Vascular causes of acute ischemic stroke in the pediatric population: The crucial role of imaging

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Abstract

Background: Acute ischemic stroke (AIS) in a pediatric patient is a rare but life-threatening medical emergency. Unlike adults, pediatric strokes result from a diverse array of etiologies, with arteriopathies and cardiac anomalies playing major roles. Given the time-sensitive nature of stroke treatment, prompt identification and rapid imaging in the emergency setting are critical to improving prognosis in pediatric patients. We aimed to enumerate the principal vascular causes of pediatric AIS and to determine the specific contribution of different imaging modalities in the etiological evaluation.
Methods: We conducted a prospective study including all children aged less than 18 years who presented with AIS between January 2020 and December 2024. The study was carried out at Hedi Chaker University Hospital in Sfax.
Results: We included 22 patients (12 boys and 10 girls). Our patients were aged between one month and 9 years. Seven patients underwent ultrasonography with Doppler, seventeen patients underwent magnetic resonance imaging and eleven patients underwent computed tomography angiography. We noted arterial dissection in 8 cases, Moyamoya disease in 8 cases, hypoplasia of the internal carotid artery in 5 cases and fibromuscular dysplasia in one case. Imaging modalities provided critical diagnostic information, with Doppler ultrasound detecting flow abnormalities in arterial dissection and hypoplasia, magnetic resonance imaging delineating vascular occlusions and stenoses, and computed tomography angiography assisting in anatomical assessment. Representative cases illustrated the spectrum of vascular pathologies and imaging findings.
Conclusion: Pediatric AIS is frequently caused by a variety of vascular disorders that require multimodal imaging for accurate diagnosis. Early recognition and targeted imaging are crucial for timely intervention and improving outcomes in this vulnerable population.

Keywords: Ischemic stroke, Pediatric stroke, vascular diseases, Neuroimaging, Arterial dissection

Introduction

Acute ischemic stroke (AIS) in a pediatric patient is defined as a stroke occurring between the ages of one month and 18 years. It is a rare but serious medical emergency, with an estimated incidence of 2.69 per 100,000 children annually [1]. Unlike adults, in whom atherosclerotic disease is the predominant cause, pediatric strokes result from a diverse array of etiologies, with arteriopathies and cardiac anomalies playing major roles [2,3]. AIS accounts for nearly half of all strokes in children [4]. The early recognition of stroke symptoms in children is often delayed due to atypical presentations and a lower clinical suspicion, which can lead to delayed intervention and worse outcomes [5]. In contrast, 80 to 85% of strokes in adults are ischemic and more readily diagnosed due to more consistent clinical patterns [6]. Given the time-sensitive nature of stroke treatment, prompt identification and rapid imaging in the emergency setting are critical to improving prognosis in pediatric patients. Emergency physicians thus play a vital role in early diagnosis and coordination of care.

Among the vascular causes in children, focal cerebral arteriopathy, moyamoya disease, and arterial dissection are increasingly recognized contributors, often requiring targeted neurovascular imaging for accurate diagnosis [7,8]. Neuroimaging plays a central role not only in confirming the diagnosis of AIS but also in uncovering underlying vascular abnormalities. Magnetic resonance imaging (MRI), magnetic resonance angiography, computed tomography angiography (CTA), and digital subtraction angiography (DSA) are pivotal in establishing an etiological diagnosis and guiding management [9,10].

In this perspective, our work aims to enumerate the principal vascular causes of pediatric AIS and to determine the specific contribution of different imaging modalities in the etiological evaluation.

Methods

We conducted a prospective study including all children aged less than 18 years who presented with AIS between January 2020 and December 2024. The study was carried out at Hedi Chaker University Hospital in Sfax.

Inclusion criteria consisted of patients under 18 years of age with AIS confirmed by neuroimaging. Patients with hemorrhagic stroke, transient ischemic attacks, or incomplete imaging were excluded.

Clinical data including demographic information, clinical presentation, vascular risk factors, and relevant past medical history were collected.

Imaging protocol involved the use of MRI with diffusion-weighted imaging (DWI) or CTA when MRI was contraindicated or unavailable. Ultrasonography with Doppler was performed when vascular flow abnormalities or arterial dissections were suspected, providing a non-invasive initial assessment of cervical vessels.

All imaging studies were independently reviewed by two experienced neuroradiologists who classified vascular abnormalities according to standardized criteria. Discrepancies were resolved by consensus.

Primary outcomes included identification and characterization of vascular etiologies of AIS and evaluation of the diagnostic yield of each imaging modality in determining stroke cause.

Results

We included 22 patients (12 boys and 10 girls). Our patients were aged between one month and 9 years. Seven patients underwent ultrasonography with Doppler, seventeen patients underwent MRI and eleven patients underwent CTA.

We noted arterial dissection in 8 cases, Moyamoya disease in 8 cases, hypoplasia of the internal carotid artery (ICA) in 5 cases and fibromuscular dysplasia in one case.

Case 1

A six-year-old boy suffering from neck pain and a motor deficiency after rapid movement of his head. Ultrasonography of the cervical portions of the right arterial carotid showed intraluminal hyperechogenic linear image at the origin of the artery (Figure 1). An MRI was performed showing dissection of the right primitive carotid artery (Figure 1). The final diagnosis was arterial dissection.

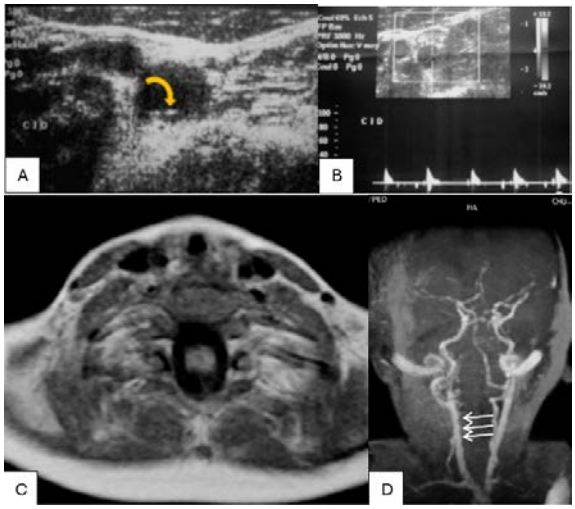

Figure 1: Ultrasonography of the cervical portions of the right arterial carotid showing intraluminal hyperechogenic linear image (arrow) at the origin of the artery: intimal flap *(A) and resistive spectrum, amortized at the origin of the right internal carotid artery (B). MRI showing dissection of the right primitive carotid artery, parietal hematoma in discreet hypersignal isosignal T1 (C) and parietal irregularity of the right primary carotid artery (arrows) (D).*

Case 2

A 4-year-old boy suffering from right hemiparesis. A cerebral angiography was performed showing important bilateral collateral circulation with filiform aspect of the left internal carotid and the sylvian arteries and occlusion of the left posterior cerebral artery (Figure 2). The final diagnosis was Moyamoya disease.

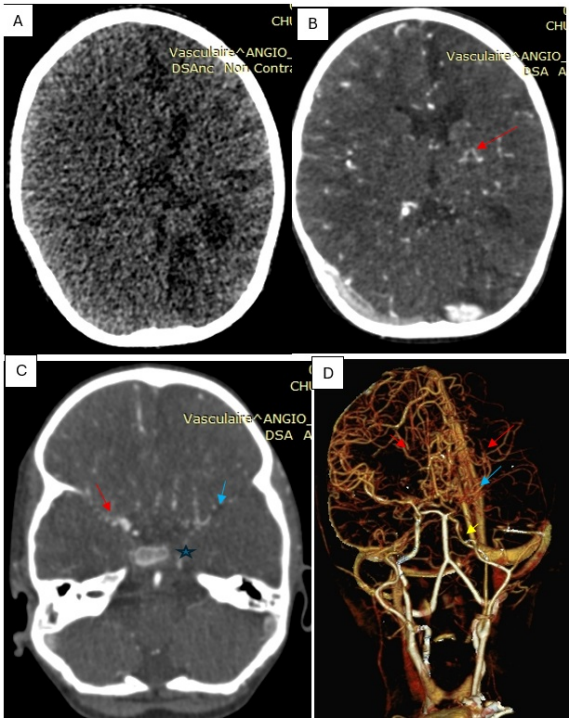

Figure 2: Computed tomography brain scan, axial section in parenchymal window without *(A) and after injection of contrast (B,C) and 3-dimensional reconstructions cerebral angiography (D) showing important bilateral collateral circulation (red arrow) with filiform aspect of the left internal carotid (yellow arrow) and the sylvian arteries (blue arrow) and occlusion of the left posterior cerebral artery (blue star).*

Case 3

Infant of 18 months, hospitalized for disorder of the state of consciousness. Computed tomography scan with MIP showed stenosis of the right internal carotid artery with development of vascular supplementation (Figure 3). The final diagnosis was Moyamoya disease.

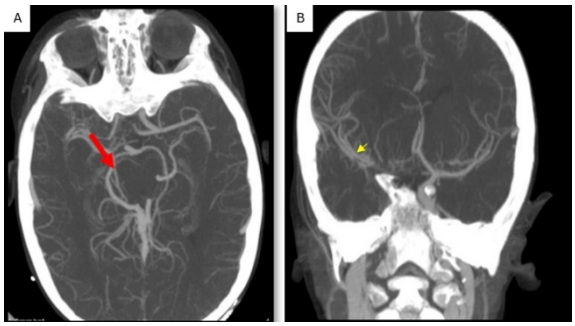

Figure 3: Computed tomography scan with MIP: showing stenosis of the right internal carotid artery (arrow)
(A) with development of vascular supplementation (yellow arrow) (B)

Case 4

7-year-old child suffering from generalized convulsions. MRI angiographic sequence showed progressive bilateral occlusion of bilateral internal carotid arteries with development of vascular supplementation. The final diagnosis was Moyamoya disease.

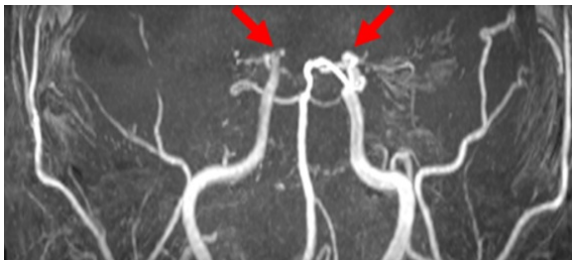

Figure 4: MRI angiographic sequence showing progressive bilateral occlusion of bilateral internal carotid arteries (arrows) with development of vascular supplementation.

Case 5

4-year-old girl, with a previous medical history of Minkowski Chauffard's disease, hospitalized for paresis of right arm of brutal installation. Computed tomography scan showed left frontal and parietal ischemic lesions. MRI angiographic sequence showed hypoplasia of the left internal carotid artery with obliteration in its distal portion. (Figure 5). Doppler ultrasound was performed. Ot showed acceleration of velocities (280 cm/s) with visible aliasing phenomenon in color coding in the right internal carotid artery without image of thrombosis or stenosis (Figure 6). An intravascular aliasing phenomenon in the right internal carotid artery with decreased flow in the left internal carotid artery (85 cm/s) were noted. The final diagnosis was hypoplasia of the internal carotid.

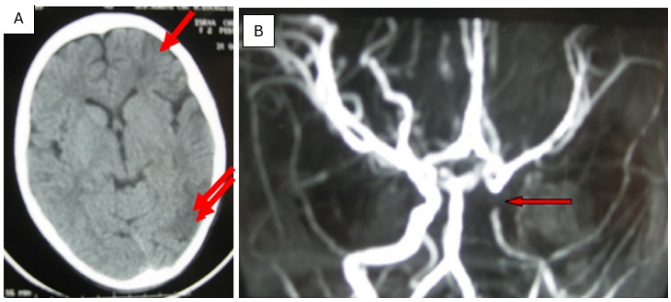

Figure 5: Computed tomography scan showing left frontal and parietal ischemic lesions
(A) and hypoplasia of the left internal carotid artery with obliteration in its distal portion (arrow) (B: MRI angiographic sequence)

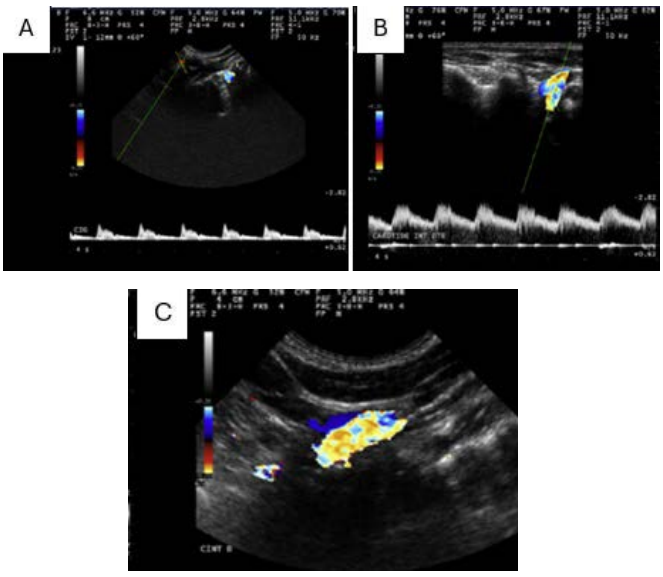

Figure 6: Doppler ultrasound showing acceleration of velocities (280 cm/s) with visible aliasing phenomenon in color coding in the right internal carotid artery without image of thrombosis or stenosis.

Case 6

A girl aged 8 years consulting for multiple strokes in the vertebral basilar territory. Computed tomography scan with injection showed pearled appearance at the origin of the right posterior cerebral artery. MRI angiographic sequence revealed short stenosis at basal trunk termination (Figure 7). The final diagnosis was fibromuscular dysplasia.

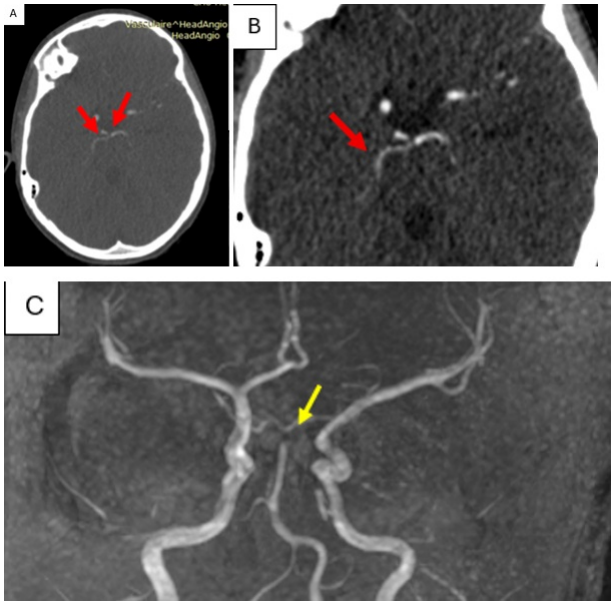

Figure 7: Computed tomography scan with injection showing pearled appearance at the origin of the right posterior cerebral artery (red arrows) *(A, B) and short stenosis at basal trunk termination (MRI angiographic sequence yellow arrow) (C)*

Discussion

Our study highlights the diverse vascular etiologies underlying pediatric AIS, with arterial dissection and Moyamoya disease being the most frequent diagnoses. Our findings emphasize the critical role of multimodal imaging in accurately identifying these vascular abnormalities, which is essential for timely diagnosis and management. The risk factors for stroke in children are congenital heart disease, infection, prothrombotic disorders, trauma, acquired and congenital vascular disease, metabolic disorders, and mitochondrial disease [11,12,13,14].

The clinical presentation is useful for localizing the lesion. The majority of pediatric ischemic strokes occur in the distribution of the middle cerebral artery, which results in hemiplegia with upper limb predominance, hemianopsia, or dysphasia [15,16]. Primarily lower extremity weakness would suggest anterior cerebral artery involvement, whereas vertigo, ataxia, and nystagmus are consistent with an ischemic event in the posterior circulation [13].

Vascular diseases alone account for one-third of cases. They are varied, including vasculitis, post-infectious causes, arterial dissection, arteriopathy (Takayasu arteritis, Moyamoya disease, cryptogenic arteriopathy), fibromuscular dysplasia, hypoplasia of the internal carotid artery, connective tissue diseases, and metabolic vasculopathy (e.g., Fabry disease) [11,17,18].

A. Arterial Dissection

Carotid or vertebral artery dissection results from a tear in the vessel lining wherein the intima separates from the media, creating a false or pseudo lumen, often accompanied by hemorrhage into the arterial wall [19,20]. Its annual incidence is 1–1.5 per 100,000 persons [14]. It can be traumatic or iatrogenic. Traumatic dissections may result from blunt trauma or rapid movement of the head in relation to the neck in any axis. While most dissections occur in the internal carotid artery, children may dissect

intracranially [21,19]. It accounts for 7% to 20% of all cases of childhood AIS [11,13].

Iatrogenic dissection may result from catheter or surgical manipulation of vessels in procedures such as angiography and endarterectomy. In rare cases, it may be related to underlying connective tissue disorders [11,19].

Ultrasonography of the cervical portions of the carotid and vertebral arteries can be useful in detailing flow aberrations, intramural hematoma, luminal thrombus, and mobile flaps [20]. It shows that velocities within the carotid bulb may decrease and are accompanied by high resistance due to stenosis that yields a biphasic pattern.

Cerebral angioscan is the most efficient modality for diagnosing dissection. An intimal tear within the vessel is often accompanied by formation of a medial or subendothelial hematoma that is readily identifiable [20]. An intramural hematoma usually manifests as a crescentic hyperdensity or suboccipital rim with thickening of the vascular wall without a change in the vessel caliber.

MRI with fat saturation has replaced conventional imaging as the gold standard for diagnosing craniocervical arterial dissection [19,20]. MR evaluation consists of three components: DWI and FLAIR for infarction, T1/T2 imaging for intramural hematoma, and MRA for vascular lumen evaluation. The classic MRI dissection finding is an eccentric periluminal rim indicative of intramural hematoma [19,20].

B. Moyamoya Disease

Moyamoya ("puff of smoke" in Japanese) is a chronic cerebrovascular disorder characterized by progressive stenosis or occlusion of the intracranial internal carotid artery and its proximal branches, accompanied by a basal collateral network [17,22,23]. Generalized cerebral atrophy is a common finding [23]. Diagnosis relies exclusively on imaging, as pathological correlation is difficult [23,22].

Pathologically, moyamoya disease is characterized by intimal thickening of the terminal portions of the internal carotid bilaterally [17,23]. Although classically described in the ICA, over 50% of patients also have involvement of the posterior cerebral arteries [22].

C. Hypoplasia of the Internal Carotid Artery

Carotid dysgenesis is classified into agenesis, aplasia, and hypoplasia. Agenesis is complete failure of development; aplasia is failure despite a precursor structure; hypoplasia is incomplete development [24,25]. In the study by Taşar et al., the prevalence of ICA aplasia or hypoplasia was 0.13% [24]. Hypoplasia is characterized by narrowing of the ICA along its entire course due to incomplete development. Diagnosis is made by identifying the absence or reduced size of the bony carotid canal on skull base CT, confirming congenital rather than acquired pathology [24,25].

D. Fibromuscular Dysplasia (FMD)

FMD is a segmentary, non-atherosclerotic, non-inflammatory vascular disease that may result in stenosis, occlusion, aneurysms, or dissection of medium-sized arteries [18,26]. It is classified into three forms: intimal (10%), medial (80–90%), and adventitial (<5%) [18]. FMD predominantly affects women (9:1 ratio) aged 15 to 50 [18]. It is suspected in hypertension before age 30, refractory hypertension, or when associated with small kidney size. Carotid and vertebral involvement is less frequent than renal (10–35%) [26].

On angio-MRI or angio-CT, characteristic findings include focal or tubular stenoses and multifocal stenoses with a "string of beads" appearance, often complicated by aneurysms [18,26]. Surgical management is less common now due to the efficacy of percutaneous transluminal angioplasty [26].

Conclusion

Pediatric arterial ischemic stroke, though rare, constitutes a true neurological emergency that demands rapid recognition and diagnostic precision. Vascular pathologies—including arterial dissection, Moyamoya disease, fibromuscular dysplasia, and congenital anomalies—are among the most important causes and may not be immediately apparent in the emergency setting. Early neuroimaging, particularly with MRI and angio-CT, is crucial not only for confirming ischemia but also for identifying the underlying vascular etiology. Strengthening awareness of pediatric stroke presentations and vascular mimics in emergency departments is essential to reduce diagnostic delays and facilitate early intervention in this vulnerable population.

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