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Case Report

Acute aortic dissection-induced paraplegia and respiratory distress mimicking the ascending flaccid paralysis of Guillain-Barré syndrome

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Abstract: Aortic dissection is a devastating disease with protean clinical manifestations. Given the high mortality within the initial few hours after onset, it is important to maintain a high level of suspicion for this diagnosis. Guillain-Barré syndrome (GBS) is an autoimmune disorder of the peripheral nervous system and is the most common cause of acute flaccid paralysis since the worldwide eradication of poliomyelitis. Here, we report on a 55-year-old woman who presented with aortic dissection-induced paraplegia and respiratory distress resembling the ascending flaccid paralysis of GBS. The patient had complained of progressive weakness of the lower extremities for 4 days. Ascending flaccid paralysis of GBS was suspected. However, chest computed tomography revealed expansion of the aorta with visible dissection, and acute aortic dissection was diagnosed. In conclusion, aortic dissection might be misdiagnosed due to its protean manifestations.

Keywords: Aortic dissection, Guillain-Barré syndrome, paraplegia, respiratory distress introduction

Introduction

Guillain-Barré syndrome (GBS) is an autoimmune disorder of the peripheral nervous system and is the most common cause of acute flaccid paralysis since the worldwide eradication of poliomyelitis. The clinical features of GBS include rapidly progressive weakness of the limbs and respiratory muscles and sensory dysfunction of the autonomic nerves [1]. A frequent mode of weakness progression is ascending paralysis, which was first described by Octave Landry. The initial paraplegia ascends quickly and may involve the respiratory muscles and cranial nerves. Ascending paralysis may sometimes arise from acute myelitis. In this case, the limb tone and muscle stretch reflexes may be reduced acutely or even absent, such as in spinal shock syndrome, leading to possible diagnostic confusion with GBS [2]. No case of ascending paralysis due to aortic dissection has been documented. Aortic dissection may have protean manifestations, including syncope, chest pain, stroke, paraplegia, anuria, pulse deficits, abdominal pain, back pain, anuria, acute congestive heart failure, and even sudden death [3]. We present a patient with paraplegia and respiratory distress resembling the ascending flaccid paralysis of GBS who was ultimately diagnosed with aortic dissection.

Case report

A 55-year-old woman had complained of progressive weakness in her lower extremities for 4 days and was referred to the Department of Neurology at the First Hospital of Jilin University. Before the onset of limb weakness, she lost consciousness for 2 days before regaining consciousness. Except for a history of cured synovitis in the knees and long-term smoking, she had been otherwise healthy, with no identified hypertension or prodromal infections. Two days

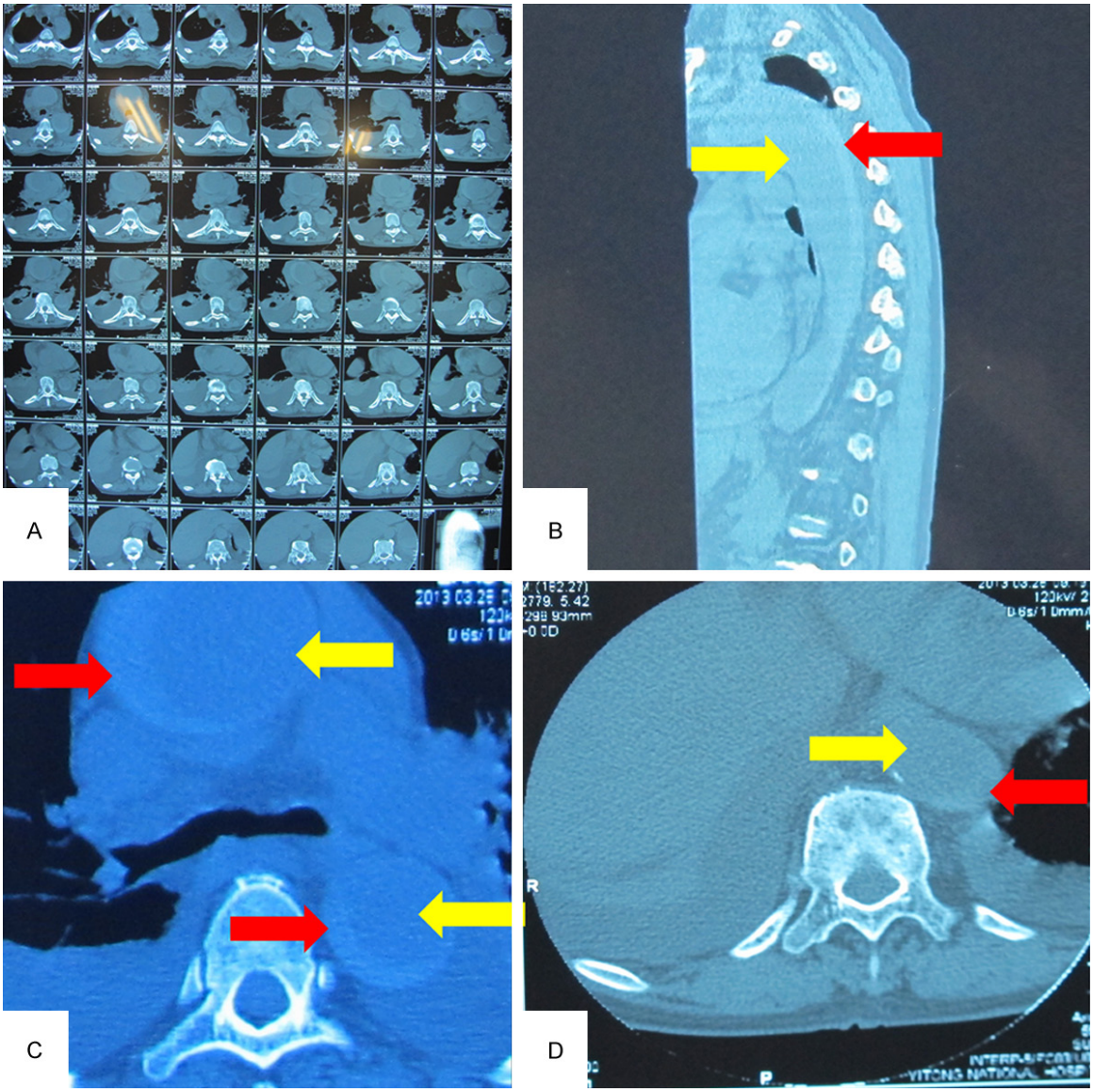

Figure 1. Chest and abdominal computed tomography (CT). (A) The chest CT images, including (B) sagittal and (C and D) axial images. In (B to D), the red arrows denote the false lumen, whereas the yellow ones denote the true lumen.

later, she complained of dyspnea and low-back pain. On general examination, she was afebrile and in an orthopnea position, with a pulse rate of 110/min, blood pressure of 158/77 mmHg, and a respiratory rate of 30/min. She was fully alert and oriented. Her neck was supple. No abnormalities were found in her cranial nerves. No involuntary movements, ataxia, or myoclonia were present. Areflexic paraplegia with muscle strength graded as 0/5 in both lower extremities was noted. Hypalgesia was noted below her knees bilaterally. Pulsation of the dorsal arteries was absent in both feet. No plantar reflexes were elicited. Babinski's sign was negative bilaterally. Head computed tomography (CT) was normal. Routine hematology studies were normal. Hepatic function tests revealed increased levels of aspartate transaminase [2473 (normal range 8-40) IU/L], alanine transaminase [1614 (normal range 8-50) IU/L], and alkaline phosphatase [134 (normal range 15-112) IU/L]. Her fasting blood glucose level was elevated [12.61 (normal range 3.9-6.1) mmol/L], as were the blood urea nitrogen [13.11 (normal range 2.5-6.1) mmol/L] and creatinine [148 (normal range 46-92) μmol/L]. The

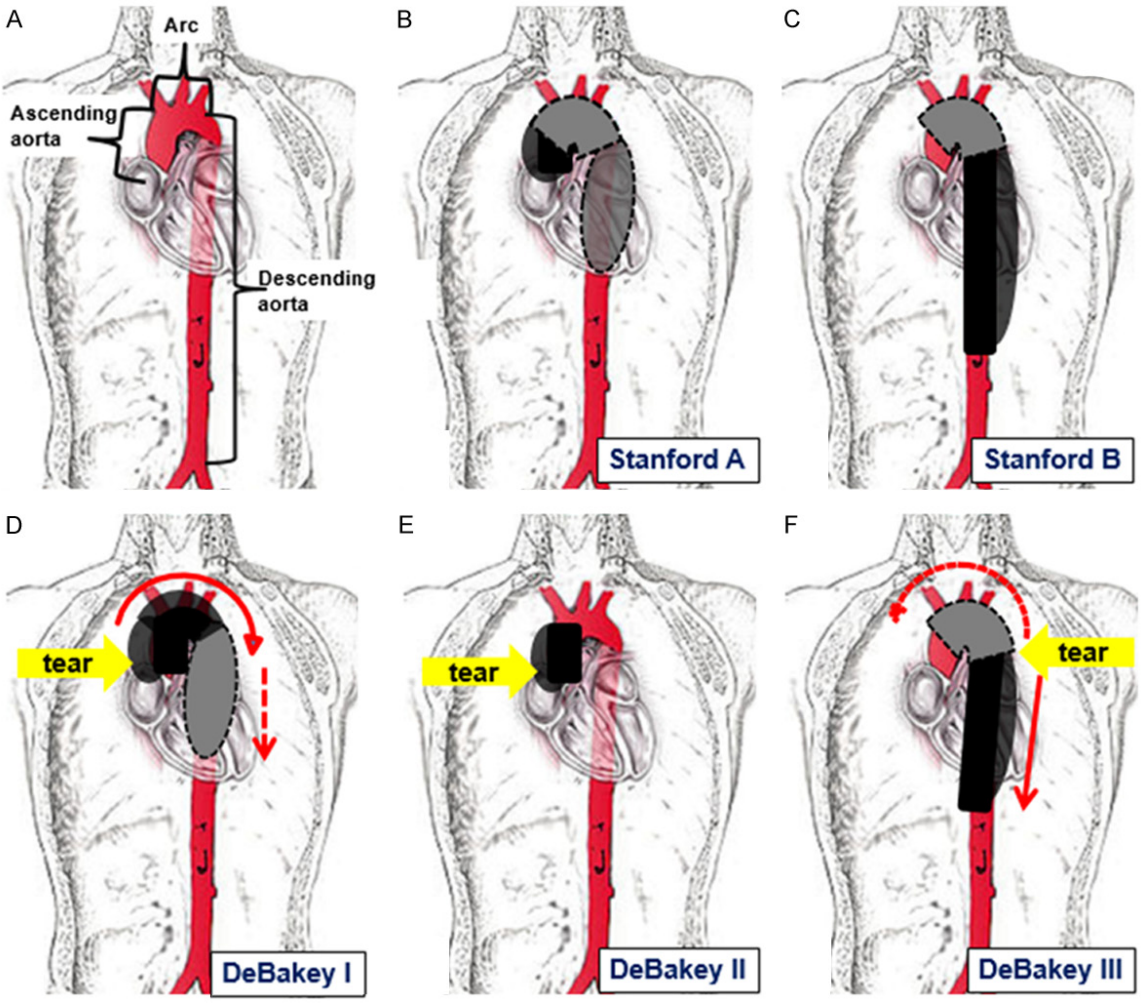

Figure 2. Schematic illustrations of aortic dissection and its classification. The classification schemes used most commonly are the Stanford and DeBakey systems. For purposes of classification, the ascending aorta refers to the aorta proximal to the brachiocephalic artery, and the descending aorta refers to the aorta distal to the left subclavian artery (A). According to the Stanford system, the ascending aorta is involved in type A (B) and not involved in type B (C), regardless of the point of origin. The DeBakey system categorizes dissections into types I, II, and III based on the origin of the intimal tear and extent of the dissection. According to this classification, dissections of type I (D) originate in the ascending aorta and propagate to the aortic arch, dissections of type II (E) originate in the ascending aorta and are confined to the ascending aorta, and dissections in type III (F) originate in the descending aorta, regardless of whether they propagate in a distal or in a retrograde manner. Surgical treatment is recommended in types I and II, but not in type III.

serum sodium [133 (normal range 137-145) mmol/L] and calcium [Ca^{2+} 1.90 (normal range 2.10-2.55) mmol/L] levels were reduced. The carbon dioxide combining power was reduced [14.1 (normal range 21-30) mmol/L]. The blood coagulation panel was abnormal [prothrombin time 22.6 (normal 9-13) s and activated partial thromboplastin time 41.6 (normal 20-40) s]. The antigen/antibody assay for hepatitis B virus was positive for HBsAg, HBeAg, and HBcAb; the antibody assay for hepatitis C virus was negative. Syphilis serology tests (rapid plasma regain), the anti-nuclear antibody series, and human immunodeficiency virus tests were all negative. A workup of tumor markers was also normal, including carcinoembryonic antigen, alpha-fetoprotein, carbohydrate antigen 242/199, cancer antigen 125, cytokeratin 19 fragment, neuron-specific enolase, free human chorionic gonadotrophin, carbohydrate antigen 153, and squamous cell carcinoma antigen. The ascending flaccid paralysis of GBS was suspected. However, the sudden loss of consciousness and absent pulsation of the dorsal

Table 1. Risk factors for aortic dissection and aneurysm [8]

Category	Diseases or factors
Genetic diseases	Marfan syndrome
	Loeys-Dietz syndrome
	Ehlers-Danlos syndrome (vascular form)
	Turner syndrome
	Bicuspid aortic valve
	Familial thoracic aortic aneurysm dissection syndrome
Inflammatory conditions	Takayasu's arteritis
	Giant cell arteritis
	Behçet's disease
Factors related to increased aortic wall stress	Hypertension
	Pheochromocytoma
	Cocaine use
	Coarctation
	Trauma
	Weight-lifting
	Smoking

artery in her feet alerted us to the possibility of aortic dissection. The chest and abdominal CT images from the local hospital were reread and revealed expansion of the aorta with visible dissection involving both the ascending and descending aorta, which is strongly suggestive of aortic dissection (**Figure 1**). After consulting cardiac surgeons, acute aortic dissection was diagnosed. Aortic CT angiography or transesophageal echocardiography was strongly recommended, but the patient's family refused to consider surgical intervention or any palliative treatment. She was discharged the on following day. A follow-up phone call confirmed that she died immediately after discharge.

Discussion

Aortic dissection is a devastating disease, which may present with a variety of clinical manifestations [3]. The DeBakey and Stanford classification schemes are commonly used for aortic dissection [4]. The DeBakey system categorizes dissections based on the origin of the intimal tear and extent of the dissection, whereas the Stanford system divides dissections according to the involvement of the ascending aorta, as shown in **Figure 2**. Based on the CT images and clinical manifestations, the aortic dissection in our case involved both the ascending and descending aortas. Therefore, as per the Stanford system, the dissection was type A. However, the origin of the tear might have been in either the ascending (DeBakey I) or descending (DeBakey III) aorta. The loss of consciousness was probably due to involvement of the aortic arch, which led to transient cerebral ischemia. Similarly, the anomalies of hepatic and renal function likely resulted from organ ischemia.

The diagnosis of GBS is based on the following typical clinical features: prodromal respiratory or gastrointestinal infections, rapid development of muscle paralysis (from onset to nadir < 4 weeks), hyporeflexia/areflexia, and albuminocytological dissociation in the cerebrospinal fluid. The progression of symptoms typically persists over the initial 4 weeks. The most frequent mode of weakness progression is ascending paralysis; the flaccid paralysis usually ascends and may gradually involve the respiratory muscles and cranial nerves. In our case, the progressive weakness of the lower limbs, hypalgesia, respiratory distress, and areflexia strongly suggested ascending paralysis. Given the fact that motor deficits are frequently accompanied by autonomic dysfunction in GBS, potentially lethal dysrhythmias can lead to acute cerebral ischemia and unconsciousness. Note that hypoxia due to respiratory failure in GBS may also cause loss of consciousness. Nevertheless, loss of consciousness is rare in GBS. Therefore, we ruled out GBS. On

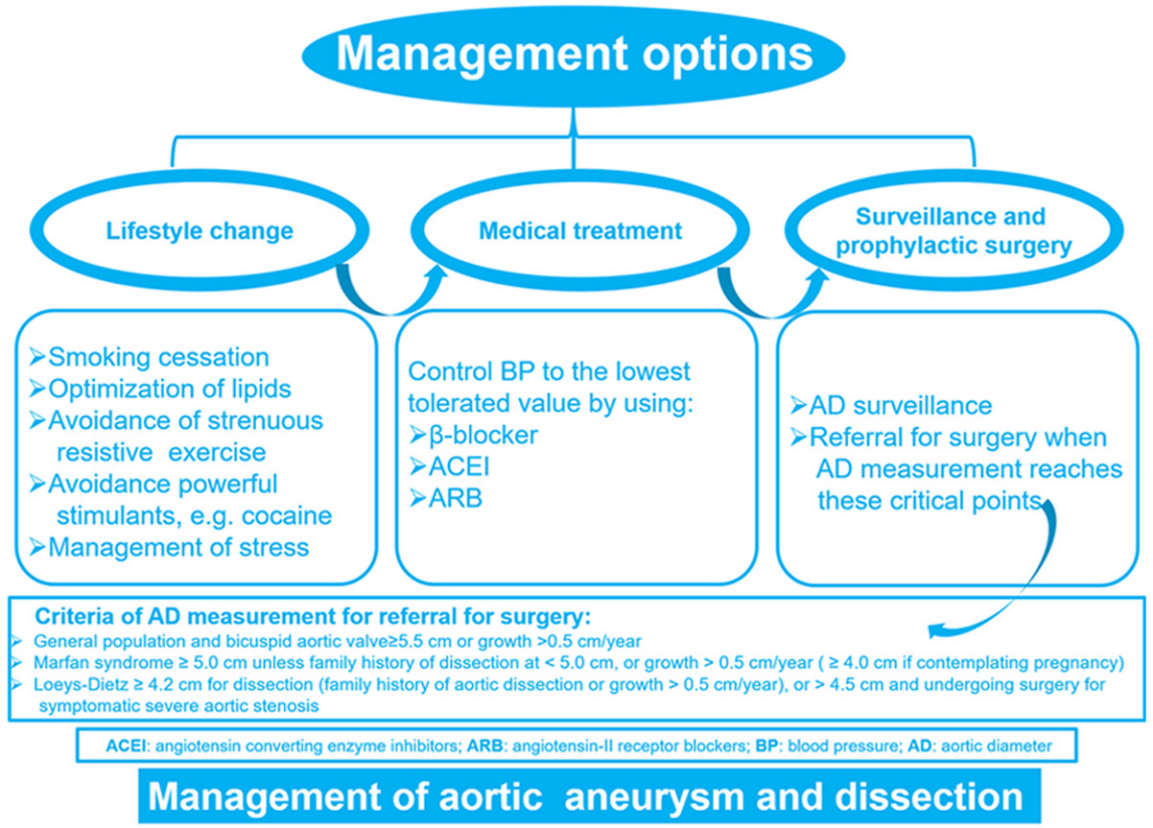

Figure 3. Management options for aortic dissection. The priority in the management of an aortic aneurysm is to control risk factors, so as to slow the expansion rate and reduce the risk of dissection or rupture. Patients should be educated to refrain from smoking, to reduce their lipid intake, to avoid strenuous resistive exercises, such as lifting heavy weights, and to abandon powerful stimulants, such as cocaine. Patients should be cautioned to manage stress as best as they can. Additionally, hypertensive patients should be treated to control hypertension to the lowest tolerated blood pressure. Although controversy exists regarding which patients may benefit from surgery, referral for surgery may be necessary when the aortic dimensions reach a critical point.

admission, the patient could not undergo CT because her posture was restricted by her orthopnea. However, the chest and abdominal CT done at the local hospital showed the expansion of the aorta and false lumen, which were highly indicative of acute aortic dissection.

Aortic dissection is defined as disruption of the media layer of the aorta, with bleeding within and along the wall of the aorta, which results in separation of the aortic layers [5]. Consequently, blood may track along the dissection plane within the media, and the aorta may rupture through the adventitia or back through the intima into the aortic lumen.

The course of aortic dissection is usually fulminant, and its prognosis is bad. Approximately 40% of the patients die immediately, 1% die each hour thereafter, and between 5% and 20% die during or shortly after surgery. Furthermore, depending on age and underlying etiology, only 50-70% survive 5 years after surgery. Consequently, early recognition is of the utmost importance. However, the identification of acute aortic dissection or rupture is often difficult and delayed. Diagnostic errors may account for deaths otherwise attributed to cardiac arrhythmias, myocardial infarction, pulmonary embolism, or mesenteric ischemia [5]. Syncope occurs in approximately 13% of the cases, and mortality is significantly higher in patients with a history of syncope [6]. Interestingly, our patient lost consciousness for 2 days. Although chest or back pain in the presence of an aortic aneurysm is a predictor of aortic rupture [7], our patient had mild back pain.

The common risk factors for aortic dissection are summarized in **Table 1** [8]. The develop-

ment of aortic dissection is associated with genetic diseases with medial degeneration (Marfan syndrome, Ehlers-Danlos syndrome (vascular form), Loeys-Dietz syndrome, Turner syndrome, inflammatory diseases of the aorta, a bicuspid aortic valve, and familial thoracic aortic aneurysm and dissection syndrome), inflammatory conditions (Takayasu's arteritis, Behçet's disease, and giant cell arteritis), conditions that increase wall stress (hypertension, pheochromocytoma, coarctation, cocaine use, and physical trauma), and smoking [8].

It is important to maintain a high level of suspicion for the diagnosis of aortic dissection. Early identification of aortic dissection from diseases with similar presentations is crucial for lowering the mortality and improving the prognosis [5]. Controversy exists regarding which patients may benefit from surgery. The management of aortic dissection by controlling risk factors may slow the expansion rate and reduce the risk of dissection or rupture (**Figure 3**) [5, 8]. Hypertensive patients should be treated to reach the lowest tolerated blood pressure and to optimize lipids [5, 8]. As smokers have twice the aneurysm expansion rate of nonsmokers and higher dissection rates, smokers should be educated to refrain from smoking. Our patient had a history of long-term smoking and identified hypertension, which may act synergistically to promote aneurysm expansion and dissection formation.

In summary, aortic dissection is usually misdiagnosed, and the treatment is delayed. Neurologists should be aware of this devastating disease and its protean manifestations.

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Disclosure of conflict of interest

None.

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