



Pediatric cerebral proliferative angiopathy presenting infratentorial hemorrhage

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Abstract

Cerebral proliferative angiopathies (CPAs) are distinct vascular malformations with varied clinical presentations and radiological findings from arteriovenous malformations (AVM) (Lasjaunias et al. in *Stroke* 39(3):878–85, 2008). They usually manifest with seizures if present supratentorial, headache, and progressive neurological deficits due to steal phenomenon or rarely with hemorrhage (Lasjaunias et al. in *Stroke* 39(3):878–85, 2008). Most of the patients are usually young females. Pediatric cases are extremely rare, with few cases reported till now. Here we report a child who presented with cerebellar bleed and diagnosed as CPA. The child was managed medically, and there was no change in caliber of the vessels after 18 months of follow-up. A short review of cases of CPA in pediatric age group presentations and management was undertaken in this case report.

Keywords Cerebral proliferative angiopathy · AVM · Infratentorial hemorrhage

Introduction

Cerebral proliferative angiopathies (CPAs) are distinct vascular malformations with varied clinical presentations and radiological findings from arteriovenous malformations (AVM) [1]. They usually manifest with seizures in supratentorial location, headache, and progressive neurological deficits due to stealing phenomenon or rarely with hemorrhage [1]. Most of the patients are usually young females [1]. Pediatric cases are

extremely rare, with few cases reported till now. Here we report a child who presented with cerebellar bleed and a short review of pediatric cases was undertaken.

Case description

A 12-year-old child presented to the emergency room with a history of being hit by a cricket ball on the head while playing. At the presentation to the emergency room, he was drowsy, obeying to commands with no focal neurological deficits. He was evaluated with CT (computed tomogram), which showed hemorrhage in the right cerebellar hemisphere size of 3 × 3.5 cm with mass effect over fourth ventricle and distortion and mild cerebral edema (Fig. 1). No hydrocephalus was noted. The patient was managed medically, and he improved in sensorium.

He was evaluated further with MRI (magnetic resonance imaging), which showed hemorrhage in the parenchyma and ill-defined clustered vessels with intervening normal neuroparenchyma in the right cerebellar hemisphere. MR angiogram showed arterial feeders from the anterior inferior cerebellar artery (AICA) and from the posterior inferior cerebellar artery (PICA) (Fig. 2b–d). Based on the findings a preliminary diagnosis of vascular malformation was made. He was further evaluated with digital subtraction angiogram (DSA), which on left vertebral injection showed an ill-defined lesion

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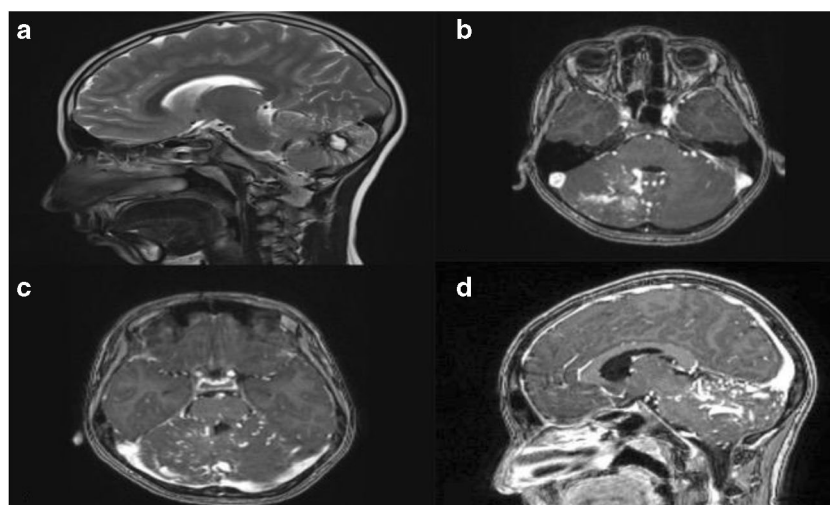
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Fig. 1 Right cerebellar hemisphere bleed with distortion of the 4th ventricle in CT brain plain and contrast images



in the right cerebellar hemisphere being fed by the superior cerebellar artery (SCA), AICA, and PICA on the right side. No obvious evidence of hypertrophy of feeding arteries or the early draining vein is seen; no definite nidus is seen (Fig. 3a–d). Based on these findings, a diagnosis of CPA was made. Due to the absence of mass effect, the child was managed symptomatically. At 3 months of follow-up, the child had recovered fully. At 1 year of follow-up, the child was fine without any deficits. Follow-up imaging at 18 months showed no change in proliferation or caliber of vessels (Fig. 4a, b).

Discussion

The total number of pediatric CPA which is reported in the literature is 13 (Table 1) including the current case, and the

mean age of all pediatric patients was 10.7 years with an age range of 2–18 years [2–4, 7–11]. Out of 13 patients, seven are female and six are male. In the original description of Lasjaunias et al., CPAs were found more common in young female (67%, $n = 33$), with a mean age of 22 years [1]. But in our review of pediatric cases, this rule was not applicable. As the individual details of pediatric patients in Lasjaunias series were not published, the exact incidence of the pediatric CPA cannot be quoted. Although Lasjaunias stated the range of CPAs in their series was from 10 to 65 years. Six patients presented with transient ischemic attacks involving motor deficits. Headache was present in six patients. Our case presented with cerebellar hemorrhage which is a very rare presentation.

Three patients underwent surgical intervention. All of them underwent indirect revascularization, with one patient having permanent deficits after 2 years of follow-up. Most of the

Fig. 2 a MRI T2 weighted image shows bleed in the right cerebellar hemisphere. b–d MRI post contrast shows ill-defined clustered vessels in the right cerebellar hemisphere. Feeders are from the anterior inferior cerebellar artery(AICA) and from the posterior inferior cerebellar artery(PICA)

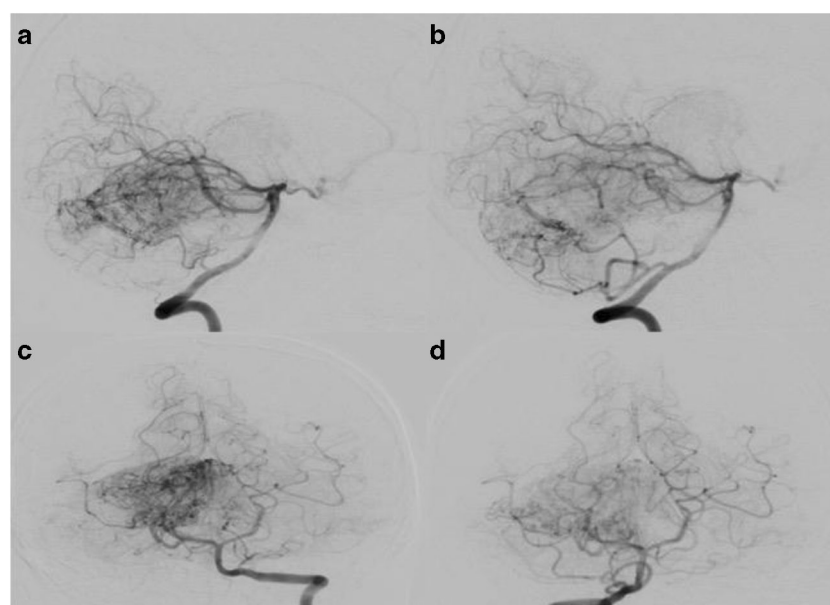
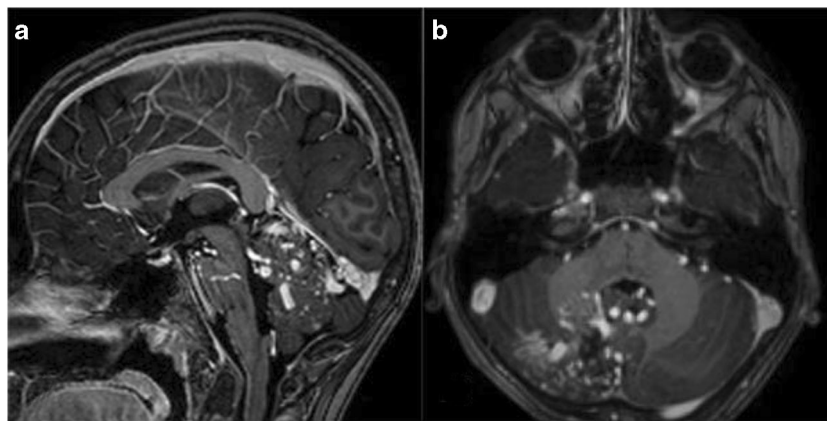


Fig. 3 **a, b** Lateral view of left vertebral injection showing ill-defined parenchymal blush in the right cerebellar hemisphere being fed by the superior cerebellar artery (SCA), AICA, and PICA. No obvious evidence of hypertrophy of vessels is seen. **c** and **d** show diffuse nidus



patients had multi-lobar clinical involvement causing ischemic symptoms probably due to vascular steal.

Epidemiology Vascular malformations are usually congenital in nature, and CPA is a rare disease and accounts for 3.4% of patients with brain AVM and commonly found in young females. Twenty-two percent of the patients had the infratentorial location of nidus [1]. They have distinct features from the classical AVM, like an absence of dominant feeders to a large area, the small size of the draining veins in comparison with the size of the nidus, the presence of intermingled brain in between the vascular channels, and absence of feeding artery aneurysms.

Unclear pathogenesis with hypoperfusion in the intermingled brain tissue and in the perimalformative area can be considered a triggering mechanism leading to cellular proliferation and endothelial hyperplasia causing uncontrolled angiogenesis, and it is also associated with segmental arterial stenosis [1, 5]. There are no known triggering mechanisms, and previous hemorrhages do not lead to increased risk. In our case, whether trivial injury has played a role in causing bleeding is not known, while some authors support the fact of increased risk of rebleed once hemorrhage has occurred because arteries have a

different origin as compared with classic brain arteriovenous malformation [1], but it is still controversial.

Clinical presentation and management

Patients usually present with seizures, followed by headache and hemorrhage [1] and some cases with a neurological deficit [2, 8]. Ischemic manifestations are usually more common than hemorrhage. Very few pediatric cases of CPA involving the posterior fossa have been reported [9]. One case of infratentorial CPA with hemorrhage has been reported in an elderly male [9].

Treatment can be varied depending on clinical features and severity of symptoms. Medical and surgical management have been described: medical management with analgesics and antiepileptic, to control patient symptoms. However, patients presenting with transient ischemic attacks are managed with indirect revascularization [3, 6] which have been successful in improving the cerebral reserve. Partial embolization of the fragile areas of the malformations had been successful in treating intractable symptoms [1]. However, there is a paucity of literature on long-term follow-up in pediatric

Fig. 4 **a, b** MRI postcontrast sagittal and axial imaging showed no change in proliferation or caliber of vessels

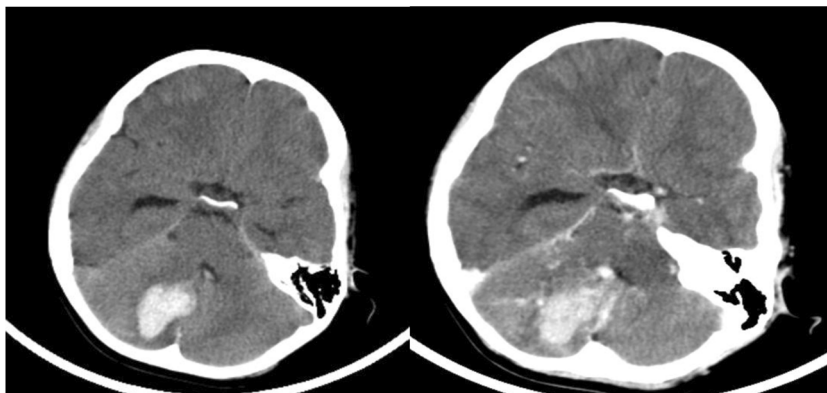


Table 1 Review of all pediatric CPA published till now

Series	Age/ sex	Clinical presentation	Location	Management	Follow-up
Fierstra et al. 2011 [2]	11/F	Seizures	Left temporal	NA	NA
Fierstra et al. 2011 [2]	11/M	Transient neurological deficits	Left frontotemporoparietal	NA	NA
Vargas et al. 2011 [3]	17/F	Headache and behavioral changes	Diffuse network	Symptomatic	NA
Vargas et al. 2011 [3]	14/M	Aphasia	Right cerebral hemisphere	NA	NA
Ellis et al. 2011 [4]	2/F	Left side weakness and foot drop	Right basal ganglia and thalamus	Revascularization (pial synangiosis, encephalomyosynangiosis, dural inversion)	12 months
Khan et al. 2013 [5]	16/F	Headaches TIA	Left parietal region	Symptomatic	NA
Kimiwada et al. 2013 [6]	13/M	Headaches and right upper limb numbness	Left frontal and parietal lobes	Symptomatic	12 months no deficits
Srivastava et al. 2015 [7]	18/M	GTCS	Left frontal	Antiepileptics	1 month (lost to follow-up)
Gold and Crawford et al. 2013 [8]	9/F	Headaches Acute hemiparesis, right homonymous hemianopia	Left cerebral hemisphere	Symptomatic medical management	12 months Similar presentation without any permanent deficits.
Peng Liu 2016 (12)	5/F	Headache and TIA	Bilateral basal ganglia and left frontal lobe	Bilateral EDAS (Indirect revascularization)	12 months Improved completely
Peng Liu 2016 (12)	4/F	Severe headaches	Right frontal parietal and basal ganglia	Symptomatic medical management	NA
Puerta et al. 2017 [9]	8/M	Acute Left hemiparesis	Right cerebral hemisphere and left basal ganglia	Indirect revascularization	No deficit in 1st year, 2 years Permanent left hemiparesis
Current case.	12/M	Trivial trauma and altered sensorium with hemorrhage in imaging	Right cerebellar hemisphere	Symptomatic	18 months No deficits

patients. Large multicenter studies are required for formulating treatment guidelines.

Conclusion

CPAs are rare and distinct vascular entities with an even lesser incidence in the pediatric population. Medical management with observation was considered in most of the patients. Surgical intervention was reserved for patients with TIA and neurological deficits. Long-term studies are required to formulate the guidelines.

Compliance with ethical standards

Conflict of interest The authors declare no conflicts of interest.

Ethics statement Patient parents have agreed for submission of the clinical and imaging details.

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