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Aortic Dissection: An Easily Missed Diagnosis when Pain Doesn't Hold the Stage

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Case series
Patients: Male, 73-year-old • Female, 70-year-old
Final Diagnosis: Aortic dissection
Symptoms: Paresis
Medication: —
Clinical Procedure: —
Specialty: Neurology

Objective: Challenging differential diagnosis

Background: Type A aortic dissection (AD) is a rare disease, with a high mortality rate. Its most common symptom is thoracic pain, which is nevertheless absent in about 6% of cases. Neurologic complications are extremely rare and include ischemic stroke and ischemic neuropathy (which are the most common as presenting symptoms), spinal cord ischemia, and hypoxic encephalopathy. These rare neurological presentations can often be missed at initial clinical examination.

Case Report: We report 2 cases of patients presenting with seemingly mild neurological symptoms. However, diagnostic tests revealed acute type A AD, and further steps were taken.

Conclusions: Although it is a rare cause of transient stroke or peripheral nerve ischemia, AD should be quickly recognized as a potential cause of new-onset neurological manifestations.

MeSH Keywords: Aortic Diseases • Ischemic Attack, Transient • Paresis

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/917179

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Background

Aortic dissection (AD) is a life-threatening surgical condition, with an expected incidence of 5 to 30 per 1 million persons per year, affecting mostly the male population [1]. The mortality rate is high, even with proper treatment, the prognosis being comparatively worse in women [2]. Age is a very important factor regarding the onset of symptoms. Age is also a key factor concerning the morpho-pathological features of the dissections afflicting the ascending aorta: most of the older patients have atheromatosis, a history of coronary angioplasty, or an intramural hematoma, while in young patients a connective tissue disorder is the most likely histological discovery. Ascending aorta dissections are classified as type A in the Stanford system, and either as type 1 (those originating in the ascending aorta and extending at least to the aortic arch) or type 2 (those limited to the ascending aorta) in the DeBakey system [3]. About 6% of patients with type A AD do not complain of pain [4]. Some of these present with neurological symptoms or heart failure [5]. Given their rarity, neurological presentations of AD can often be missed at initial clinical examination, a fact that is highlighted in the present paper, in which we summarized the cases of 2 patients with acute type A AD who presented with transient and painless neurological manifestations, all of which were caused by arterial obstruction.

Case Report

Case 1

A 73-year-old white male with a history of diabetes mellitus, arterial hypertension, New York Heart Association (NYHA) class II heart failure, atrial fibrillation, and mechanical aortic valve replacement (10 years previously) for severe aortic stenosis, who was on oral anticoagulant therapy with an international normalized ratio (INR) in the range of 4, was hospitalized for 2 transient episodes of left hemiparesis and dysarthria, with relapse of symptoms after admission. His daily medication consisted of amlodipine, ramipril, metformin, and acenocoumarol. On physical examination there were no pathological findings. The patient was afebrile, without any localized pain, with normal blood pressure (120/80 mmHg, under antihypertensive therapy). Cardiac auscultation showed a regular rate and rhythm without any murmur, while assessment of the peripheral vascular system found symmetrical pulse in all peripheral arteries. The neurological examination at the time of admission did not reveal any neurological deficits, the clinical picture being suggestive of a right carotid transient ischemic attack. The cerebral computed tomography (CT) examination at admission highlighted only an ischemic stroke sequela in the right posterior cerebral artery territory, multiple chronic lacunar strokes in the basal ganglia and bilaterally in the thalamus, with no evidence of acute ischemic lesions, which might explain the transient neurologic deficits. Electrocardiographic (ECG) examination revealed sinus rhythm, 80 beats per minute, and atypical left bundle branch block (also present on an ECG recorded 2 years ago). Cervical and transcranial Doppler showed normal flow rates in the cervico-cerebral arteries, without any arterial stenosis. Blood analyses highlighted slight hyperglycemia (180 mg/dL), the patient being known with diabetes mellitus, and an INR value of 4 in the context of warfarin treatment, with no other abnormalities. We have not determined the troponin level at the time of the hospital admission, as the patient did not show any clinical or ECG signs of myocardial infarction. Given the lack of carotid stenosis and the presence of aortic mechanic valve, the possibility of a valvular vegetation was taken into consideration. Transthoracic echocardiography did not reveal any vegetation, but demonstrated instead a dissection flap with intraluminal thrombus in the ascending aorta and moderate aortic regurgitation. The intimal flap did not involve the sinus of Valsalva. Chest CT and angio CT confirmed the existence of a dissection flap at the origin of the aorta (Figure 1A) and extending into the ascending aorta (Figure 1B) over a distance of approximately 50 mm, with a parietal thrombus of maximum diameter of 8 mm. In the ascending aorta the maximal axial diameters were 58/48 mm, while the minimal axial dimensions were approximately 32/50 mm. The aortic arch and the descending aorta

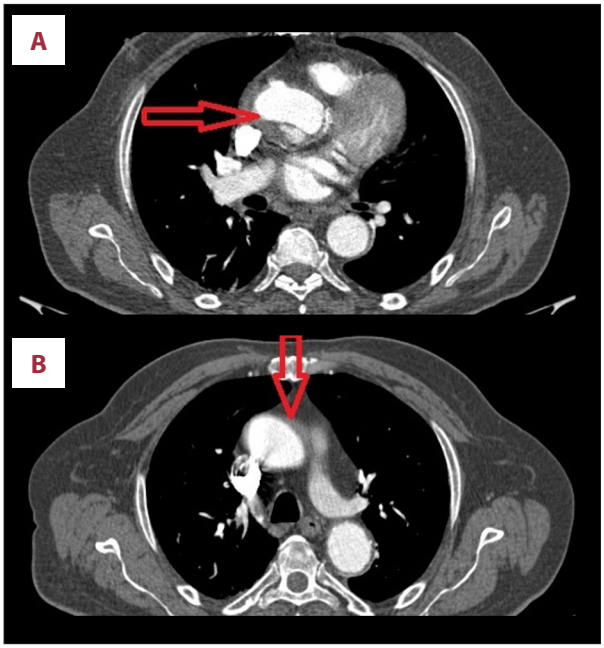

Figure 1. (**A**) Case 1: computed tomography angiography of the aorta showing the true and false lumen at the origin and filling defect-clot (horizontal red arrow). (**B**) Case 1: a true and false lumen of ascending aorta with a parietal thrombus (vertical red arrow).

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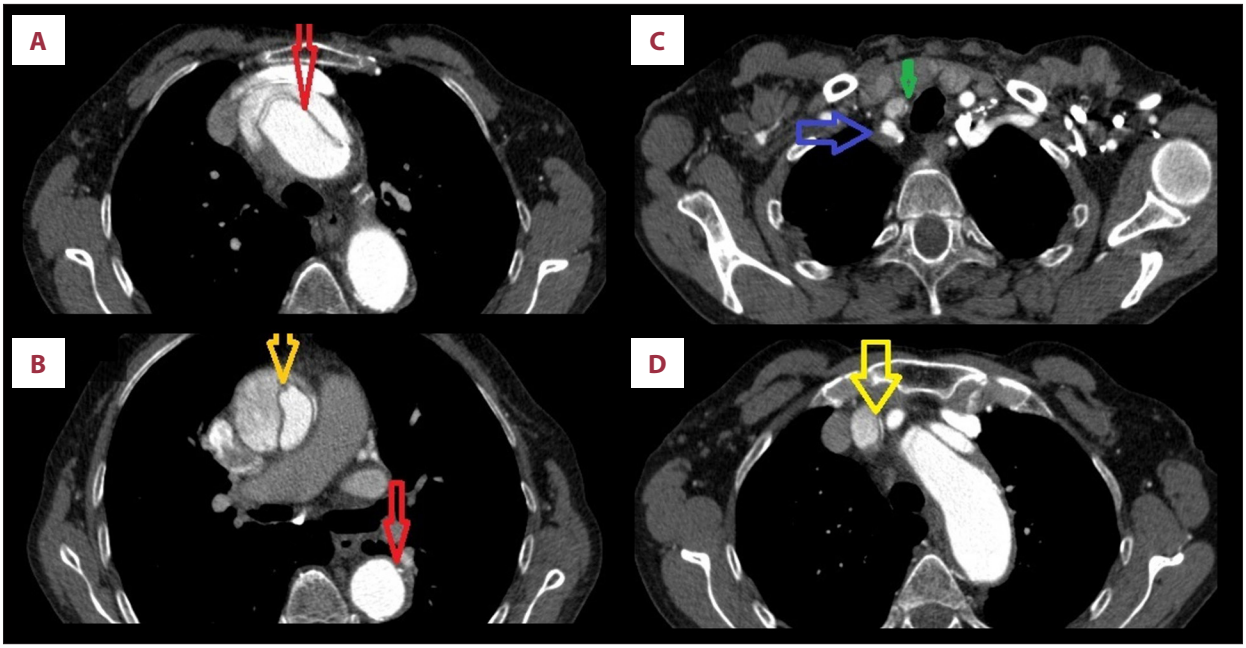

Figure 2. (**A**) Case 2: computed tomography angiography image shows aortic arch dissection (red arrow). (**B**) Case 2: thrombosis of the false lumen in the descending thoracic aorta (red arrow) and flap of dissection into ascending aorta (yellow arrow). (**C**) Case 2: dissection of right common carotid artery (green arrow) and right subclavian artery with filling defect-clot (blue arrow). (**D**) Case 2: dissection of the brachiocephalic trunk (yellow arrow).

appeared normal, so the dissection fold did not extend to the brachiocephalic trunk. We have no data regarding a possible dissection extension to the coronary arteries, as these were not evaluated. The left bundle branch block had been existing long before the current vascular event, so it could not have been explained by a concurrent coronary artery dissection.

After the patient was evaluated to determine his eligibility for cardiovascular surgery, we decided on conservative treatment. Even if the surgical intervention would have been indicated, it could have not been performed as the patient refused any invasive procedure. We initially treated him with unfractioned heparin (12 units/kg/hour) for 2 weeks and subsequently with acenocoumarol. During the entire hospitalization (4 weeks), the patient did not complain of any pain. The clinical and neurological evolution was uneventful, and the patient had no neurological deficits at discharge. We were not able to perform follow-up imaging because the patient refused any further examination. At the 3-month appointment, the patient was fully recovered without any neurological deficits or cardiac manifestations.

The most probable cause of the transient ischemic attacks in this patient was artery-to artery embolism from the mural thrombus. An atherothrombotic or hemodynamic mechanism was less plausible as ultrasonography revealed no stenosis in the right internal carotid and its branches. On the other hand, the diagnosis of AD was almost certain as the patient had some of the appropriate risk factors (gender, age, arterial hypertension) and CT-angiography is credited with 87% to 100% specificity (and 83% to 94% sensitivity) [6] for AD.

Case 2

A 70-year-old white female, with a history of rheumatoid arthritis (RA) (treated with methotrexate 7.5 mg as a single weekly dose and folic acid) and arterial hypertension (treated with candesartan 16 mg once daily) presented to the Emergency Department (ED) for a right transient brachial monoparesis without any other associated neurological signs. Physical examination revealed absent right radial and brachial pulse, but normal pulse in all other peripheral arteries. Blood pressure was undetectable in the right upper extremity, but normal in the left arm was 125/75 mmHg. Cardiac auscultation revealed a regular rate and rhythm, without any murmur. The patient denied any significant thoracic, neck, or abdominal pain but described a slight, intermittent interscapular discomfort. The patient stated that this manifestation had been episodic in nature for the last few months and assigned it to RA, and therefore she didn't ask a physician to evaluate her, consequently no clinical, laboratory or imagistic assessment regarding the cause of this symptom was performed prior to her current presentation at the ED. She denied that this episodic pain was associated with high blood pressure (she stated that under her current medication her blood pressure was controlled). On neurological examination, the patient did not

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exhibit any abnormalities. Given the clinical context of the symptoms (lack of pulse in the radial and the right brachial arteries and transient monoparesis, as well as the slight interscapular discomfort), ischemic neuropathy in the context of a type A AD rather than related to RA was suspected. ECG examination revealed sinus rhythm, 64 beats per minute, without pathological changes. Echo Doppler ultrasound highlighted low velocity monophasic waveform in the right subclavian and common carotid arteries and a dissection flap with intraluminal thrombus. The left common carotid and subclavian arteries were normal. Transthoracic echocardiography revealed numerous atheromas in the aortic root and dissection of the ascending aorta and aortic arch with extension of the dissection into the right common carotid artery. Additionally, moderate aortic insufficiency (presumably secondary to AD) was noticed with reversed flow in the descending aorta, representing approximately 50% of the anterograde flow, as evaluated by means of velocity-time integral. The flap most probably involved the aortic valve, as the patient had aortic regurgitation.

Chest computed tomography and angio CT confirmed the presence of the dissection flap starting at the origin (Figure 2A), extending into the ascending aorta (Figure 2B) and the aortic arch, and further into right common carotid artery (Figure 2C), subclavian artery (Figure 2C), and brachiocephalic trunk (Figure 2D), as well as the thrombosis of the false lumen in the descending thoracic aorta (Figure 2B). The patient was transferred to another hospital and underwent open-heart surgery to repair the vessel (24 hours after symptoms onset). Excision of the intimal tear and obliteration of entry into the false lumen were performed initially. Afterwards, the aorta was reconstructed with interposition of a synthetic vascular graft. Unfortunately, in the early postoperative period after surgical repair, the AD was complicated by an acute right hemispheric ischemic stroke, probably due to intraoperative brain malperfusion. Blood pressure level was closely controlled during the surgery, thus there is no possibility that an uncontrolled blood pressure could have caused an extension of the flap responsible for the ischemic stroke. The patient died 5 days later.

Discussion

Classically, the clinical picture has an abrupt onset and peaks over the following minutes to hours [11]. According to the International Registry of Acute Aortic Dissection (IRAD), painless type A AD was associated with a worse prognosis (1% to 2% per hour mortality risk if left untreated) leading to congestive heart failure, ischemic cerebral events, and comparatively more syncopal episodes [7]. The prevalence of ischemic stroke is approximately 10% in all patients with AD [8]. Ischemic strokes in patients with AD is a consequence of either stenosis or arterial occlusions or probably caused by shock secondary to cardiac tamponade. The 2 possible causes of arterial occlusion are the expansion of the dissection in the major branches such as the brachiocephalic trunk or common carotid arteries, and thromboembolism from the aorta into the major arteries [9]. The topography of stroke secondary to AD involves more commonly the anterior circulation than the posterior circulation [10,11] because these great arteries originate precisely from the aortic arch, while the vertebral arteries emerge from the subclavian arteries [10]. Only one-fifth of patients with impaired supra-aortic vessels have a preoperative ischemic stroke, the rest being asymptomatic [12]. Mortality was not found to be higher among patients with stroke, if the AD was rapidly diagnosed [12].

After the stroke, which is the most common initial neurological finding [12], ischemic neuropathy represents the second neurological manifestation in the context of an AD. Neuropathy is the consequence of dissection extending into a major artery of the extremities (aortic bifurcation, iliac, femoral and subclavicular arteries), interfering with the blood supply of that limb, thus affecting the vasa nervorum of the peripheral nerves. The progression to ischemic necrosis of the extremities is largely dependent on the degree of collateral circulation [12]. The clinical manifestations of an ischemic neuropathy are variable [severe, distal pain in the extremity (not conforming to the distribution territory of their nerve trunks), numbness, coldness, paresthesia, pulse deficit] but more frequently a monoparesis or unilateral distal limb numbness are found at examination [12]. Our cases are remarkable by the absence of pain and by the low intensity and varied nature of the symptoms. In the first described case, although the symptoms suggested an internal carotid artery stenosis, no atheromatosis was found on ultrasound examination. Another possible mechanism could be cardioembolism, originating from a thrombus obstructing the prosthetic metallic valve. Thromboembolism is the most common complication of mechanical valves; however, left atrium enlargement and atrial fibrillation could increase the risk of cardioembolism, even in patients on the correct anticoagulation treatment [13]. The AD was a surprising finding taking into account the absence of pain, and the fact that AD after aortic valve replacement is a rare complication [14,15], occurring in only about 1% of cases [15]. The second patient was found to have transient right arm paralysis that was relieved in 5 minutes with no pain or paresthesia, but with pulse deficit in the right brachial and radial arteries. There is evidence that up to 85% cases of RA show clinical manifestations of neuropathy: mononeuritis multiplex, sensorimotor neuropathy, and entrapment neuropathy attributed to drug toxicity, amyloidosis, an autoimmune mechanism [16], or necrotizing vasculitis of the vasa vasorum [17]. On the other hand, cardiac disease (ischemic heart disease, cardiomyopathy, pericarditis, vasculitis of coronary artery, arrhythmia, and valve diseases) is a well-recognized complication in RA patients [18].

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However, we cannot exclude a causal link between the AD and RA. We found a case report in the literature stating that AD may be related to RA through the phenomenon of vasa vasorum ischemia [17]. The ischemic neuropathy in our second patient could be attributed to the occlusion of the brachiocephalic trunk and subclavian artery, these arteries being involved in AD. Neurological rapid improvement in such cases is probably the result of only transient arterial occlusion when dissection is propagating [9]. However, in the second patient it is difficult to differentiate a transient paralysis secondary to ischemic neuropathy from lack of blood flow due to AD with malperfusion of the right upper extremity. Limb ischemia secondary to AD is caused by involvement of a side branch orifice into the dissection or obliteration of the true lumen by an expanding false lumen. On the other hand, ischemic neuropathy could be attributed to occlusive dissection of the subclavian artery affecting the vasa nervorum of the peripheral nerves, as described in other case reports were limbs paralysis secondary to AD were classified as ischemic neuropathy [19].

It should be noted that ischemic events affecting other territories (such as lower limbs) may be the dominant clinical feature of AD [20]. Hence, the main symptom at presentation of AD may be not thoracic pain but ischemia of one or multiple organs, which may eventually lead to treatment directed to the affected territory. In some of these cases, targeting only the ischemic organ may be feasible, provided that the patient is afterwards closely monitored.

Conclusions

Atypical presentation of AD (no pain and neurological symptoms dominating the clinical picture) may be misleading, resulting in misdiagnosis: putting forward a neurological diagnosis and missing AD altogether, and therefore the opportunity to timely perform the appropriate investigational and therapeutic procedures. Therefore, being a vital emergency, AD should be part of the differential diagnosis in patients with neurologic manifestations, even when another mechanism seems more plausible and the telltale manifestations of AD are absent.

Conflicts of interest

None.

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