Case Report Multiple spontaneous isolated arterial dissections: a rare case report

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Abstract: Spontaneous dissections in multiple arteries are a rare condition with clinical presentation varying from asymptomatic conditions to sudden death. We present a rare case where a routine thoracic computed tomography (CT) scan showed a type B aortic dissection. Medical records showed that the patient previously had been diagnosed with bilateral spontaneous isolated internal carotid artery dissections, which caused an attack of amaurosis fugax a few months earlier. The patient was asymptomatic during the admission with type B aortic dissection. However, the patient had a high blood pressure which was medically treated. A new CT scan confirmed earlier findings and revealed a spontaneous isolated dissection in the superior mesenteric artery. No progression was seen when the scan was compared to a new CT scan performed 10 days later. The type B aortic dissection was considered to be chronic and stable with no need for vascular intervention. This case report illustrates a rare condition of four isolated arterial dissections. The present case demonstrates the necessity of further examinations, which should be considered carefully when a patient presents with several independent arterial dissections.

Keywords: Dissection, internal carotid artery dissection, multiple artery dissections, spontaneous dissection, superior mesenteric artery dissection, type B aortic dissection

Introduction

Patients with several spontaneous isolated arterial dissections are rare. Arterial dissections can result in high morbidity and mortality, as these patients can represent some of the most serious vascular emergencies. However, some patients are asymptomatic despite having dissections located in arteries known to be highly symptomatic and fatal.

A dissection is a result of a tear in the tunica intima, the arterial inner wall, which creates a false lumen in the vessel wall. This may compromise branch vessels, which results in symptoms of organs and tissue supplied by the vessel. One of the most important vessel dissections to know as a physician, is aortic dissection, which is classified according to Standford type A and B. Type A and B refer to the localization of the inlet either being proximal or distal to the left subclavian artery, respectively. The annual incidence has been

reported to be 7.2 per 100,000 [1]. The clinical presentation ranges from incidental finding with no symptoms to severe chest/abdominal and back pain or sudden death.

Spontaneous isolated dissections in the internal carotid artery can cause stroke and are overrepresented in younger individuals presenting with stroke compared to older. The incidence is estimated to be around 3 per 100,000 for spontaneous dissection [2], while the incidence of dissections in the bilateral carotid artery is estimated to be 1/5 of these cases [3, 4]. Clinical presentation varies from being asymptomatic to a condition with transient cerebral ischemia and classical signs of stroke.

Spontaneous isolated superior mesenteric artery dissection (SISMAD) is a condition with a reported incidence of 9 per 10,000 [5]. The presentation varies from the patient being asymptomatic to acute abdominal pain due to e.g. bowel ischemia or peritonitis.

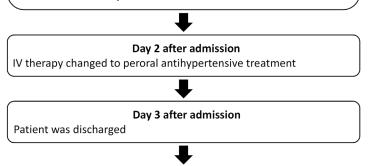
Six months prior to presentation

Attack of amaurosis fugax. CT scan revealed bilateral dissections in internal carotid arteries. Discharged with normal vision few days later



Admission to the Department of Cardiology

Routine CT scan showed aortic dissection type B. Patient was asymptomatic. Blood pressure was treated with IV Labetolol. New ECG-gated CT scan showed organs supplied from the true lumen and a separate dissection in the superior mesenteric artery. Stationary internal carotid artery dissections



Day 10 after admission

CT scan with no worsening. Planned for a new CT scan and clinical control in 1 year

Figure 1. Timeline of case report.

Common for all three abovementioned types of arterial dissections is that the aetiology can be spontaneous, due to trauma or secondary to genetic connective tissue disorders e.g. Ehlers-Danlos syndrome, Marfan syndrome or fibromuscular dysplasia.

To our knowledge, a combination of isolated dissections in aorta, bilateral carotid arteries and superior mesenteric artery has not been reported previously. In this case report, we present a patient case with spontaneous isolated dissections in four vessels.

History of presentation

A 72-year-old man was admitted at the Department of Cardiology after a routine thoracic CT scan revealed a type B aortic dissection. The CT scan was performed due to a malignant suspected infiltration in the right upper lung.

Past medical history

The patient had a medical history of being a former smoker, hypertension, polymyalgia rheu-

matica and bilateral internal carotid artery dissections. Bilateral internal carotid artery dissections were discovered during an admission 6 months prior on a CT scan performed due to an episode of amaurosis fugax (Figure 1). At that time, no other dissections were seen on the CT scan. The patient was discharged after a few days of admission without complaints or abnormal clinical findings. The family medical history included a father who suffered from stroke and a mother with breast cancer. No sudden cardiac deaths or death at birth were reported in the family.

Investigations

On admission to Department of Cardiology, the patient was asymptomatic, denying any recent symptoms of dyspnoea, or chest or back pain. However, the patient had severe backpain when performing physical work a few years ago.

Examination by the physician at time of admission was normal. The patient showed no phenotypical signs of any connective tissue diseases such as Marfan or Ehlers-Danlos syndrome. Blood chemistry revealed a C-reactive protein of 29 mg/L, a D-dimer of 0.7 FEU/L and normal arterial blood gas.

Management

The blood pressure was initially treated with intravenous Labetalol (Table 1). The initial CT scan did not include arcus aorta nor the head or neck vessels. To assess if thoracic endovascular aortic repair (TEVAR) was an option, a new electrocardiogram-gated CT scan with contrast was performed. The scan revealed a type B aortic dissection originating 5 cm above the diaphragm, extending to the left common iliac artery (Figure 2). There was a dissection membrane in the superior mesenteric artery, which was without connection to the aortic dissection. The entire extent of the false lumen of the aortic dissection had flow. A small haematoma was found in the descending aorta and a bigger inlet 5 cm above the diaphragm, but no extra-

Table 1. Vital parameters at admission

Blood pressure	152/90 mmHg
Heart rhythm	55 beats/minute
Respiratory rate	16 breaths/minute
Oxygen saturation (Periphery pulse oximetry)	96%
Temperature	36.8°C

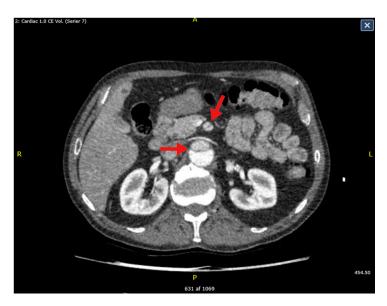


Figure 2. CT scan showing the dissection in aorta and superior mesenteric artery.



Figure 3. CT scan showing bilateral internal carotid artery dissections.

vascular bleeding was found. The maximal diameter of aorta was 45 mm. Retrospectively, it was possible to identify a little calcified plaque in the middle of the lumen of the

descending aorta, located at the origin of the dissection membrane, in the previous CT scan performed 3 months prior. Thus, it was concluded that the aortic dissection was already present for at least 3 months prior. No visible dissections were found in the neck vessels near aorta, but the dissections in the internal carotid arteries were unchanged during the past 6 months, which was the reason for no urgent intervention (Figure 3).

On the second day of admission, Labetolol treatment was discontinued and oral antihypertensive treatment was intensified. The patient was discharged after three days of admission and had a CT scan 10 days later, which showed stationary conditions. The patient was planned for a clinical control CT scan with a 1-year follow up.

Discussion

In this case, the patient presented with four spontaneous isolated arterial dissections. The patient was first diagnosed with spontaneous isolated dissections in the bilateral internal carotid arteries which resulted in amaurosis fugax. Later, the patient was diagnosed with asymptomatic type B aortic dissection. Furthermore, an asymptomatic dissection in the superior mesenteric artery was revealed. The type B aortic dissection was considered stable and chronic. Intervascular intervention was considered unnecessary.

Only few case reports exist describing multiple spontaneous dissections as presented in this case, especially with the rare combination of four spontaneous

dissections in different vessels (**Table 2**). However, the prevalence of dissections may be underreported as the patients can be asymptomatic.

Multiple spontaneous arterial dissections

Table 2. Publications of case reports with multiple arterial dissections with involvement of 3 or more dissections

Article	Year	Gender	Age	Comorbidity	Symptoms	Dissections	Treatment
Spontaneous multiple arterial dissections presenting with renal infarction and subarachnoid hemorrhage in a patient under treatment for infertility [8]	2005	Female	36	5 years infertility treatment	Left flank pain, nausea and vomit. On 4^{th} day of admission, the patient lost consciousness	4 dissections (Bilateral renal arteries (RA) and superior mesenteric arteries (SMA) on admission day On 4 th day: dissection of right vertebral artery)	Heparin injection for renal infarction Intravascular embolization
Spontaneous bilateral carotid and vertebral artery dissections associated with multiple disparate intracranial aneurysms, subarachnoid hemorrhage and spontaneous resolution. Case report and literature review [9]	2007	Female	29	None	48 hours of confusion culminating in total collapse. Neck and shoulder pain and affected consciousness	3 dissections (Left vertebral artery, right common carotid artery and distal cervical left internal carotid artery). Furthermore, basilar aneurysm and posterior cerebral artery (PCA) aneurysm was found.	Ventricular drainage, craniotomy, clipped PCA aneurysm, low molecular weight heparin
Spontaneous Multiarterial Dissection Immediately after Childbirth [10]	2012	Female	32	Papillary thyroid cancer	Intermittent neck and chest pain starting immediately after delivery. One week post-partum, patient presented with intermittent chest and neck pain. 6 days later neck pain radiating to chest	3 dissections (Left vertebral artery, proximal left anterior coronary artery (LAD) and left internal mammary artery)	Coronary artery bypass grafting (CABG) with saphenous vein graft to LAD
Sequential multiple visceral arteries dissections without aortic involvement [11]	2013	Male	54	Hypertension, diverticular disease and smoking	Sudden upper back and abdominal pain At day 10 new onset of severe thoracic and abdominal pain After 2 months: severe upper back pain	4 dissections (On admission: SMA Day 10: Worsening of SMA + carotid artery (CA) + bilateral RA's After 2 months: stationary conditions)	On admission: conservative strategy On day 10: Endovascular repair of SMA After 2 months: observation and discharge
Spontaneous bilateral cervical internal carotid and vertebral artery dissection in a Japanese patient without collagen vascular disease with special reference to single-nucleotide polymorphisms [12]	2016	Male	52	Migraine with aura	Headache without aura	4 dissections (Left internal carotid artery, right internal carotid artery and bilateral vertebral arteries)	Not reported
Spontaneous dissections of multiple visceral arteries: an extremely rare case [13]	2017	Male	55	Frequent alcohol consumption	Diffuse abdominal pain and nausea. Vomiting, diarrhea and abdominal distention	4 dissections (Proximal celiac artery, superior mesenteric artery and bilateral renal artery)	Conservative strategy and endovascular intervention on day 3
Bilateral vertebral artery dissection and unilateral carotid artery dissection in case of Ehlers-Danlos Syndrome Type IV [14]	2018	Female	50	None reported at admission. Later diagnosed with Ehlos-Danlos type IV	Severe headache. Developed right cervical pain and dizziness at 3 days after admission	3 dissections (Bilateral vertebral artery and right internal carotid artery)	Initially conservatively, but later stented
A rare case of 7 simultaneous arterial dissections and review of the literature [15]	2019	Female	51	Smoking, migraine, appendectomy and caesarian delivery	Sudden onset right flank pain. Within 24 hours abdominal pain	7 dissections (Celiac trunk, splenic artery, right renal artery, inferior mes- enteric artery, bilateral external iliac arteries and right carotid artery)	Endovascular repair with stent graft
Spontaneous dissections involving multiple coronary arteries and a vertebral artery over 7 years [16]	2019	Female	38	None reported	2011: Chest pain after delivery 2017: Syncope 2018: Chest pain after delivery and VF requiring defibrillation	5 dissections 2011: Spontaneous carotid rtery dissection (SCAD) in LAD and right coronary artery (RCA). 2017: Distal right vertebral artery. 2018: SCAD of left circumfex arter (LCx) and first obtuse marginal (OM1)	2011: CABG 2017: Conservative strategy 2018: Conservative strategy and implanta- tion of implantable cardioverter defibrillator (ICD) for secondary prevention

Multiple spontaneous arterial dissections

Spontaneous Bilateral Internal Carotid and Vertebral Artery Dissections with Dominant-hemisphere Circulation Maintained by External Carotid Artery- ophthalmic Artery Anastomoses [17]	2019	Female	49	Migraine and C6-7 cervical discectomy and fusion	3 hours of mild right-hand weakness and mild headache	4 dissections (Bilateral internal carotid and vertebral artery)	Endovascular intervention
Multiple cervical artery dissections after alemtuzumab [18]	2019	Female	40	Rapid remitting multiple sclerosis (RRMS), orthostatic hypotension and bradycardia when treated with high dose steroids	Three days after alemtuzumab, the patient complained of painful visual blur. The day after she developed acute aphasia and right hemiplegia	3 dissections (Bilateral carotid arteries and right vertebral artery)	Heparin and antiplatelet therapy
Endovascular stenting in a rare case of multiple spontaneous visceral arterial dissections [19]	2020	Male	Middle aged	None	Sudden-onset sharp left sided back pain and costovertebral angle tenderness	3 dissections (Celiac artery and bilateral renal arteries)	Stenting the bilateral renal arteries
Spontaneous multiple arterial dissection in a COVID-19-positive decedent [20]	2022	Male	40	Unspecified mito- chondrial myopathy	At admission: weakness, fatigue, short- ness of breath, fever Day 11: Abdominal pain and shortness of breath	4 dissections (Splenic artery, inferior mesenteric artery and bilateral renal arteries)	Unsuccessful cardiopulmonary resuscitation

RA: Renal artery; SMA: Superior mesenteric artery; PCA: Posterior cerebral artery; LAD: Left anterior descending coronary artery bypass graft; CA: carotid artery; SCAD: Spontaneous coronary artery dissection; RCA: Right coronary artery; LCX: Left circumflex; OM1: First obtuse marginal; ICD: Implantable cardioverter-defibrillator; RRMS: Rapid remitting multiple sclerosis; CPR: Cardiopulmonary resuscitation.

The aetiology may often remain unknown. Cardiovascular disease and hypertension have been suggested as risk factors to arterial dissections. Among inherited conditions such as Ehlers-Danlos, Marfan and Loyez-Deitz syndromes, less than 3 visceral arteries are usually involved [13]. In this case, the aetiology was unclear. However, the patient had underlying risk factors (Former smoker, male gender, the patient's age and familial disposition to stroke). Spontaneous multiple arterial dissections can be caused by monogenic connective tissue disorders such as Ehlers-Danlos, Marfan and Loyez-Deitz syndromes. One case report found that patients with multivessel arterial dissections were more likely to be <45 years of age, more likely to have associated pseudoaneurysms and more likely to have involvement of extracranial cerebral arteries [4]. Furthermore, they found that the patients had a higher incidence of fibromuscular dysplasia when compared to patients with single arterial dissections [4]. A different study from 2008 found that patients with multiple extracranial arterial dissections had impaired vasomotion, which might predispose them to dissections [6].

Because of the rare incidence of multiple arterial dissections in general, there is no consensus on the diagnostic approach or therapeutic strategy. Moreover, only very few case reports have a long follow up time. There are currently no clinical guidelines suggesting follow up protocols. However, a review from 2014 regarding dissections in the mesenteric artery, concluded that medical therapy was indicated with intensive surveillance. Furthermore, invasive intervention could be necessary if signs of ischemia, aneurismal enlargement or rupture were developed [7]. This is consistent with the approach for this patient.

Conclusion

In conclusion, we report a rare case of multiple spontaneous arterial dissections including aorta, which has not been reported previously. A conservative strategy was chosen with antihypertensive treatment and no worsening was observed on the control CT scan 10 days after discharge compared to the scan at admission.

Disclosure of conflict of interest

None.

Abbreviations

ACA, Anterior cerebral artery; CA, Celiac artery; CABG, Coronary artery bypass grafting; CPR, Cardiopulmonary resuscitation; ECG, Electrocardiogram; ICA, Internal carotid artery; ICD, Implantable cardioverter-defibrillator; LAD, Left anterior descending artery; PCA, Posterior cerebral artery; RA, Renal artery; RCA, Right coronary artery; RRMS, Relapsing remitting multiple sclerosis; SCAD, Spontaneous coronary artery dissection; SMA, Superior mesenteric artery; VA, Vertebral artery; VF, Ventricular fibrillation.

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References

- [1] Smedberg C, Steuer J, Leander K and Hultgren R. Sex differences and temporal trends in aortic dissection: a population-based study of incidence, treatment strategies, and outcome in Swedish patients during 15 years. Eur Heart J 2020; 41: 2430-2438.
- [2] Schievink WI, Mokri B and Whisnant JP. Internal carotid artery dissection in a community. Rochester, Minnesota, 1987-1992. Stroke 1993; 24: 1678-1680.
- [3] Townend BS, Traves L and Crimmins D. Bilateral spontaneous carotid artery dissection. J Clin Neurosci 2005; 12: 592-594.
- [4] Hassan AE, Zacharatos H, Mohammad YM, Tariq N, Vazquez G, Rodriguez GJ, Suri MF and Qureshi AI. Comparison of single versus multiple spontaneous extra- and/or intracranial arterial dissection. J Stroke Cerebrovasc Dis 2013; 22: 42-48.
- [5] Foord AG and Lewis RD. Primary dissecting aneurysms of peripheral and pulmonary arteries: dissecting hemorrhage of media. Arch Pathol 1959; 68: 553-577.
- [6] Baracchini C, Tonello S, Vitaliani R, Giometto B, Meneghetti G and Ballotta E. Vasomotion in multiple spontaneous cervical artery dissections. Stroke 2008; 39: 1148-1151.
- [7] Garrett HE Jr. Options for treatment of spontaneous mesenteric artery dissection. J Vasc Surg 2014; 59: 1433-1439, e1-2.
- [8] Wakino S, Tawarahara K, Tsuchiya N, Kurosawa Y, Sugihara T and Ando K. Spontaneous multiple arterial dissections presenting with renal infarction and subarachnoid hemorrhage

Multiple spontaneous arterial dissections

- in a patient under treatment for infertility. Circ J 2005; 69: 368-372.
- [9] Marshman LA, Ball L and Jadun CK. Spontaneous bilateral carotid and vertebral artery dissections associated with multiple disparate intracranial aneurysms, subarachnoid hemorrhage and spontaneous resolution. Case report and literature review. Clin Neurol Neurosurg 2007; 109: 816-820.
- [10] Brantley HP, Cabarrus BR and Movahed A. Spontaneous multiarterial dissection immediately after childbirth. Tex Heart Inst J 2012; 39: 683-686.
- [11] Bonardelli S, Battaglia G, Zanotti C, Cervi E, Guadrini C and Giulini SM. Sequential multiple visceral arteries dissections without aortic involvement. Ann Vasc Surg 2013; 27: 497, e9-13.
- [12] Abe A, Nito C, Sakamoto Y, Nogami A, Hokama H, Takahashi S, Kirita K, Ueda M, Ishimaru Y and Kimura K. Spontaneous bilateral cervical internal carotid and vertebral artery dissection in a Japanese patient without collagen vascular disease with special reference to singlenucleotide polymorphisms. J Stroke Cerebrovasc Dis 2016; 25: e114-7.
- [13] Le TB, Jeon YS, Hong KC, Cho SG and Park KM. Spontaneous dissections of multiple visceral arteries: an extremely rare case. Ann Surg Treat Res 2017; 92: 225-229.
- [14] Ohshima T, Miyachi S, Isaji T, Matsuo N, Kawaguchi R and Takayasu M. Bilateral vertebral artery dissection and unilateral carotid artery dissection in case of Ehlers-Danlos syndrome type IV. World Neurosurg 2019; 121: 83-87.

- [15] Amitai Komem D, Sukenik Halevy R, Griton Y, Shnaker A, Levy Y, Weissmann J and Fajer S. A rare case of 7 simultaneous arterial dissections and review of the literature. Vasc Endovascular Surg 2019; 53: 617-622.
- [16] Khalid N, Rogers T and Waksman R. Spontaneous dissections involving multiple coronary arteries and a vertebral artery over 7 years. Eur Heart J 2019; 40: 322.
- [17] Golub D, Hu L, Dogra S, Torres J and Shapiro M. Spontaneous bilateral internal carotid and vertebral artery dissections with dominanthemisphere circulation maintained by external carotid artery-ophthalmic artery anastomoses. Neurosurg Focus 2019; 46: E6.
- [18] Durand-Dubief F, Marignier R, Berthezene Y, Cottin J, Nighoghossian N and Vukusic S. Spontaneous multiple cervical artery dissections after alemtuzumab. Mult Scler 2020; 26: 381-383.
- [19] Moran J, Galla N and Ranade M. Endovascular stenting in a rare case of multiple spontaneous visceral arterial dissections. Vasc Endovascular Surg 2021; 55: 269-272.
- [20] James C and Peterson DC. Spontaneous multiple arterial dissection in a COVID-19-positive decedent. Am J Forensic Med Pathol 2022; 43: 52-54.