Cerebral Dural Sinus Thrombosis
- Case Report -

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Abstract

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- 증례보고-

Cerebral dural sinus thrombosis (CDST) has been described as a rare disease with a variety of patho- etiological factors. The diagnosis of CDST is difficult due to various symptoms and signs, none of which is specific to CDST. But timely diagnosis is critical for effective management. The introduction and widespread use of computed tomography (CT), magnetic resonance imaging (MRI) and cerebral angiography made early diagnosis of CDST possible. In particular, MR venography is the most useful tool for establishing a correct diagnosis quickly. In early literature, mortality ranked between 30% and 50% but in more recent series it is between 5.5% and 30%. With the advent of diagnostic and therapeutic tools, early diagnosis and proper management has made the prognosis better. The appropriate therapy for CDST, however, has been the subject of much controversy. Individual variations of the venous system and collateral vessels are key factors to decide the proper treatment. In this report, we present two cases with symptomatic CDST treated without open surgical or direct endovascular interventions with good outcome.

KEY WORDS Cerebral dural sinus thrombosis, MR venography, Collateral vessels.

Introduction

Cerebral dural sinus thrombosis (CDST) has been described as an uncommon cause of ischemic stroke. It can often be clinically difficult to diagnose. The incidence of CDST is still somewhat unknown. The initial symptoms and signs are often vague. But the introduction and widespread use of CT, MRI and cerebral angiography made early diagnosis of CDST possible. But the appropriate therapy has not been established. Individual variations of the venous system and collateral vessels are key factors to decide the proper treatment. In this report, we describe two symptomatic patients with CDST who was successfully treated without open surgical or direct endovascular interventions.

Case Reports

1. Patient 1
A 43-year-old right handed woman had a 3-day history of headache and was admitted to the neurosurgical department when she developed symptoms of mild anomic aphasia. A CT scan of the brain demonstrated a hemorrhagic infarct in the left temporal lobe, which was suggestive of cerebral dural sinus thrombosis (Fig. 1A). The results of clotting screening tests, including protein C and antithrombin III, were normal. MRI revealed thrombosis of the
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left transverse and sigmoid sinuses in addition to the Remorrhagic infact(Fig. 1B). Cerebral angiography was performed. It demonstrated thrombosis of the left transverse and sigmoid sinuses, extending into the proximal internal jugular vein(Fig. 1C). Heparin was not administered because of the recent occurrence of intra-cerebral hemorrhage.

Fig. 1. A] Computed tomographic scan of the brain demonstrating a large hemorrhagic infarct in the left temporal lobe. B] Axial T1-weighted MR image revealing thrombus in the entire left transverse sinus as high signal intensity(arrows) and intraparen-chymal hematoma in the left temporal lobe(arrow heads). C] Digital subtraction angiogram of the left internal carotid artery, anteroposterior view, venous phase, showing occlusion of the left transverse and sigmoid sinuses(arrow). D] Follow-up angiography obtained 3 weeks after ictus, showing partial recanalization of the transverse sinus(arrows) and occlusion of the sigmoid sinus(arrow heads).
as well as the good neurologic status of the patient. Therapy was directed at control of intracranial pressure (ICP) and maintenance of adequate hydration. On day 5, repeated neurological examinations showed that she was fully awake and conscious; however, a moderate language disturbance persisted. The patient improved progressively and repeated MRI investigations revealed no further bleeding or infarction. On day 10, intravenous administration

![Fig. 2. A] Contrast enhanced CT scan of the brain showing a hemorrhagic infarct in the right occipital lobe with an empty delta sign (arrow). B] T1-weighted sagittal MR image revealing high signal intensity (arrow) in the posterior portion of the superior sagittal sinus, indicating thrombosis and also shows thrombosed veins (arrow heads). C] MR venogram, sagittal view. No flow signal in the posterior portion of the superior sagittal sinus is seen, indicating thrombosis (arrow). D] Anteroposterior digital subtraction venogram showing partial obstruction of the superior sagittal sinus (arrows) and a few anastomotic meningeal veins (arrow heads).]
of mannitol was discontinued. But she was continued on seizure prophylaxis with valproate. On day 20, the patient became neurologically intact. Repeated cerebral angiography showed an occlusion of the distal transverse and sigmoid sinus but slow filling of the proximal transverse sinus (Fig. 1D). After 1 month, the patient had returned to her premorbid condition without a language disturbance.

2. Patient 2

A 73-year-old right-handed woman, who was receiving anti-hypertensive medications, was admitted to the neurosurgical department because of a headache for 7 days. A neurological examination showed that she was fully awake. But she complained of a headache and visual disturbance. A CT scan of the brain demonstrated a left occipital hemorrhagic infarct with an empty delta sign in superior sagittal sinus, which was suggestive of CDST (Fig. 2A). MRI investigations revealed thrombosis of the superior sagittal sinus and the torcular Herophili (Fig. 2B and 2C). Studies on clotting parameters and other associated factors revealed no definitive abnormalities. The therapy was directed at reducing ICP, maintenance of adequate hydration and maintenance of a normal arterial blood pressure. No additional thrombolytic anti-coagulant therapy was performed because of the patient’s refusal. The next day, cerebral angiography was performed. It demonstrated a partially obstructed superior sagittal sinus. It also showed collateral pathways from the superior sagittal sinus to the left transverse and the sigmoid sinus via anastomotic meningeal veins (Fig. 2D). The patient improved progressively and 1 month later, discharged from the hospital. At that time, she was fully independent, despite left homonymous hemianopia.

Discussion

A wide variety of pathological conditions may cause or predispose to CDST (26,8,9). The symptoms and signs are also variable ranging from nonspecific symptoms of headache, focal neurologic deficit, to more severe manifestations such as coma. The incidence of CDST is still unknown because it goes unrecognized in many patients. It has been reported as a rare disease in most autopsy series. Towbin (8) reported CDST in 9% of 182 consecutive autopsies. Averback (1) insists in his report that CDST in young adults is an under-recognized disease. But Preter et al. (7) suggested that CDST was not uncommon. With the advent of diagnostic tools, the detection rate is being increased. CDST is frequently seen in association with infection, trauma, connective tissue diseases such as Behcet’s disease, cancer, cardiac disease, diabetes mellitus, inflammatory bowel disease, hormonal changes (puerperium, use of oral contraceptives) and dehydration (26-28). But in about one-quarter of cases, no causes could be found (27) as in our two cases. In our cases, the etiology of CDST was not established. The neurologic symptoms, signs and their mode of onset are extremely variable. Preter et al. (7) described 6 main symptoms and signs, those included such symptoms and signs as headache, papilledema, hemiplegia, seizure, confusion and dysphasia. The most frequent symptom and sign was headache (74%) and papilledema (45%). Headache is much more frequent symptom in CDST than in arterial cerebral ischemia. The anatomical basis for headache may be the presence of densely populated nerve terminals in the connective tissue in close contact to the sinuses (9). Clinical manifestations of a patients with an arterial thrombotic disease usually develop suddenly, which suggests that it is a single event, monophasic process. But the patient with CDST often shows a gradual increase of symptoms or a fluctuating course. The fluctuating symptoms imply that CDST is a continuing process (9). In our cases, the symptoms of the patients developed and progressed gradually. The radiologic diagnostic criteria of CDST, including the delta sign on a contrast enhanced CT, the signal characteristics of thrombus and lack of flow within the sinus on MRI, have been well described (9,10). But CT will demonstrate only secondary effects of CDST. The thrombus within the sinus usually was not detected on a CT scan. MRI is more sensitive in demonstrating the thrombi and the parenchymal changes. In our cases, MRI demonstrated thrombi in the transverse and sigmoid sinus in patient 1 and in the superior sagittal sinus in patient 2. Yuh et al. (10) described MR findings of CDST and grouped those findings into three patterns according to the signal changes on T2-weighted image and the presence of hematoma. In their report, the frequent observation of brain swelling without associated vasogenic edema (high signal on T2-weighted images) suggests an absence of blood-brain barrier breakdown in the areas affected by CDST. They explained that the abnormal signal on T2-weighted images demonstrated the transudate in the interstitial space which was made by
the increased shift of bulk water. Thus, the high signal intensity on T2WI should not always be considered as an infarction. It could represent interstitial edema—a reversible process. Those findings are characteristic on MRI in patients with CDST. In arterial thrombotic disease, a high signal intensity on T2WI usually represents fixed infarctions. Hemorrhage in arterial cerebral ischemia usually occurs in the reperfusion period especially when distal migration of the embolic fragment from the initial site occurs. But hemorrhage in CDST is due to the increased venous and capillary pressure. In arterial ischemic thrombotic disease, ischemic damages to the vessels and neurons probably precedes the hemorrhage. In CDST, the increased capillary pressure leads to blood leakage but only later to damage of the other components of the tissue including neurons. Those findings explain well that the great reversibility of the patients with CDST even when the patient is in the worst neurological conditions. In the patients with CDST, cerebral blood flow as measured by Xenon single photon emission computed tomography (SPECT) is decreased but the blood flow can be increased considerably after stimulation with acetazolamide. In contrast, in arterial cerebral ischemia, the reduced blood flow in the ischemic region cannot be increased after acetazolamide stimulation. The cerebral blood flow (CBF) changes indicate that despite a decrease in blood flow due to venous occlusion, on the arterial side there is no maximal dilation to overcome the reduction of CBF. In the report presented by Villiringer et al., normal or slightly decreased cerebrovascular reserve capacity in CDST makes the prognosis better than in arterial stroke. In recent years, the mortality has decreased from a range of 30 to 50% to a range of 5.5 to 30%. Preter et al. reported good outcomes without permanent neurological deficits in 86% of their patients with CDST. Several therapeutic options are available. The choice of the treatment depends on the severity of the clinical symptoms, the location of thrombi, the extent of thrombi and the presence of collateral vessels. The patients with mild symptoms and normal imaging studies of the brain parenchyma can be managed with systemic anticoagulation, ICP management and adequate hydration. But individual variations of the collateral vessels should be considered. In our cases, the patient 2 had a few anastomotic meningeal veins from the superior sagittal sinus to the transverse-sigmoid sinus. The presence of collateral vessels are very important factor to decide the treatment of choice. Even in the patients with hemorrhagic transformations of the moderate-sized infarcted area, the patient with abundant collateral vessels could achieve good outcomes with the medical treatment alone. In patient 1, repeated cerebral angiography revealed partial recanalization of the affected vessels. The only partial recanalization of the occluded dural sinus is sometimes necessary to obtain a desirable clinical outcome. Preventing further propagation of thrombosis and improving the blood rheology are essential to the treatment. In arterial thrombotic disease, hemorrhage is due to reperfusion and so improvement of blood flow by heparin increases the risk of hemorrhage. Bleeding in CDST is due to the increased venous pressure. Therefore heparin induced improvement of outflow decreases the risk of hemorrhage. When the patient showed rapid neurological deterioration with extensive thrombosis, direct thrombolytic therapy should be considered. Several reports have shown reasonable safety and efficacy of direct endovascular thrombolytic therapy with infusion of urokinase or tissue Plasminogen Activator (tPA). Several treatment options are available but, because of the characteristics of this disease, the proper treatment must be tailored to the needs of the individual patient. As in our cases, recanalizations of the occluded dural sinus, variations of the venous system and the opening of collateral veins are important factors to decide the treatment. Although the size of hemorrhagic infarction was considerable, the patients showed good recoveries with or without minor sequelae.

**Conclusion**

These cases document good outcomes in patients with CDST treated medically. Correct and prompt diagnosis relies on a high index of suspicion, particularly in those patients with vague symptoms. Because the prognosis of the patients with CDST is better than those reported in the early literature, the patients with CDST should be treated aggressively. And the treatment should be tailored to the needs of the individual patient.

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References

5) Kim SY, Suh JH[ Direct endovascular thrombolytic therapy for dural sinus thrombosis[ Infusion of Alteplase. AJNR 18[ ] 639-645, 1997

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— 국문 조록 —

뇌경막 정맥동 혈전증 및 비교 연구

뇌경막 정맥동 혈전증의 임상적 특성과 치료

중심 단어: 뇌경막 정맥동 혈전증, 치료방법, 임상적 특성.