Korean Journal of Pediatrics Vol. 52, No. 9, 2009
DOI : 10.3345/kjp.2009.52.9.1044
■ Case report ■

Spontaneous intracranial internal carotid artery dissection in a child with psoriasis

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= Abstract =

A 13-year-old girl with psoriasis of the elbow, trunk, and face suddenly developed a severe headache followed by left hemiparesis and facial palsy. Brain magnetic resonance imaging showed an acute infarction of the right temporofrontal lobe and basal ganglia on the T2- and diffusion-weighted images. Cerebral angiography showed pre-occlusive irregular scalloped stenosis (99%) in the proximal M1 segment of the right middle cerebral artery and a web-like stenosis at the supraclinoid portion of the right internal carotid artery (ICA) suggestive of a spontaneous intracranial ICA dissection. The patient was administered a low dose of dipyridamole, and a rehabilitation program was initiated. Headache, left motor weakness, and facial droop improved within a week. However, mild left facial palsy and reduced fine motor function of the left hand were still present after 3 weeks. We report a rare case of spontaneous intracranial ICA dissection in a child with psoriasis. (Korean J Pediatr 2009;52:1044-1047)

Key Words : Internal carotid artery dissection, Psoriasiss

Introduction

Dissection of the internal carotid artery (ICA) is a cause of pediatric ischemic stroke, but not commonly reported[1-3]. Dissection of the ICA can occur spontaneously without an identified etiology or in the context of trauma or physical exertion as trivial as lifting a heavy object, coughing or straining during a bowel movement[2-4]. The etiology of spontaneous intracranial ICA dissection is not well understood, but environmental and genetic risk factors have been implicated[2-5].

The epidemiology of psoriasis shows that a number of diseases are associated with psoriasis more often than expected compared to individuals without psoriasis[6]. These include arthritis, Crohn's disease, cardiovascular disease, hypertension, diabetes, obesity, and chronic oropharyngeal infections[6]. There have been a few case reports of cerebrovascular events in patients with psoriasis[7-9]; most suspected cases have been associated with thromboembolic disorders[7]; rarely with moyamoya disease[8]. However, cerebral arterial dissection involving the intracranial ICA has not been previously reported in a patient with psoriasis. Here we report a case of spontaneous intracranial ICA dissection in a child with psoriasis.

Received : 22 June 2009, Revised : 14 August 2009,
Accepted : 22 August 2009
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Case report

A 13-year-old girl had sudden development of a severe right hemi-cranial headache while riding a bicycle, which was immediately followed by a contralateral hemiparesis. The patient was transferred to the Chonnam National University Hospital from a local clinic because of the sustained symptoms including a left facial palsy, slurred speech and altered mental status. The brain computed tomography (CT) revealed no abnormal findings. The patient had no history of neck or head trauma or unusual physical exertion. At the time of the event, there was no preceding febrile illness. The patient was relatively healthy without previous vascular risk factors. The patient was treated for psoriasis. Psoriasis was diagnosed six years previously; the patient had chronic relapses; the psoriasis was aggravated in recent days prior to presentation. The family history for acute stroke was unremarkable.

The brain magnetic resonance imaging (MRI) showed ill-defined hyperintense lesions involving the right temporofrontal lobe and basal ganglia on the T2 and diffusion weighted images, consistent with the acute stage of a cerebral infraction without hemorrhage (Fig. 1). The MR angiography and cerebral angiography showed pre-occlusive irregular scalloped stenosis (99%) in the proximal M1 segment of the right middle cerebral artery (MCA) and a web like stenosis in the supraclinoid portion of the right ICA (Fig. 2), suggestive of a dissection of the ICA. In addition, there was a marked decrease in blood flow at the right MCA. However, the left carotid and posterior circulation appeared normal.

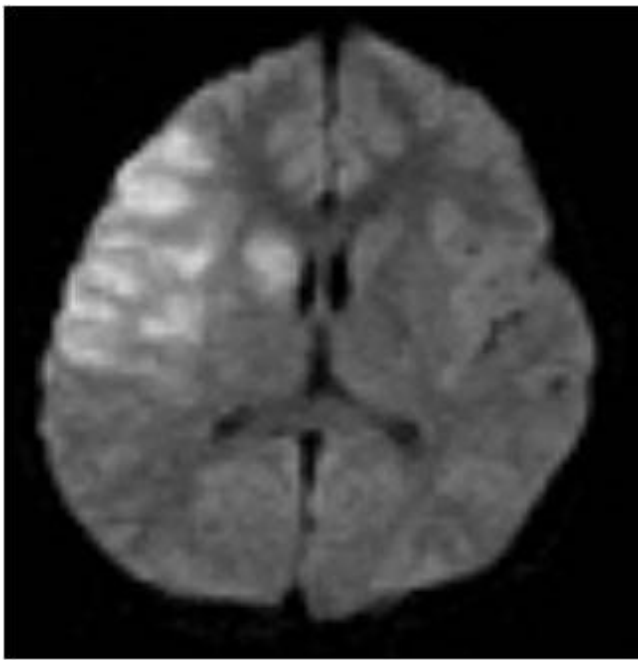

Fig. 1. The magnetic resonance diffusion-weighted image shows hyperintense lesions involving the right temporofrontal lobe and basal ganglia suggesting the acute stage of cerebral infraction in the right middle cerebral artery region.

The laboratory findings were all normal, including: a complete blood cell count, erythrocyte sedimentation rate, plasma lactate and pyruvate levels, antinuclear antibody, rheumatoid factor, anti-double-stranded DNA antibody, fasting lipid profiles, antithorombin-III, fibrinogen, protein C and S, prothrombin time, activated partial thromboplastin time, and cerebrospinal fluid analysis. The electroencephalography showed bifrontal slow delta waves but no epileptiform discharges. The electrocardiography, echocardiography and abdominal ultrasound were all normal.

The patient was treated with low dose dipyridamole (5 mg/kg/day) for antiplatelet activity to prevent stroke recurrence. The skin lesions including multiple erythematous ring shaped patches with scales on the elbows, back, buttocks, chest, and face (Fig. 3) were confirmed to be psoriasis by skin biopsy and treated by a dermatologist. A rehabilitation program was initiated for the residual ab-

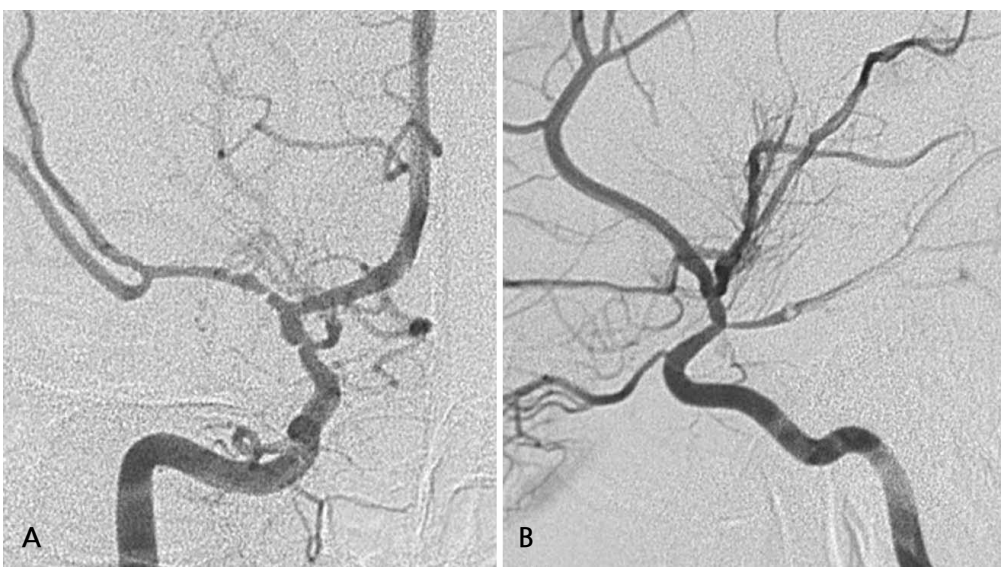

Fig. 2. Right anteroposterior (A) and lateral (B) cerebral angiography shows pre-occlusive irregular scalloped stenosis (99%) in the proximal M1 segment of the right middle cerebral artery (A) and a web-like stenosis in the supraclinoid portion of the right internal carotid artery (B) consistent with a dissection of the intracranial internal carotid artery.

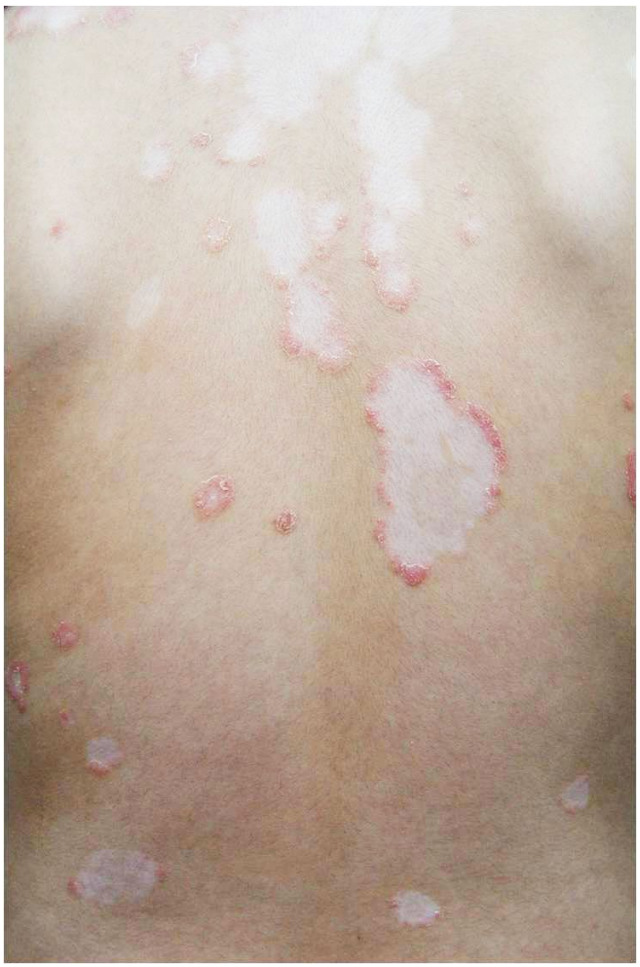

Fig. 3. The skin lesion on the back with multiple erythematous ring-shaped patches and scales suggests psoriasis.

normalities from the stroke.

The headache and the left motor weakness and facial droop improved within a week. However, the mild left facial palsy and dull fine motor responses of the left hand remained even at three weeks after the stroke. The brain CT followed-up on the eighth day, confirmed an infarction in the right temporofrontal lobe and basal ganglia with hemorrhagic transformation. A brain MRI and MR angiography were recommended two months after the stroke but was refused by the parents. The patient was then lost to follow up after discharge.

Discussion

Spontaneous *intracranial* ICA dissections are considered rare and uncommon when compared with those of the vertebrobasilar system or the ICA's cervical portion that are mobile and more susceptible to mechanical factors in the pathogenesis of dissection[3-5]. However, in children than in adult, an *intracranial* ICA dissection is more commonly reported[1]. Interestingly, adults with intracranial dissections were reported to have a younger age at onset than those with extracranial dissections[1]. The typical angiographic findings associated with an intracranial ICA dissection are: the string sign, a double lumen, irregular scalloped stenosis and vessel occlusion usually seen when the dissection involves the subintimal and intramedial layers[5].

The etiology of spontaneous intracranial ICA dissection is not well understood, but it is likely multifactorial[4]. Many environmental and genetic risk factors have been found in patients with spontaneous ICA dissections such as:[4] heritable connective tissue disorders including Ehlers-Danlos syndrome type IV, Marfan syndrome, autosomal dominant polycystic kidney disease, and osteogenesis imperfecta[3, 4]; an underlying vasculopathy such as fibromuscular dysplasia[2, 5], cystic medial necrosis, an intimal fibroelastic aberration, atherosclerosis[5], α1-antitrypsin deficiency, and hyperhomocystinuria[2], as well as infections[2, 4]. However, psoriasis has not been reported previously in patients with ICA dissection.

In patients with psoriasis, there have been a few case reports of cerebrovascular events[7-9]; most suspected cases were associated with thromboembolic disorders[7], rarely with moyamoya disease[8]. McDonald CJ et al.[7] reported that 29 patients (11.5%), among a total of 253 patients with psoriasis that had one or more episodes of thromboembolic events such as a cerebrovascular accident, myocardial infarction, thrombophlebitis, or pulmonary embolization. Stratigos AJ et al.[9] described a case of spontaneous clearing of psoriasis after a stroke in an elderly woman with chronic plaque psoriasis.

The temporal association of these two disorders in our case, of psoriasis and spontaneous intracranial ICA dissection, is suspected to have occurred coincidentally. Psoriasis is a common cutaneous disorder affecting up to 2-3% of western populations[6]. However, we also suspect that the psoriasis might have preceded the spontaneous intracranial ICA dissection and somehow might have been associated with it. Psoriasis is not common in eastern countries including Japan[6], and in our case the psoriatic lesions were aggravated several days before the stroke. However, further reports are needed to confirm an association.

한글 요약

소아에서 건선과 함께 진단된 자발성 두개강 내 내경 동맥 박리 1례

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내경 동맥 박리는 드물기는 하나 소아 허혈성 뇌경색의 한 원인이며, 선행된 외상 후 발생하는 경우가 흔해 두개강 내 보다는 두개강 외에 발생하는 경우가 많다. 건선은 흔한 피부 질환 중 하나이며, 서양인에 비해 동양인에서는 그 빈도가 낮고 다른 전신 질환과 동반되어 보고되기도 한다. 건선은 혈전 색전증과 연관된 허혈성 심뇌혈관계 질환과 연관되어 보고된 바가 있기는 하지만, 혈관 박리와 연관되어 보고된 경우는 드물다. 저자들은 이전에 건선으로 치료 받고 최근 피부 증상이 악화되던 중 우측 반신 마비와 안면 마비가 발생해 내원한 13세 여아에서 자발성 두개강 내 내경 동맥 박리를 진단하여 보고하는 바이다.

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