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Case Report

Primary Internal Carotid Artery Vasculitis in a Child: A Rare Entity and Challenging Diagnosis

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Abstract Childhood primary central nervous system (CNS) vasculitis is a potentially reversible but serious inflammatory brain disease that manifests with acute or gradual onset neurological and psychiatric symptoms in children and adolescents. It can affect large or small blood vessels and can be categorised by size, location, aetiology or progression. Neurovascular imaging is crucial and mandatory to confirm the diagnosis of CNS vasculitis and may help to obviate the need for brain biopsy. We aimed to report a case of primary internal carotid artery vasculitis in a child, with presenting clinical and imaging findings.

Key words Central nervous system; Child vasculitis; MRI; Stroke

Introduction

Vasculitis is a heterogeneous group of pathologies characterised by inflammation of blood vessels that can be a fatal disease or cause severe and permanent disabilities.[1] Features of vasculitis vary greatly and classified depending on affected blood vessels or organs, aetiology (infectious/non infectious), local or systemic clinical manifestations and demographic characteristics (age, sex, ethnicity). Central nervous system vasculitis (CNS), a primary single organ vasculitis, may not be a component of systemic vasculitis. Primary CNS vasculitis may involve large intracranial arteries and frequently results in territorial cerebral infarction.[2,3] The diagnosis of primary vasculitis in childhood can be challenging and difficult due to rarity and variety of clinical manifestations. Herein, we present a case of primary ICA vasculitis in a 14-year-old boy with clinical and imaging features.

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Case Report

A 14-year-old boy was admitted to an external centre with the complaints of headache and weakness in the left leg. Initial non contrast-enhanced computed tomography (CT) was taken and found no abnormality. Two days after the first symptoms appeared, he was admitted to our hospital due to progression of his complaints. He had an episode of sudden loss of sensation in his left arm and leg, progressive weakness and difficulty speaking. Neurological examination revealed facial asymmetry, dysarthria and 3/5 motor strength in both upper and lower extremities. No family history and past medical history of an ischaemic event were recorded. Repeat head CT revealed hypodense areas and swelling of the grey-white matter in the right inferior frontal lobe extending to insular lobe and putamen (Figure 1a). On suspicion of acute ischaemic stroke, the patient underwent contrast-enhanced magnetic resonance imaging (MRI) combined with diffusion-weighted imaging (DWI) and magnetic resonance angiography (MRA). Diffusion restriction in the right frontotemporal lobe involving, insular lobe, centrum semiovale, putamen, internal and external capsule were compatible with acute ischaemic infarction (Figures 1b & 1c). MRA and contrast enhanced MRI showed concentric vessel wall thickening and prominent enhancement with marked narrow lumen, near-occlusive stenosis in the petrosal-cavernosal segment of the right internal carotid

artery (ICA) and resulting in acute ischaemia-infarct zones (Figure 2). To rule out ICA dissection, CT angiography was examined and no dissection flap was seen. Rheumatoid-inflammatory markers in serum and cerebrospinal fluid were all within the normal ranges. The virology screening tests were negative. The final diagnosis of primary ICA vasculitis with unknown aetiology was made and was started with immunotherapy including enoxaparin and high dose methylprednisolone. Subsequently, methylprednisolone 1 mg/kg/day was given for 3 days and then gradually reduced to 0.5 mg/kg for maintenance, combined with enoxaparin 6000 U/day for anticoagulation. After the immunotherapy and anticoagulation, the patient recovered well. MRI and MRA examinations revealed that vascular wall thickening and contrast enhancement in the right ICA segments and lumen narrowing had regressed, and chronic infarct areas were limited to the lentiform nucleus (Figure 3). He was discharged after 4 weeks of hospitalisation. At discharge, he continued to have mild weakness in his left arm and leg, and he was directed to continue physical therapy. A follow-up examination four months later revealed no neurological deficits.

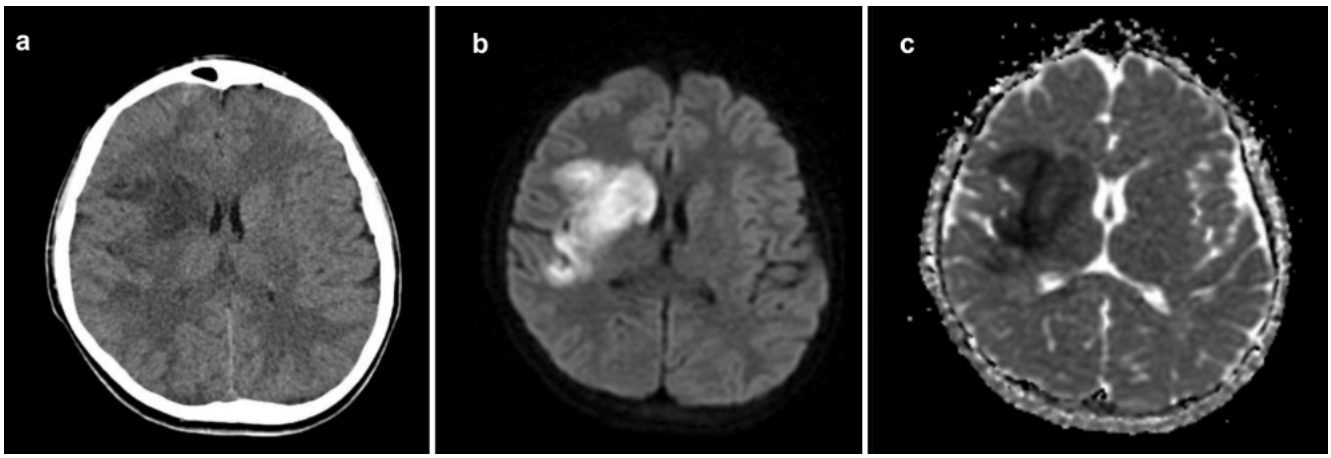

Figure 1 Non-contrast-enhanced axial CT image (a) showing hypodense areas and subtle loss of grey-white matter differentiation in the right frontotemporal lobe extending to putamen. (b,c) Diffusion weighted images (B 1000-b and ADC-c) demonstrate the diffusion restriction within these hypodense areas. These findings are suggestive of an ischaemic infarct by occlusion of the right ICA or branches.

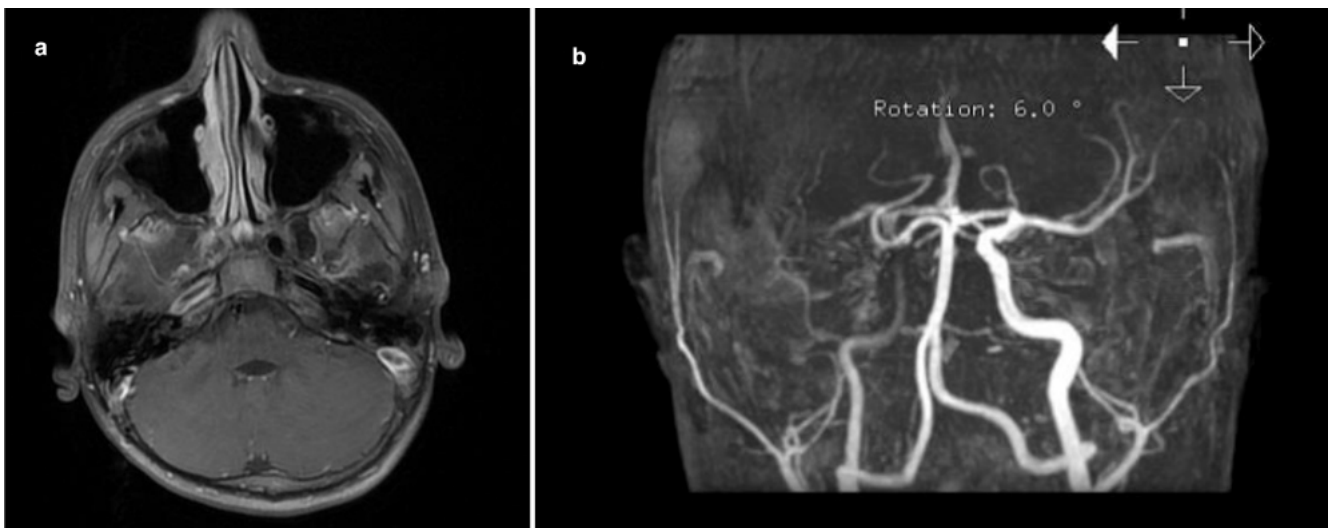

Figure 2 Contrast enhanced axial T1 weighted MR image (a) shows concentric vessel wall thickening and enhancement in the petrosal-cavernosal segment of the right ICA. MR angiography coronal image (b) shows markedly narrow lumen, near occlusion-severe stenosis in the petrosal-cavernosal segment of the right ICA.

Discussion

Vasculitis is defined as the presence of inflammation in the blood vessel walls, arteries, veins or capillaries.[2] If the aetiology of the inflammation is unknown, it is called primary vasculitis. On the other hand if there is an underlying disease then it is called secondary vasculitis. Since the pathophysiology has not yet been elucidated in most cases, it has become difficult to develop better diagnostic criteria for systemic vasculitis. The International Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitides (CHCC) convened and proposed a nomenclature system for the most common forms of vasculitis which provided names and definitions.[2,3] However, there are still several forms of atypical vasculitis including primary CNS vasculitis that do not fit in this classification. The primary CNS vasculitis is rare in children and only a few cases have been reported so far.[4,5]

In childhood, clinical presentation of primary CNS vasculitis is highly variable and depends on multiple factors such as the affected vessel, the extent of disease, the vascular sites involved, and the inflammatory pathophysiology. Ischaemic stroke and infarction resulting from vasculitis are more common clinical manifestation in infants and children. The diagnostic criteria are an acquired and otherwise unexplained neurological deficit, classical angiographic or histopathological features of CNS vasculitis, and the absence of another systemic disorder to explain these features.[2,6]

Primary CNS vasculitis can be classified as non-progressive or progressive vasculitis. If recurrent ischaemic events occur new ischaemic lesions develop on brain imaging, it is defined as progressive disease. Non progressive form is more common and affects boys more frequently than girls. It usually presents with unilateral stroke, severe headache and normal inflammatory markers in serum or cerebrospinal fluid. Imaging is crucial to confirm the diagnosis of CNS vasculitis. MRI typically demonstrates unilateral proximal vessel stenoses with gadolinium wall enhancement and multifocal parenchymal ischaemic lesions.[2,6,7]

Conventional angiography and MRA should be considered as diagnostic and monitoring tool for vessel stenosis.[6,7]

When primary central nervous system angiitis is clinically suspected in childhood, a brain biopsy may be necessary; supportive laboratory and neuroradiological studies and negative angiography may be necessary.[6,8] Accordingly, we noticed similarities in our case and non-progressive disease including male patient, stroke presentation, normal serum markers, affected unilateral ICA and not recurrence. Given the characteristic clinical presentation together with the vessel wall MRI findings

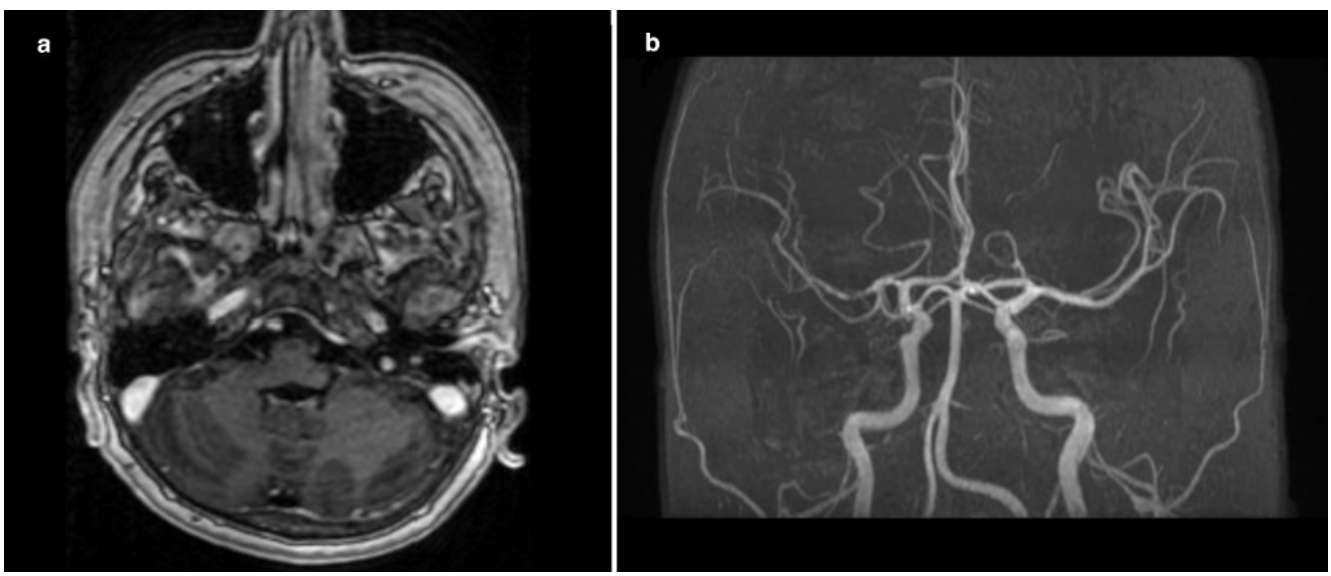

Figure 3 On control scans, Contrast enhanced axial T1 weighted MR image (a) and MR angiography coronal image (b) reveal vessel wall thickening and enhancement were regressed and significant improvement in lumen diameter of the right ICA and improved distal perfusion.

and exclusion of alternative diagnoses, brain biopsy was not considered necessary in our patient.

In large vessel CNS vasculitis, the main differential diagnosis includes infective vasculopathies, vasoconstrictive disorders, and rare genetic/metabolic diseases. Among the infections, postvaricella angiopathy is the most common infectious cause of vascular stroke in children and typically causes a unilateral inflammatory vasculopathy of the supraclinoid internal carotid, and/or proximal middle and/or anterior cerebral arteries.[6,9] For this reason, viruses, especially varicella zoster along with infective causative agents, were investigated in our patient and none were detected.

Also ICA vasculitis may be associated with inflammatory rheumatological diseases including Takayasu arteritis and giant cell arteritis. Especially the presence of cerebral infarction with large vessel vasculitis suggested Takayasu's arteritis in the first stage.[10] However, rheumatologic panel negativity and only ICA involvement ruled out the diagnosis of rheumatological diseases. Since the patient was in a paediatric age group, Moyamoya disease was also considered in the differential diagnosis. However, the absence of cerebral atrophy and collateral formation on MRI ruled out this diagnosis.[2,6]

Treatment recommendations include long-term corticosteroids to prevent recurrent ischaemic attacks and improve long-term neurological outcome and anticoagulation therapy for secondary stroke prophylaxis.[6]

Primary CNS vasculitis is an uncommon but potentially treatable cause of paediatric ischaemic stroke. Early recognition through comprehensive clinical evaluation and multimodal neurovascular imaging is essential for prompt diagnosis, exclusion of important mimics, and timely initiation of immunosuppressive therapy. This case highlights the added value of contrast-enhanced vessel wall MRI in establishing the diagnosis and monitoring treatment response, while potentially avoiding invasive procedures such as brain biopsy. Awareness of this rare entity may facilitate earlier diagnosis and improve neurological outcomes in affected children.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Conflicts of Interest

There are no conflicts of interest.

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