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Case Report

Isolated Cortical Vein Thrombosis with Long Cord Sign

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Isolated cortical vein thrombosis (ICVT) is a rare disease, accounting for less than 1% of strokes. A 46-year-old woman presented with progressive left side weakness. Magnetic resonance (MR) imaging with T2*-gradient echo (T2*-GE) sequence showed long cord sign at the right frontal cortex. The patient was treated with low molecular weight heparin, followed by oral warfarin for 6 months. The 3-month follow-up MR imaging showed recanalization of the previously thrombosed cortical vein. She was completely recovered without neurological deficits after 6 months. This provides that MR imaging with T2*-GE sequence can help to diagnosis the ICVT and outcomes of the ICVT are generally favorable.

Key Words : Cortical vein thrombosis · Cord sign · Hemorrhagic infarction.

INTRODUCTION

An isolated cortical vein thrombosis (ICVT) causing a venous hemorrhagic infarction without involving venous sinuses or deep cerebral veins is rare and accounts for less than 1% of strokes[8]. An ICVT is more difficult to diagnose than a cerebral venous thrombosis because the symptoms are non-specific and there is high variability in the number, size, and localization of the cortical vein[2,11,12]. Recently, T2*-gradient echo (T2*-GE) sequence magnetic resonance (MR) imaging has high sensitivity to all paramagnetic hemoglobin and shows linear hypointensity because of the magnetic susceptibility effect (MSE) at the site of the venous thrombosis[2]. Therefore, it is particularly useful for diagnosing an ICVT by detecting a thrombosed cortical vein−the cord sign and typically described long cord signs are rarely reported[1,6,10,12].

In this case report, a patient presented with an ICVT in the right frontal lobe that was diagnosed by the long cord sign on the T2*-GE MR imaging.

CASE REPORT

A 46-year-old female visited the emergency department because of progressively worsening left side motor weakness and hypoesthesia. Patient denied any history of trauma, infection or hypercoagulable disorders. The only medication she reported using was oral contraceptives. Neurological examination revealed alert mentality, stable vital signs, strength of 2/5 in the left extremities, and mild hypoesthesia in the left limbs. There was suspicion of stroke, and MR examinations including T2*-GE and MR venography (MRV) were performed. Fluid-attenuated inversion recovery (FLAIR) images demonstrated increased signal intensity in the right frontoparietal lobe (Fig. 1A). There was no definite intracranial vascular abnormality on MR angiography (MRA). MRV showed patency of all the major venous sinuses and deep cerebral veins (Fig. 1B). T2*-GE images showed linear hypointensity in the right posterior and the central sulcus because of the thrombosed cortical vein (Fig. 1C). Laboratory examinations, including the coagulation factors, were normal. These findings suggested acute ICVT. The patient was treated conservatively without an anticoagulant. After 1 day of admission, the patient presented worsening of left motor weakness (strength of 1/5) and displayed focal involuntary movement in the left lower extremity. Follow-up MRI was performed, and demonstrated hemorrhagic conversion in the previous lesion. Digital subtraction angiography (DSA) was performed to identify other vascular lesions that could possibly have caused the hemorrhage. DSA images showed a defect of the cortical venous structure in the junction of the right frontoparietal area compared with the left side, without other vascular malformations (Fig. 2A, B). The patient was treated with antiepileptic (levetiracetam) and anticoagulation (low molecular

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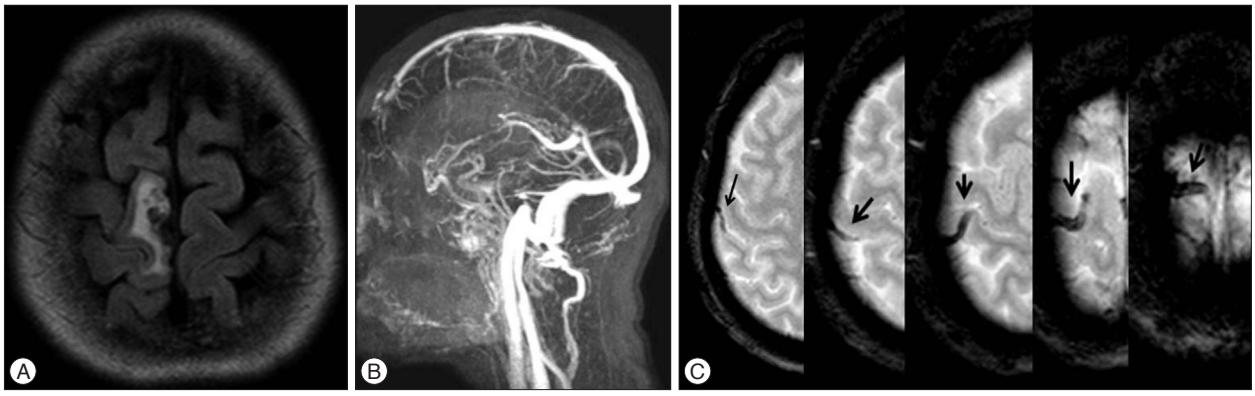

Fig. 1. A : Fluid-attenuated inversion recovery image demonstrating the initial presenting venous infarction. B : Magnetic resonance venography showed the patency of the major venous sinuses and deep cerebral veins. C : On T2*-gradient echo sequence image, long linear hypointensity along the right central and paracentral sulci (arrows) was observed–the long cord sign.

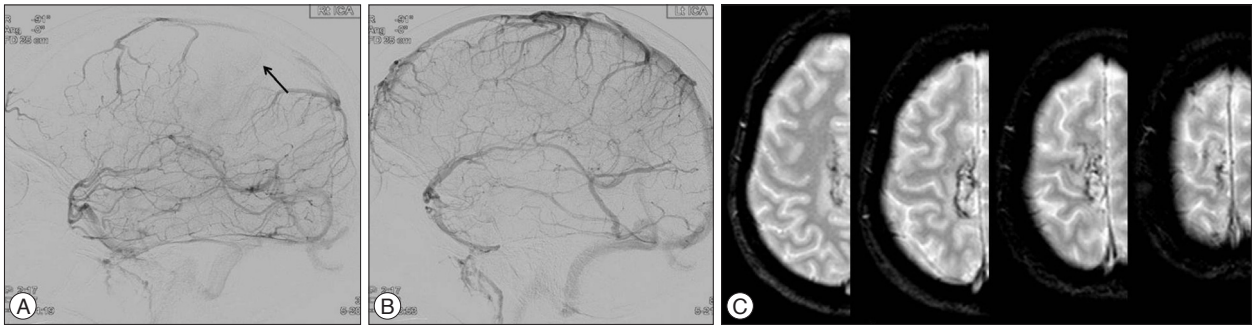

Fig. 2. A : DSA of the right side showed a defect of the cortical vein (arrow) as compared with the left side. B : No definite abnormality on DSA of the left side. C : The 3-month follow-up T2*-gradient echo showed recanalization of the thrombosed cortical vein. DSA : digital subtraction angiography.

weight heparin, LMWH) medications. After 1 week, the anticoagulant was changed from LMWH to oral warfarin. Motor weakness in the left limbs gradually improved. The patient was able to walk with a cane at discharge. At 3 month follow-up, T2*-GE imaging showed resolution of the hemorrhagic venous infarction and recanalization of the thrombosed cortical vein (Fig. 2C). The patient completely recovered and discontinued the medications after 6 months.

DISCUSSION

There are a limited number of case studies and case series describing ICVT, and its incidence and etiology are unknown. Recently, Coutinho et al.[3] published a systematic review of ICVT case reports and case series. The authors found reports of the following underlying conditions : use of oral contraceptives or steroids, infection, malignancy, postpartum status or pregnancy, intracranial hypotension after a lumbar puncture, vasculitis, inflammatory systemic disease, and genetic or acquired thrombophilia[4-7]. Boukobza et al.[2] reported the incidence of ICVT was 6.3% of all cerebral venous thrombosis (CVT) cases. However, ICVT might frequently be overlooked for the following reasons : 1) non-specific symptoms from ICVT involvement in the silent cortical vein, 2) rapid compensation by collateral vessels and recanalization, and 3) thrombosed small veins at the cortical level that are difficult to detect using routine MR imaging.

Diagnosis of ICVT is based on the identification of cortical vein thrombosis without venous sinus and deep vein involvement. The majority of ICVT occurrences are diagnosed by neuroradiological modalities such as CT, MR images, and DSA. CT scans can directly detect ICVT, which is shown as a hyperintensive structure. However, indirect signs such as sulci infarction or hemorrhage are more frequently observed[2,9]. MRI is the most appropriate diagnostic modality for investigating ICVT and can directly detect cortical thrombus. MRI can also identify secondary changes of cortical venous congestion around the involved vein, including swollen gyri and hemorrhagic conversion. Additionally, MRV is able to identify patency of dural sinuses, and can show defects of the large cortical vein resulting from cortical vein thrombosis relative to the non-involved side[2,6]. Cord sign is shown with high intensity on the T1 weighted image, low signal intensity on the T2*-GE sequence, and high signal intensity on the FLAIR image. It is the most important diagnostic sign of ICVT. However, cord sign is rare. The long cord sign observed in our case is particularly rare. T2*-GE sequence improves detection of ICVT because of susceptibility effects of the deoxyhemoglobin in the thrombosed cortical vein[2,6,9]. ICVT diagnosis could be missed when routine MRI and MRV are performed because the cortical vein is variable with respect to number, size, and localization. Thus, cord sign is

rarely observed in those sequences. Therefore, a T2*-GE sequence must be included when ICVT is suspected. In our case, the cord sign was not observed on FLAIR, T1, and T2 sequences. However, it was observed on the T2*-GE sequence. When MR series fail to identify the ICVT, DSA is occasionally required to confirm vascular abnormality. Vascular abnormality may include the following : delayed local venous drainage, defect of the cortical vein compared to the normal side, collateral vessels, and intact patency of the sinuses.

There is no consensus regarding the management of ICVT. However, anticoagulation with therapeutic dose of heparin or low molecular weight heparin is recommended because it is known to be moderately more beneficial than placebo[1,2,9,12]. In some reports, ICVT patients completely recovered with conservative care without anticoagulation[6,12]. There is no consensus regarding duration of oral anticoagulant in ICVT patients. Duration of oral anticoagulant therapy in cases of ICVT depends on whether there is a complete resolution of the thrombus on follow-up images. Oral anticoagulant therapy is typically recommended for 3−6 months in patients with recurrent symptoms and genetic or acquired thrombophilia[2,8,9,12]. In our case, the 3-month follow-up T2*-GE sequence showed recanalization of the thrombosed cortical vein. However, we stopped the anticoagulant after 6 months because the patient's symptoms were persistent.

There are no prospective or control studies on ICVT. Use of anticoagulation, the existence of a parenchymal hemorrhage or infarction, and length of the thrombosed cortical vein do not significantly affect the clinical outcomes in the reported case series[1,2,6,9,12]. The majority of patients with ICVT have acceptable recovery.

CONCLUSION

ICVT might be overlooked because its symptoms are non-specific, and the diagnostic modalities for detecting a thrombus in the cortical vein are limited. We report the case of a patient with ICVT and long cord sign on the T2*-GE sequence. Our case demonstrates that the T2*-GE sequence is useful for detecting ICVT, and length of the involved vein in a patient with ICVT is not relevant to prognosis.

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