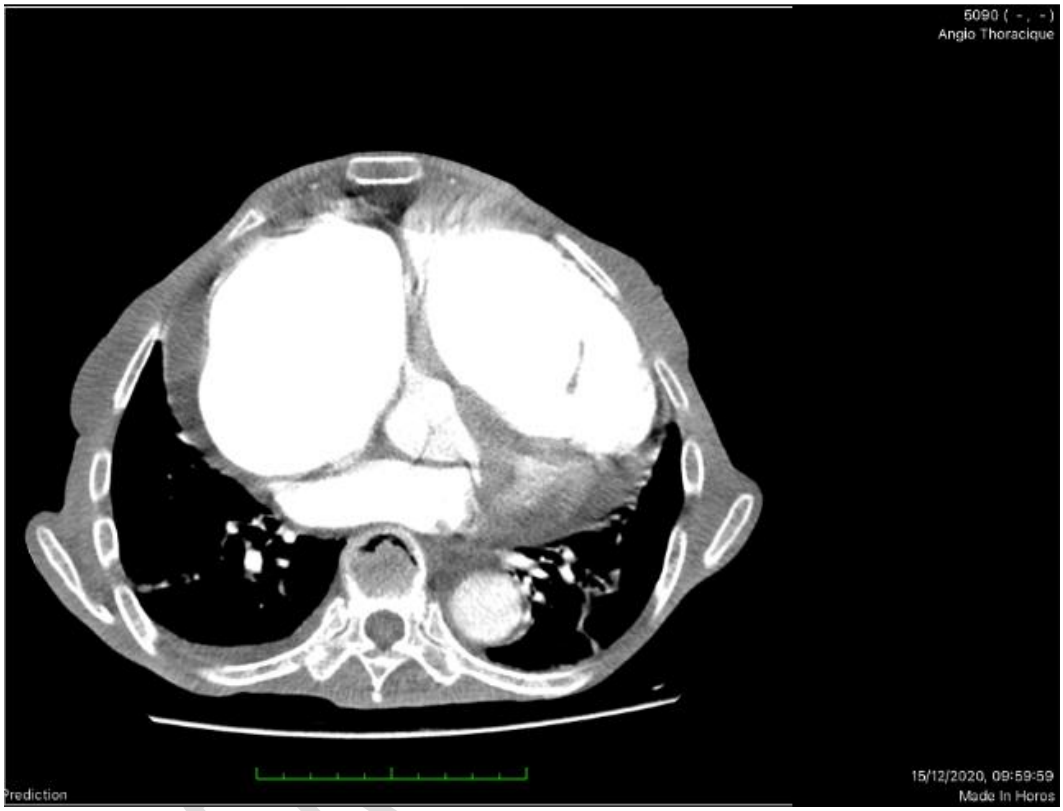

Figure 1: Thoraco-Abdominal CT with contrast confirming the Stanford type A thoracic aortic dissection above the aortic valve.

DISCUSSION:

Acute AD is one of the life-threatening cardiovascular emergencies associated with significant in-hospital mortality (27,4%) and an incidence between 2.6 to 3.5 per 100 000 person-years, while in-hospital mortality is 27.4%. **(3, 4)** The Stanford classification system defines two types of aortic dissection, type A which involving the ascending aorta and type B limited to the aorta distal to the left subclavian artery. This classification helps to

separate type A which needs surgical management, whereas type B usually requires only medical management.

Hypertension is the most significant risk factor for AD (which was absent in our case). **(5, 6)** Other risk factors include age, smoking, dyslipidemia, cocaine use, hereditary disorders (Marfan syndrome, Ehlers-Danlos syndrome, Turner syndrome, bicuspid aortic valve, coarctation of the aorta, preexisting aortic aneurysm), vasculitis, trauma, and iatrogenic factors. **(7)**

AD happens when an intimal tear leads to the leakage and propagation of blood through the aortic media creating a false lumen. The dissection may propagate proximally and can involve the heart or distally the descending aorta and its major branches. Pathogenesis is related to factors that contribute to aortic wall medial degeneration and those that increase aortic wall stress.

Chest pain is one of the most common symptoms in aortic dissection and its absence makes the diagnosis difficult for the clinician. It's classically described as sudden onset of tearing chest pain, often in the interscapular region and may be associated with syncope, dyspnea, and weakness **(7)**. However, this classical clinical presentation may be absent. **(8)** 4.5%-6% of ADs are painless, and without risk factors (as in our patient) and the chances of missing the diagnosis are very high in those cases. **(5, 9)** The initial complaints of these patients do not suggest cardiovascular system pathology (As in our presented case). For this reason, the time from admission to a definite diagnosis was significantly longer.

Diagnosis depends on clinical presentation and imaging. High index of suspicion is very important in at-risk patients.

Magnetic resonance imaging (MRI), computed tomography (CT) or transesophageal echocardiography (TEE) are diagnostic with sensitivities of almost 98.3%, 98.3%, and 97.7%, respectively Transthoracic echocardiography (TTE) has only 59.3% sensitivity. **(10)** MRI, CT and TTE imaging usually reveal an intimal flap allowing rapid and accurate diagnosis of AD. **(11)** (In our case, TTE and CT were enough for the diagnosis. MRI was not performed).

Treatment of AD depends on the type of dissection and associated complications. Type A AD usually requires emergent surgery. Whereas type B AD is usually treated medically, but it may need surgical or endovascular intervention when it is

associated with complications. Beta-blockers are important in the management of AD (used for our patient). **(12, 13)**

CONCLUSION:

Atypical presentation of aortic dissection without chest pain may be more frequent than previously reported, that physicians should always keep in mind and widen their differential diagnosis, especially with symptoms of dyspnea and do not hesitate to perform a transthoracic echocardiogram at the slightest suspicion since it is easy to perform and has a non-negligible sensitivity and specificity.

REFERENCES:

1- Khan IA, Nair CK: Clinical, diagnostic, and management perspectives of aortic dissection . Chest. 2002, 122:311-328.

2- Juang D, Braverman AC, Eagle K. Aortic Dissection. Circulation, 2008; 118: e507-e510.

3- Clouse WD, Hallett JW, Schaff HV, et al., editors. Acute aortic dissection: population–based incidence compared with degenerative aortic aneurysm rupture. Mayo Clinic Proceedings; Elsevier, 2004.

4- Meszaros I, Morocz J, Szlavi J, et al. Epidemiologyand clinicopathology of aortic dissection: A population–based longitudinal study over 27 year.Chest, 2000; 117(5): 1271–78.

5- Alter SM, Eskin B, Allegra JR: Diagnosis of aortic dissection in emergency department patients is rare . West J Emerg Med. 2015, 16:629-631.

6- Park SW, Hutchison S, Mehta RH, et al.: Association of painless acute aortic dissection with increased mortality. Mayo Clin Proc. 2004, 79:1252-1257.

7- Hagan PG, Nienaber CA, Isselbacher EM, et al. The International Registry of Acute Aortic Dissection (IRAD): New insights into an old disease. JAMA, 2000; 283(7): 897–903.

8- Hagan PG, Nienaber CA, Isselbacher EM, et al.: The International Registry of Acute Aortic Dissection (IRAD): new insights into an old disease. JAMA. 2000, 283:897-903.

9- Tsai TT, Nienaber CA, Eagle KA: Acute aortic syndromes. Circulation. 2005, 112:3802-3813.

10- Nienaber CA, von Kodolitsch Y, Nicolas V, et al.: The diagnosis of thoracic aortic dissection by noninvasive imaging procedures. N Engl J Med. 1993, 328:1-9.

11- Fisher ER, Stern EJ, Godwin JD 2nd, Otto CM, Johnson JA: Acute aortic dissection: typical and atypical imaging features. Radiographics. 1994, 14:1263-1271.

12- Nienaber CA, Clough RE: Management of acute aortic dissection . Lancet. 2015, 385:800-811.

13- Pape LA, Awais M, Woznicki EM, et al.: Presentation, diagnosis, and outcomes of acute aortic dissection: 17- year trends from the International Registry of Acute Aortic Dissection. J Am Coll Cardiol. 2015, 66:350-358.