

Childhood Primary Angiitis of the Central Nervous System Presenting as Isolated Middle Cerebral Artery Dissection

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Isolated middle cerebral artery dissection and primary angiitis of the central nervous system are two rare but serious causes of ischemic stroke in children. We report a case of a patient who presented with headache, left-sided hemiparesis, and left hemi-hypoesthesia with acute cerebral infarction in the right middle cerebral artery territory. Brain magnetic resonance imaging showed isolated right middle cerebral artery dissection with arterial wall contrast enhancement extending beyond the length of the dissection and diffuse narrowing with stenosis in the right posterior cerebral artery suggesting angiitis. After exclusion of other diagnoses the patient was treated with corticosteroids and dual antiplatelet therapy. Follow up magnetic resonance imaging performed 4 months later showed partially improved contrast enhancement in the right middle cerebral artery and persistent narrowing and focal stenosis of the right posterior cerebral artery. Although rare, primary angiitis of the central nervous system should be considered in the differential diagnosis of acute stroke in children. High-resolution vessel wall imaging magnetic resonance studies can provide important information in determining the etiology of stroke.

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Isolated middle cerebral artery (MCA) dissection is a rare but serious cause of acute ischemic stroke in children. In most cases, the etiology is traumatic or idiopathic. Childhood primary angiitis of the central nervous system (cPACNS) is also a rare condition and only recently has been recognized as a potentially fatal cause of childhood stroke.¹ Childhood PACNS is categorized into two subtypes: angiography-negative small vessel cPACNS, and angiography-positive large vessel cPACNS. Angiography-positive large vessel cPACNS is further divided into progressive and nonprogressive disease. Although there are no universal management guidelines for cPACNS, treatment usually consists of a combination of immunosuppressive and antithrombotic agents to prevent disease progression and com-

plications.² Herein we report a case in which cPACNS presented with unilateral dissection of the right MCA and multiple stenoses of the right posterior cerebral artery (PCA).

CASE

A 14-year-old healthy male with no significant previous medical history presented with acute sudden onset left-sided hemiparesis after a paroxysmal but severe headache during defecation. Recent history of trauma to the head and neck was denied. Initial neurological examination revealed left hemi-hypoesthesia whilst signs of extinction, anosognosia, and asomatognosia

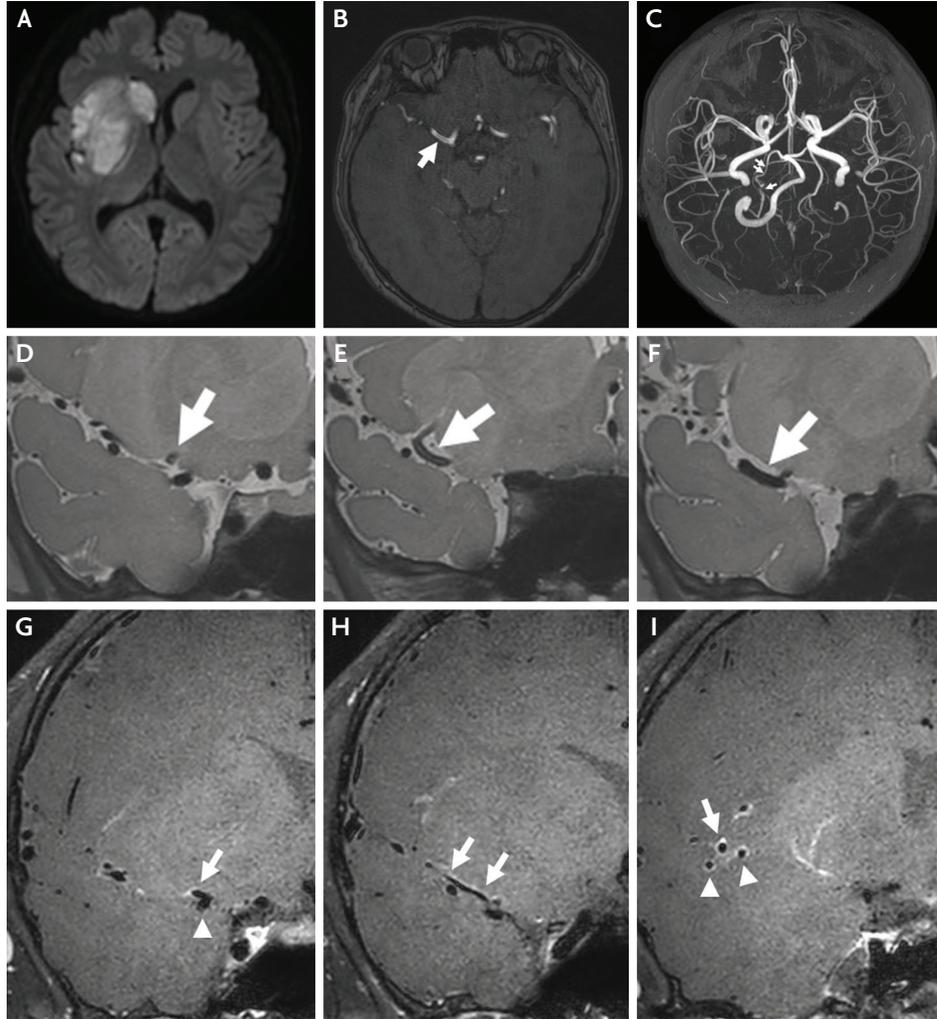


FIG. 1. Initial brain magnetic resonance imaging revealing (A) acute infarction in the right MCA territory on axial T2 FLAIR and (B) diffuse double lumen-like lesion at the right MCA (arrow) on axial time-of-flight angiography. High-resolution vessel wall imaging showing (C) diffuse narrowing and multiple stenosis of the right PCA (small arrows), (D, E) intimal flap (arrows) at the superior division of the right MCA, (F) with no involvement of the inferior division of the right MCA (arrow), and diffuse arterial wall enhancement at the (G) proximal M1 (arrow), (H) mid M1 (arrows), and (I) M2 segments of both the superior (arrow) and inferior (arrowheads of G, I) divisions.

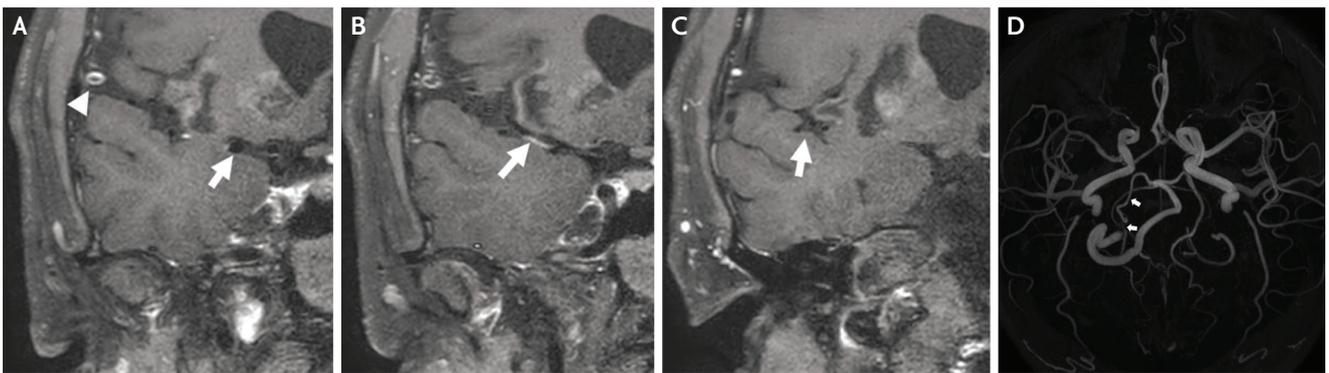


FIG. 2. Follow-up coronal T1 contrast enhanced magnetic resonance imaging. (A) Improvement of wall enhancement at the right proximal M1 (arrow). Concentric wall enhancement at the right M3 (arrowhead of A) can also be seen. (B) Right mid-M1 with remaining wall enhancement (arrow). (C) M2 segments with resolved contrast enhancement (arrow). (D) Magnetic resonance angiography showing persistent narrowing and focal stenosis of the right PCA (arrows).

were not found. The initial National Institute of Health Stroke Scale score was 11 (facial palsy 2; left arm 4; left leg 4; sensory 1). He had no systemic symptoms, including fever, recent weight change, skin rash, arthritis, and dyspnea. Brain magnetic resonance imaging (MRI) was performed and showed acute right MCA infarction with a diffuse double-lumen sign at the right M1 (Fig. 1A, B).

Further workup via high-resolution intracranial vessel wall imaging showed early bifurcation of the right MCA at the proximal M1 segment. Arterial dissection was noted only at the superior division on proton density imaging. (Fig. 1D-F). There was arterial wall enhancement not only at the superior division but also at the M2 segment of the inferior division (Fig. 1G-I). Diffuse narrowing of the right PCA with multiple stenosis was also seen (Fig. 1C). The multiple intracranial lesions were unexplainable with only isolated arterial dissection and were therefore considered suggestive of angiitis. Family history was negative for rheumatologic or cerebrovascular disease. On cerebrospinal fluid (CSF) examination, CSF color was slightly yellowish, opening pressure was 2 cm H₂O, white blood cell count was 4/mm³, red blood cell count was 600/mm³, and protein was 36.0 mg/dL. Infection workup and cytology testing of the CSF were negative. Serum inflammatory markers including ESR (3 mm/hour) and CRP (0.05 mg/dL) were within normal limits. Other than serum antinuclear antibodies (1:80), no laboratory evidence of antibody-mediated disorders was found. Therapy with high-dose intravenous methylprednisolone (1,000 mg per day for 5 days, switched to prednisolone 60 mg on day 6) and dual antiplatelet therapy was started, and the patient was transferred to another hospital for rehabilitation. 4 months later follow up brain MRI showed improved right M1 stenosis and contrast enhancement, but narrowing and focal stenosis of the right PCA persisted (Fig. 2). His 1-year functional status as assessed on the modified Rankin scale was 3.

DISCUSSION

MCA dissection in children is rare. Nevertheless, phy-

sicians should consider intracranial artery dissection in the differential diagnosis of ischemic stroke in children. Concerning the etiology of dissection, although the prevalence of traumatic or idiopathic dissection is higher, the possibility of cPACNS should also be considered. When cPACNS is suspected, high-resolution vessel wall imaging magnetic resonance studies can be helpful. Since cPACNS is a rare and unfamiliar entity, it can be challenging to diagnose. However as untreated cPACNS is associated with high morbidity and mortality, early diagnosis and initiation of immunomodulatory therapy are of great importance.

Ethics Statement

This study was approved by the Institutional Review Board (IRB) of Boramae Medical Center (IRB No. 20-2022-42). The IRB waived the need for informed consent.

Availability of Data and Material

The authors confirm that the data supporting the findings of this study are available within the article.

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Conflicts of Interest

No potential conflicts of interest relevant to this article was reported.

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