CASE REPORT

Benign Angiopathy of Central Nervous System

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Benign angiopathy of central nervous system (BACNS) is rare in children. We report a seven-year-old boy presenting with sudden severe headache and progressive external ophthalmoplegia. Magnetic resonance angiography (MRA) showed diffuse segmental narrowing of major cerebral arteries. Following a course of glucocorticoid, there was complete resolution of vascular lesions and follow-up MRA did not show any evidence of new lesions.

Key words: *Angiopathy, Benign, Central nervous system, Ophthalmoplegia, Primary angiitis.*

Primary angiitis of the central nervous system (PACNS) is rare in children. The disease usually runs a progressive, fatal course and is histopathologically characterized by the presence of marked granulomatous vasculitis(1). A subset of patients with PACNS has a relatively benign clinical course and is termed as benign angiopathy of central nervous system (BACNS)(1). Correct diagnosis requires a high degree of suspicion along with knowledge of other diseases that may masquerade as vasculitis (1).

CASE REPORT

A seven-year-old boy presented with history of sudden onset of severe headache, vomiting and low-grade fever for seven days associated with increasing diplopia, blurring of vision and photophobia for three days. There was no history of head injury, physical weakness or sensory loss. There was no past history of febrile illness with rash suggestive of varicella. No similar illness was reported in any other family members.

On examination, he was well oriented and interactive. A resting tachycardia (heart rate 166/min) and hypertension (116/84 mmHg) was found. Bilateral ptosis, mydriasis, loss of direct and consensual reflex and severe limitation of eye movements on all directions were noted. Fundoscopy was normal. No other focal neurological deficit was found. Other systems were normal.

Baseline hematological study was normal. Cerebrospinal fluid showed a normal cell count with a high protein content (200mg %). An awake EEG record showed slow background activity suggesting encephalopathy. A diffuse high signal intensity involving the occipito-parietal cortical and sub cortical regions were shown in T2 weighted MRI. MR angiography revealed a diffuse narrowing of right sided internal carotid artery, bilateral middle cerebral arteries and vertebrobasilar arteries (***Fig.*1**). Autoantibody screen, serum homocysteine, protein C, protein S, antithrombin III, antiphospholipid antibody levels were normal. HIV-ELISA was non reactive.

A presumptive diagnosis of primary CNS angiitis was considered. Treatment was initiated with intravenous dexamethasone given for one week followed by oral prednisolone (1 mg/kg/day) for four weeks. Hypertension was treated with oral nifedipine. Major symptomatic improvement was noted within two weeks and he became asymptomatic by 3rd week. Prednisolone was tapered and discontinued in another two weeks. MR

angiography two and six months later showed complete resolution of all the previous lesions and no evidence of any new lesion.

Considering the complete resolution of the angiographically demonstrated vascular narrowing following a course of steroid treatment a final diagnosis of benign angiopathy of central nervous system (BACNS) was made.

DISCUSSION

Diagnostic criteria of primary angiitis of CNS were proposed in 1988 with clinical description of 46 cases worldwide. These criteria included (*i*) an unexplained neurologic deficit despite aggressive diagnostic evaluation; (*ii*) a high probability angiogram for arteritis and/or histopathologic evidence of arteritis confined to the CNS; and (*iii*) exclusion of all those disorders capable of mimicking the angiographic findings or associated with vascular inflammation of the CNS(1). The clinical presentations are highly variable but the triad of headache, organic brain syndrome and multifocal neurodeficit is highly suggestive of the condition. Systemic symptoms are mostly absent in the affected patients as is laboratory evidence of inflammation(2).

Blood tests are not helpful in the diagnosis of CNS vasculitis. Cerebrospinal fluid (CSF) analysis is usually abnormal in patients with PACNS, showing pleocytosis and elevated protein levels, though it is usually normal in BACNS. CSF study is also helpful in ruling out vasculitic mimics(2). The most important diagnostic aid remains the MRI and MRA. Brain biopsy is considered gold standard for confirming diagnosis and starting prolonged immunosuppressive therapy in this condition(3). MRA findings include vascular beading and absence or cut off of one or more vessels seen in single or multiple vascular beds. Reversibility of angiographic abnormalities is essential for diagnosis of BACNS. Reduced sensitivity for smaller caliber vessels remains the major drawback of angiography(4).

BACNS is treated with tapering course of oral prednisolone and nifedipine for about 6 months which results in complete radiological recovery and appearance of no new lesions. On the contrary, PACNS cases usually need prolonged treatment with cyclophosphamide, methotrexate, azathioprine and prednisolone along with several MR imaging for monitoring of activity(5).

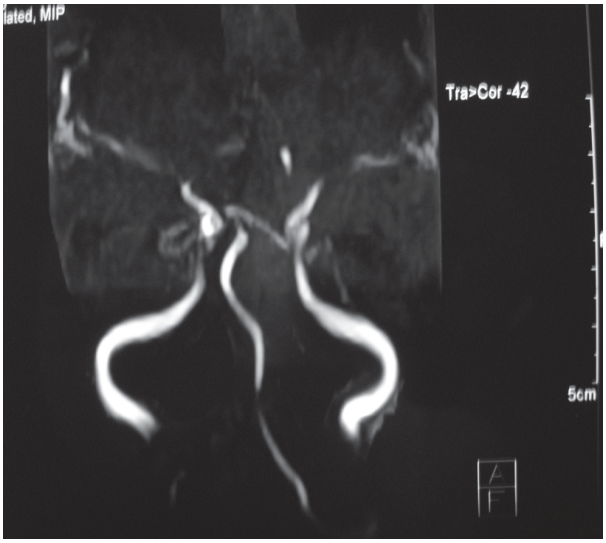

FIG.1 *Diffuse narrowing of right internal carotid and bilateral middle cerebral and vertebrobasilar arteries.*

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