CASE REPORT

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Postpartum Cerebral Angiopathy - Presenting with Both Hemorrhagic and Ischemic Stroke

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Postpartum cerebral angiopathy(PPCA) is a benign and, reversible cerebrovascular disease in the postpartum period. The authors report here a 29-year-old woman who had severe headache, very poor consciousness and visual disturbances as the symptoms of PPCA. The clinical and radiological evaluation and treatment of this patient are disscussed.

KEY WORDS : Postpartum · Stroke.

Introduction

Stroke is an uncommon disease in young woman, but it is believed that during pregnancy and the postpartum period, the risk of stroke is increased[5,7,8,12]. The overall adjusted relative risk of stroke is 2.4 per 100,000 pregnancies(95percent confidence range, 1.6 to 3.6) and attributable risk is 8.1 strokes per 100,000 pregnancies(95percent confidence range, 6.4 to 9.7) in the United States[5]. Stroke is the 5th. leading cause of maternal death and it account for about 10% of all maternal deaths in North America[2]. Postpartum cerebral angiopathy (PPCA) is one of the rare clinical entities of stroke. It has a relatively benign clinical course that presents with headache, nausea/vomiting, blurred vision or scotoma, seizure, a decreased consciousness level, and focal neurological deficit in the postpartum period[9,13]. In this study, the authors present a case of PPCA that presented with both hemorrhagic and ischemic stroke, and we have also conducted a review of the relevant medical literatures.

Case Report

A 29-year-old woman was presented to the emergency room with the chief complaint of severe headache and progressive deterioration of her consciousness level that occurred 5days after the normal spontaneous vaginal delivery of her second baby. She'd had an uneventful course of pregnancy. She had no signs of infection or deep vein thrombosis, and there was no significant past medical history, especially regarding the hypertension or eclampsia. On admission, she was normotensive without edema which is usually seen in eclamptic patients[13]; but she was deeply drowsy and had mild right hemiparesis. Visual system was not checkable, but the pupillary reflex was prompt. All hematologic, rheumatologic, and cardiologic work ups including electrolytes, blood count, coagulation test, and renal function test(for proteinuria and hyperuricemia), hematologic antithrombin III, protein C and S, immunologic rheumatoid factor, lupus anticoagulant, antinuclear antibody, erythrosedimentation rate and C3-C4 concentration, electrocardiography and echocardiography revealed no remarkable abnormalities.

The cerebrospinal fluid analysis was normal. Initial brain computed tomography(CT) scan showed about 15cc of intracerebral hematoma(ICH) in the left mesial temporal lobe and intraventricular hemorrhage(IVH) in the left lateral and third ventricles. Despite administering mannitol, her consciousness level progressively deteriorated. Magnetic resonance imaging(MRI) and Magnetic resonance angiography(MRA) were obtained 3days and cerebral angiogram was obtained 5days after the symptom onset. The MRI demonstrated ICH in the mesial temporal lobe, IVH in the lateral and third ventricles and there was an ischemic signal along the cingulum and medial frontal lobe. The MRA and cerebral angiogram demonstrated a diffuse multiple narrowing of the distal intra-

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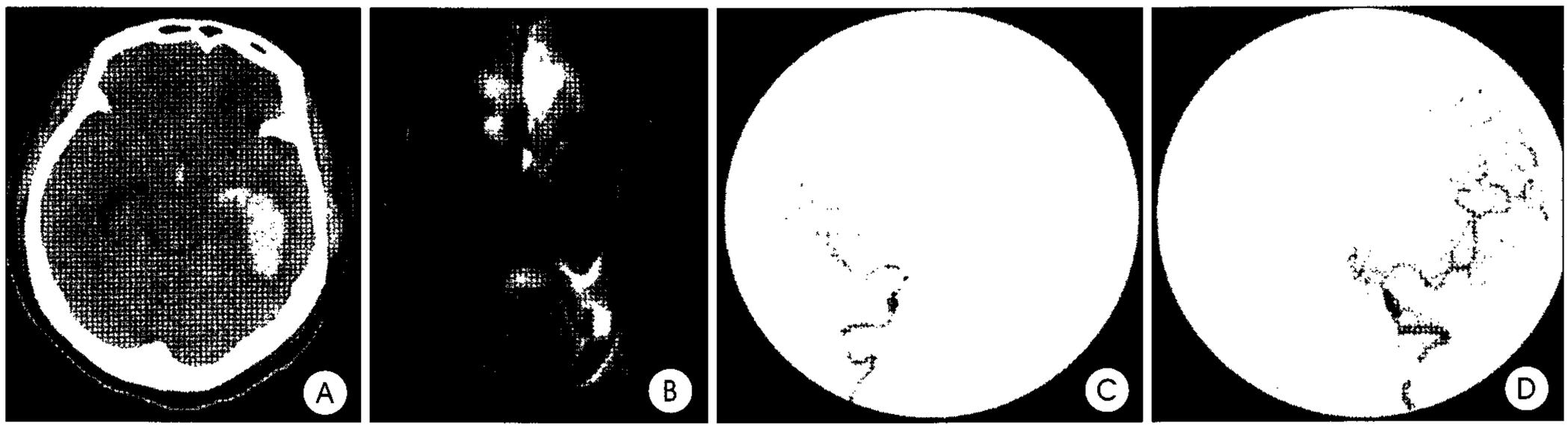

Fig. 1. Initial computed tomography, diffusion weighted magnetic resonance image at 10th day after symptom onset and cerebral angiograms at 12th day after symptom onset. These show intraventricular hemorrhage, cerebral infarction and multiple segmental narrowing of intracerebral arteries.

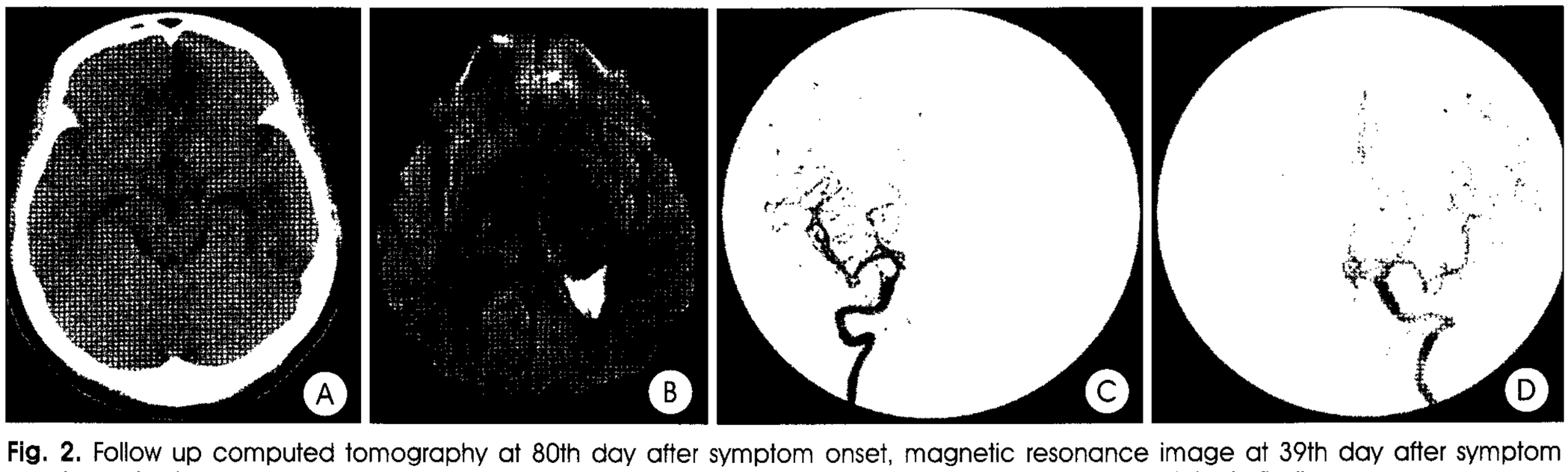

Fig. 2. Follow up computed tomography at 80th day after symptom onset, magnetic resonance image at 39th day after symptom onset, cerebral angiograms at 48th day after symptom onset. These show much improved state of radiologic findings.

cerebral artery, proximal middle cerebral artery, anterior cerebral artery and vertebrobasilar artery (Fig. 1). Thus, she was diagnosed as PPCA because she did not meet the clinical criteria for the diagnosis of eclampsia, such as proteinuria, edema, hypertension, and convulsion. She was given nimodipine 48mg/day and methylprednisolone 1,000mg/day intravenously for 1week, and drugs were then tapered gradually over a week. Her consciousness level and her neurological deficits were improved, however she complained of blindness. Both pupils were isocoric and the pupillary light reflex was prompt. At 49days after the symptom onset, repeated cerebral angiogram revealed that the previously noted multiple segmental narrowings became much improved. The follow-up brain MRI showed decreased ischemic signal in the frontal lobe and there were no abnormal signals in both occipital lobes (Fig. 2). Visual evoked potential(EP) was done and it revealed cortical blindness. Her vision has been improving and she could count fingers 30cm in front of her eyes 2months after the first symptom onset.

Discussion

Postpartum cerebral angiopathy(PPCA) as the cause of ischemic and hemorrhagic stroke is a rare clinical entity and it has been known as a benign cerebrovascular disease of the postpartum period[9,13]. The majority of PPCA occur during the first week. PPCA presents with a sudden onset of headache, nausea/vomiting, blurred vision or scotoma, seizure, a decreased consciousness level, and a focal neurological deficit after the normal delivery. The initial presentation is usually severe. The patients have no history of eclampsia and laboratory tests are usually normal. The cerebrospinal fluid analysis is normal or it shows a moderate pleocytiosis or elevated protein. CT/ MRI findings were cerebral edema, infarction, subarachnoid hemorrhage or intraparenchymal hemorrhage. Cerebral angiography revealed the classic picture of multiple segmental narrowings or alternating stenosis with ectasia of multiple intracranial arteries, and these findings were not confined to the area of hemorrhage. These angiographic features were more prominent in the posterior circulating system[13]. In terms of the angiopathy, the etiology of PPCA is unknown. For the classic angiographic feature, the etiology of PPCA is presumed to be transient vasoconstriction, inflammation or acute and transient attack of hypertension. However, the rapid improvement of clinical symptoms and the resolution of the angiographic abnormalities within several weeks suggest transient vasoconstriction rather than a true inflammatory vasculitis. The experimental data by Garner et al have shown

that the acute elevation of blood pressure can produce areas of vasospasm and dilatation, and some reported cases of PPCA have been associated with acute and transient attack of hypertension[13]. The use of sympathomimetic drugs such as ergonovine and bromocriptine that have direct vasoconstrictive effects may also produce PPCA[1,3,8].

There is no concordant therapeutic conclusions for PPCA but calcium antagonists and high dose steroid therapy have been shown as effective treatment in several reports[1,3,10]. More recently, balloon angioplasty, a proven effective treatment method for cerebral vasospasm from other causes such as subarachnoid hemorrhage has been applied by Ringer et al and it was effective for improving both the angiographic narrowing and cerebral blood flow[11]. Repeated transcranial doppler(TCD) or transcranial color-coded sonography(TCCS) for detecting abnormal increased flow velocity are useful tools for the diagnosis in suspected patients or for follow up in confirmed patients[6,9]. There has been some mortal cases reported[7], but this disease generally has a relatively benign clinical course and the patients with PPCA show rapid clinical improvement after high dose steroid therapy, calcium antagonists, or angioplasty.

Conclusion

Postpartum cerebral angiopathy(PPCA) is known as a benign and a reversible cerebrovascular disease that can occur during postpartum period. The labratory findings are usually within normal limits. CT/MRI shows cerebral hemorrhage, ischemia, or both. The cerebral angiogram demonstrates the characteristic feature of multiple segmental narrowing in the majority of the intracranial vessels. Calcium antagonists, steroid and angioplasty are effective treatment. Recognition of the possibility of PPCA is important for the preclusion of unnecessarily aggressive and potentially toxic immunosuppressive therapy for a presumed vasculitis.

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Commentary

Tremendous changes of body physiology occur during pregnancy and the post partum period, which may explain high incidence of strokes in perinatal period. But interestingly the postpartum period is associated with much higher incidence of stroke than during pregnancy[6]. It is presumable that acutely elevated blood pressure during the labor or acute change of body physiology after delivery may cause vasospasm of the already hormonally influenced cerebral vessels. But vasospasm may occur as the cerebral vasoconstriction syndrome[5] of unknown etiology. Severe vasoconsticting and vasodilatation can result in cerebral infarction and/or hemorrhage[6]. Stroke may occur in pregnant women without any history of eclampsia or pre-existing cerebral vascular lesions. Prolonged vasospasm may progress to irreversible vasculitis in the long run[1]. Besides, large reduction of blood volume or hormonal changes during the postpartum period may precipitate cerebral infarction[3].

Though the patient was normotensive at admission and had no past history of hypertension or eclampsia, she probably had a period of acutely fluctuating blood pressure at some time during the perinatal period.

Vertebrobasilar system has relatively poor sympathetic innervation that these arteries are easily affected by the acutely rising blood pressure. The reason why the posterior circulating system of this case was affected more prominently could be explained by this anatomical characteristics[2,4].

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