Spontaneous Occlusion of Several Cerebral Venous Sinuses Mimicking Parkinson Disease

Amin Jahanbakhshi, Mehdi Moghaddasi, Alireza Tabibkhooei, Masoumeh Najafi

Skull Base Research Center, Hazrat Rasoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran

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Abstract- Idiopathic occlusion of nearly all cerebral venous sinuses in association with the widespread formation of dural arteriovenous fistulas (AVF) is an extremely rare condition. The cause-and-effect relationship between thrombosis and AVF is not known, but a disturbance in venous flow and distant stagnation has been mentioned as probable pathomechanisms. We introduce a patient that was misdiagnosed as Parkinson's disease and treated accordingly for weeks. Then, rapidly-progressive dementia and shortly after that, an intracerebral hemorrhage occurred, and the diagnosis was established after Magnetic Resonance Imaging and Angiography. There were a whole venous sinus system thrombosis and the formation of numerous dural arteriovenous fistulas. The mechanism and diagnostic nuances are described in this paper, and the treatment options and prognosis are discussed.

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Keywords: Several venous sinus thromboses; Multiple dural arteriovenous fistulas; Parkinson disease

Introduction

Occlusion of several cerebral venous sinuses as a complication of hypercoagulable states such as antiphospholipid syndrome has been reported previously (1). These patients are usually presented with signs and symptoms of increased intracranial pressure (ICP) or hemorrhagic infarcts. Multiple Dural Arteriovenous fistula (AVF) is another rare condition that has been reported a few times and may have an association with multiple dural sinus occlusion (2). The exact pathomechanisms and the cause-and-effect relationship between extensive thrombosis and multiple dural AVF are not known (2). The association with movement disorder is very rare (3). Timing of disease progression has not been reported; however, we found no radiologic sign of thrombosis or dural AVF in MRI performed three years ago for other reasons.

We introduce a rare case of idiopathic thrombosis of the whole venous sinus system and widespread dural AVFs. After a misdiagnosis of Parkinson's disease and dementia, the disease was finally discovered after an intracerebral hemorrhage occurred.

Case Report

Clinical examination

A patient is a 57-year-old man presented with balance and gait disturbance, cognitive problems, and slowness of movement. The symptoms, slowness of movements, and gait disturbance had started insidiously for one month before the first presentation. Clinical diagnosis of Parkinson's disease is made, and oral treatment had been started. Later, rapid deterioration of gait disturbance, automatism, speech difficulty, and finally, a disorder of orientation with a fluctuating nature developed. At presentation, GCS was 11-12. The patient is hospitalized in neurology service with the first impression of Parkinsonism and rapidly-progressive dementia. There is no history of seizure, and other neurological examinations revealed no positive finding.

Imaging and paraclinical findings

Laboratory blood tests for infectious and rheumatologic factors, including antiphospholipid antibodies, coagulation factors, and even prion disease (because of rapidly-progressive dementia), were checked and were normal. Three years ago, he was presented with benign positional vertigo, and an MRI performed that time revealed no abnormality. However, the MR scan performed at the presentation revealed multiple tortuous vasculatures dispersed in whole-brain

Corresponding Author: M. Najafi
Skull Base Research Center, Hazrat Rasoul Akram Hospital, Iran University of Medical Sciences, Tehran, Iran
Tel: +98 21 6650 3890, Fax: +98 21 6650 3890, E-mail address: masy.najafi@gmail.com

(Figure 1). However, these findings were not followed by an angiography until a rapid deterioration of consciousness occurred, which was caused by a spontaneous intracerebral hemorrhage. At this time, neurosurgery consultation was requested, and an emergent craniotomy was performed (Figure 2). Intraoperatively huge bleeding veins were abundant, and hemostasis was achieved hardly using vascular clipping and cottonoid buttress. A few days later, the cottonoids were removed, and the patient underwent an angiography. Obliteration of nearly all cerebral venous sinus was seen, and multiple extracranial bypass veins were evident. In addition, early filling of some extracranial veins proved the presence of Arteriovenous (AV) fistula (Figure 3). After consultation with an endovascular interventionist, the obliteration of these AV fistulae was tried. The most important feeders were bilateral occipital and superficial temporal arteries that were obliterated in two sessions (Figure 4).

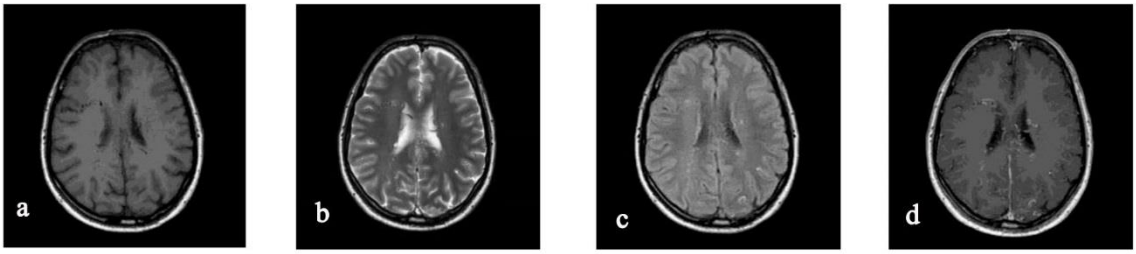

Figure 1. Axial T1-weighted (a), T2-weighted (b), FLAIR (c) and Gadolinium-enhanced (d) MRI showing the abnormal tortuous veins

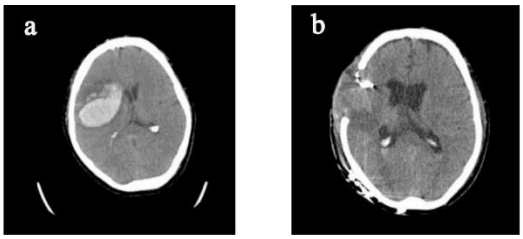

Figure 2. Intracerebral Hemorrhage treated with craniotomy and multiple clipping

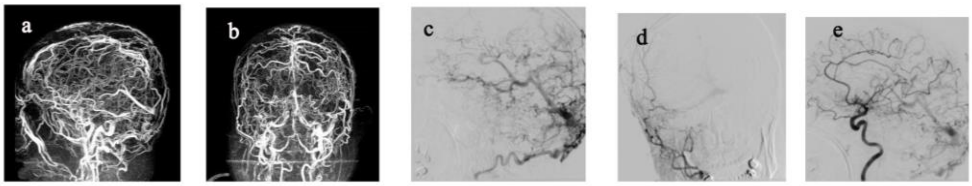

Figure 3. Magnetic resonance venography (a,b) and digital subtracted angiography (c,d,e) showing the absence of normal venous sinuses and several AVF

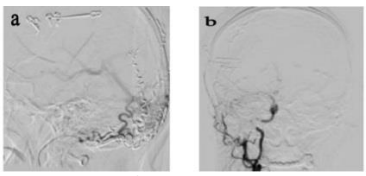

Figure 4. Digital subtracted angiography after endovascular obliteration of the venous fistula

A few days later, the patient developed hydrocephalus and underwent a ventriculoperitoneal shunt procedure; however, after shunt placement, he developed subdural hematoma that was evacuated emergently. The patient was stable thereafter, and GCS was fluctuating around 8 to 10. Unfortunately, two

months after admission, a severe sepsis occurred that led to his death.

Discussion

The mechanisms that underlie the thrombosis of several venous sinuses and the formation of several AV fistula are not completely known. Either of these two conditions may be the cause or the effect of the other(2). Congenital, infectious, traumatic, and autoimmune and hypercoagulopathic causes have been described elsewhere (4, 5). Extensive and simultaneous occlusion of the venous sinuses has been reported in 71% of cases with multiple AV fistulas (6). Inflammatory pathomechanisms may be involved that cause the release of angiogenic factors (4). Therefore, venous sinus thrombosis triggers the inflammatory responses leading to the formation of AV fistula. On the other hand, AV fistula causes stagnation and turbulence of venous flow distant to the venous sinuses and pave the road for several venous sinus thromboses. Chronology and order are not well understood; however, in the current case. There was no clue to a venous abnormality in the MRI performed 3 years ago, showing that this time is the maximum time needed for the process to be completed.

In patients with multiple dural sinus fistula, the occurrence of cortical venous reflux is common, and this may lead to cerebral ischemia or intracranial hemorrhage (7). Moreover, in multiple AV fistula, venous hypertension in the deep venous system occurs more frequently (8), and this may underlie the movement disorders seen in the current patient.

Clinical presentation of multiple AV fistula is usually more aggressive than single AV fistula. Rapidly progressive venous hypertensive encephalopathy may cause hemorrhage, seizure, or neurological deficits. The rapid progression and extensive brain involvement differentiate it from simple AVF and cause a general decline in brain function and may cause memory loss, behavioral changes, and dementia (9, 10). That is why the current patient was misdiagnosed as Parkinson's disease and had taken anti-Parkinson disease medication for several weeks.

The diagnosis of multiple AV fistula is suggested by initial imaging. Non-contrast brain CT scans may not help in uncomplicated cases but sometimes may show blood stasis. However, MRI may give several clues. Signal abnormalities and hyperintense areas (leukoaraiosis) in white matter and multiple serpentine veins with fluid voids may be seen (11) (Figure 1). MRI can be used for monitoring of progression or improvement of venous hypertension (12). Digital Subtracted angiography (DSA) is considered as a gold standard of diagnosis. But, because of the complex dynamics of venous circulation in multiple AV fistula, visualization of all AVFs may be challenging (2).

Due to the extensive involvement of the brain and marked abnormality in the venous circulation, treatment is essential. Obliteration of fistula can be achieved either by endovascular technique, microsurgically, or by stereotactic radiosurgery. Obliteration is usually performed in a staged manner, and the fistulas with cortical reflux with a higher Borden/Cognard classification have priority (2). In the current case, a combination of techniques was used. Because of intracerebral hemorrhage, we obliterated some venous channels with multiple clips. Then endovascular obliteration was performed in two stages. If hydrocephalus happened, it should be noted that shunting in such patients may be complicated by hemorrhage either by making injury to several veins in the passage of ventricular catheter or reduction of ICP that may cause changes in the venous circulation. The prognosis is usually poor, and the treatments are usually not able to stop the progression of the disease (13, 14). However, improvement of symptoms, including dementia with treatment, has also been reported (11).

This report introduces a case of multiple dural sinus thrombosis and multiple dural arteriovenous fistulae that was presented with a movement disorder and misdiagnosed to have Parkinson's disease and later, while he was evaluated for rapidly progressive dementia, intracerebral hemorrhage occurred. It can be highlighted the importance of high suspicion for venous disease in unusually presented patients. Although the treatment may not stop the progression of disease in extensive cases, it can postpone some complications such as ICH.

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