Thrombosis of the Vein of Galen: A Case Report

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Abstract- Thrombosis of the deep cerebral venous system is a rare entity. We report a case of 68-year-old male presenting with headache, left side weakness, and alteration of consciousness at admission. He was diagnosed as deep cerebral venous thrombosis (DCVT) of the vein of Galen on the basis of brain magnetic resonance imaging (MRI) findings. Anticoagulant therapy with heparin was constituted and he made a good recovery. DCVT often involves bilateral thalami and sometimes basal ganglia with obscure clinical manifestations. Therefore, early diagnosis may not be always straightforward. Brain MRI findings with unique “circular” region of signal change in bilateral thalami and basal ganglia can be very helpful for a proper diagnosis. Although early anticoagulation therapy is usually recommended, the optimal treatment of this rare disease needs further investigation.

Key Words: Cerebral deep venous thrombosis, Vein of Galen, Anticoagulant therapy

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INTRODUCTION

Cerebral venous thrombosis is an uncommon disease. The most frequent thrombotic sites are the lateral, cavernous, and superior sagittal sinuses. On the other hand, thrombosis of the deep venous system, such as internal cerebral veins, vein of Galen and straight sinus occurs far rarer with a worse prognosis¹⁻³. As non-specific clinical features make early diagnosis of DCVT difficult, the application of brain MRI is helpful. Once DCVT is confirmed, anticoagulant therapy could be started to prevent further thromboembolism¹⁻⁵.

CASE REPORT

A 68-year-old male suffered from acute left hemiparesis and alteration of consciousness. He soon went to our emergency service on the same day. He had had new onset of headache for 3 days before going to the emergency service. He denied any preexisting systemic disease. The neurological examination revealed a drowsy state of consciousness with intact cranial nerves. There were also hemiparesis, relative hyperreflexia, and extensor-type plantar response on the left side. The other physical findings were unremarkable. Laboratory find-
ings, including complete blood count, biochemical profiles, and autoimmune as well as tumor markers were all within normal limits, except for the elevated fibrinogen (511 ug/L; normal < 375). Emergent non contrast-enhanced brain computed tomography (CT) revealed non-specific, ill-demarcated hypodense areas in bilateral thalami and basal ganglia (Fig. A). He was admitted at the same day under the diagnosis of cerebrovascular disease and was treated with oral antiplatelet agent (aspirin) initially. Brain MRI was performed 2 days after admission (5 days after onset of symptoms). It showed “circular” hyperintensity on axial T2-weighted imaging (T2WI) in bilateral thalami and basal ganglia (Fig. B) and a hyperintense signal ascribable to the thrombosed vein of Galen on T1-weighted imaging (T1WI) (Fig. C). Besides, diffusion-weighted imaging (DWI) showed hypointensity in the right basal ganglion, suggesting hemorrhagic transformation (Fig. D). Thrombosis of the cerebral deep venous system was highly suspected. Finally, the appearance of thrombosis of vein of Galen on the three-dimension time of flight magnetic resonance angiography (3D TOF MRA) (Fig. E) confirmed the diagnosis. Heparin was administered immediately for 3 days and then was shifted to oral warfarin. He recovered well from headache, alteration of consciousness and left hemiparesis in a week. After 1-year of oral warfarin treatment, follow-up brain MRI revealed resolution of thrombosis of vein of Galen on 3D TOF MRA (Fig. F).

**DISCUSSION**

There are a number of causes that lead to cerebral
venous thrombosis\(^1\-\(^2\), including hypercoagulative state, oral contraceptives, pregnancy\(^4\), malignancy\(^5\), dehydration and infection. The only risk factor found in this reported case was the elevated fibrinogen level.

Deep cerebral veins drain the deep white matter, thalami and basal ganglia into bilateral internal cerebral veins, which start behind the foramina of Monro and are located in the roof of the third ventricle near midline. The internal cerebral veins run posteriorly and then unite with each other and the basal vein of Rosenthal to form the vein of Galen. The vein of Galen is a short structure passing under the splenium of the corpus callosum. It curves around the splenium and joins the inferior sagittal sinus to form the straight sinus. Isolated DCVT\(^6\), which means thrombosis of deep cerebral veins without concomitant superior sagittal sinus involvement, typically makes venous infarction in bilateral thalami and sometimes basal ganglia with brain edema\(^3\-\(^8\). However, DCVT with unilateral venous infarction has been reported\(^8\). Also, the venous infarction can be associated with hemorrhagic transformation as shown in this case, where hemorrhagic transformation in the right basal ganglia with brain edema\(^3\-\(^8\). However, DCVT with unilateral venous infarction has been reported\(^8\). Moreover, DCVT with unilateral venous infarction has been reported\(^8\).

Because of the relative insensitivity of non contrast-enhanced brain CT to infarction, the radiological diagnosis of DCVT may be missed if no other neuroimage studies are done. However, contrast-enhanced CT may not confirm the diagnosis of DCVT either, if the thrombus is too small to be visualized as the “empty delta” sign\(^9\). In this case report, brain CT showed a subtle, ill-defined low density in bilateral thalami and basal ganglia. We should be alert for the possibility of DCVT when there are imaging findings of this kind in brain CT. Although cerebral angiography is considered as a definitive diagnostic test for DCVT, many recent reports have also demonstrated the value of brain MRI\(^1\-\(^3\). In this case, brain MRI revealed “circular” hyperintensity in bilateral thalami and basal ganglia in T2WI. This pattern of lesion distribution can not be totally explained by the involvement of one single arterial territory. On the other hand, the possibility of deep venous territory infarction should be taken into consideration. In this regard it should be noted that the infarction area may be only confined to bilateral thalami in some cases of DCVT and survey of top of basilar artery occlusion is needed under such circumstances\(^13\). We finally established the diagnosis of venous infarction due to occlusion of vein of Galen by the imaging findings of 3D TOF MRA\(^1\-\(^3\), in which the thrombosed vein of Galen can be evidently visualized. Magnetic resonance venography (MRV) is considered as another helpful diagnostic tool and is often indicated at very early (before day 5) or late (after 6 weeks) stages when routine MRI shows equivocal signs or false-negative findings\(^11\). For this reported case, we did not arrange MRV because at first the obscure clinical findings were not specific for cerebral venous thrombosis, and because MRI performed 5 days after onset of symptoms and showed confirmatory imaging findings.

Whether there is hemorrhage or not, anticoagulation with heparin is the mainstay of acute-phase treatment for DCVT\(^1\-\(^3\). Heparin may arrest the thrombotic process and was reported to reduce the mortality rate\(^1\). Heparin is usually followed by oral anticoagulation for 3-6 months or longer, with target international normalized ratio of 2.5, to prevent recurrent thrombosis if there is evidence of coagulopathy\(^2\-\(^3\). The hypointensity in the right basal ganglion in DWI suggested hemorrhagic transformation in our case. He did recover well with anticoagulant therapy. After 1-year treatment of oral warfarin, the follow-up MRI revealed resolution of the thrombosed vein of Galen in 3D TOF MRA. Combined therapy with local thrombolysis might be useful for DCVT in view of the chance of restoring flow more rapidly than heparin alone\(^1\-\(^1\), but there is no evidence that the clinical outcome is better because of the greater risk of hemorrhage\(^5\). Further clinical trials are needed to establish the optimal treatment for DCVT, an infrequent but potentially serious or even fatal condition.
In summary, DCVT is such a rare, potentially fatal disease that early diagnosis is essential for timely treatment. However, early diagnosis is usually difficult due to its non-specific clinical manifestations and obscure imaging findings in brain CT. When there are infarcts with edema involving bilateral thalami or basal ganglia, one should have DCVT in the list of differential diagnosis. The pattern of brain MRI findings is an uncommon one, but is distinctive for DCVT. This case report reminds neurologists that brain MRI could be as important as neurological examination in the diagnosis of many central nervous system diseases. The unique imaging findings of DCVT therefore should be kept in mind because the prognosis could be very different with or without an appropriate treatment in time.

REFERENCES