# Case Report

# Intracranial Hypertension in Unruptured Arteriovenous Malformation: Case Report

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*A report of intracranial hypertension in unruptured cerebral arteriovenous malformation (AVMs) is presented. A 16-years-old obese female presented with a first episode of acute severe headache and papilledema. Non-contrasted computer tomography scan demonstrated no evidence of hemorrhage or hydrocephalus. The magnetic resonance imaging brain shows the unruptured AVMs at right temporal area.*

*The AVMs was urgently embolized by glue. Headache completely disappeared within two weeks. The papilledemagradually improved within two weeks and completely recovered within six weeks. Patients with unruptured AVMs should undergo early intervention, either by means of surgical excision or embolization, so as to avoid the permanent deficits of optic nerve such as decreased visual acuity and impaired visual field and optic atrophy.*

***Keywords:*** *Unruptured cerebral arteriovenous malformation, AVMs, Intracranial hypertension*

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Intracranial hypertension is a state of increased intracranial pressure due to disequilibrium of intracranial components such as brain parenchyma, cerebral blood volume, and amount of cerebrospinal fluids[1,2]. The disequilibrium might be caused by tumor, hydrocephalus, hemorrhage, etc. Clinical manifestations are headache, vomiting, and papilledema, which are commonly found in examination. The underlying conditions have to be immediately discovered and treated in order to avoid permanent morbidity and even mortality. For idiopathic intracranial hypertension, the etiology is unknown. The diagnosis is to rule out all other possible causes. Neuroimaging studies are necessary to exclude intracranial lesion. The treatments include medication, diversion of cerebrospinal fluid or optic nerve fenestration and others which depend on visual function[2].

Cerebral arteriovenous malformations (AVMs) are congenital lesions. Patients remain asymptomatic unless AVMs rupture a common cause of intracranial hypertension. In case of unruptured AVMs, clinical manifestation could be seizure or headache. These may be controlled with medications. Intracranial hypertension in unruptured AVMs is a rare condition but should be taken into consideration as a cause of mentioned symptoms.

A number of 38% to 65% of patients with AVMs have hemorrhage. Approximately, 15% to 35% of AVM ruptured patients display seizure as an initial symptom. Less than 10% of cases are diagnosed with neurological deficits[3,4]. For ruptured AVM, the patient must be treated to avoid rebleeding. Multimodality of treatment such as surgery, endovascular and radiosurgery should be tailored for an individual patient.

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## Case Report

A 16-years-old teenage girl gained 30 kilograms of her weight in the last 12 months, presented with acute headache and vomiting. Physical examination revealed no sign of either neurological deficits or papilledema. Non-contrasted Computerized Tomography scan showed no evidence of hemorrhage or hydrocephalus but there was anisodense area at the right posterior inferior temporal lobe. She was prescribed medication for her symptom. A week later, she returned to the hospital because

of progressive headache and poor vision. Physical examination divulged acute severe papilledema which definitely indicated the high intracranial pressure. Optic nerve function was evaluated. The patient demonstrated a visual acuity of 20/40 and an enlarged blind spot (Fig. 1A&B). A provisional diagnosis was intracranial hypertension and she was admitted to hospital for further investigations and management. Magnetic resonance imaging (MRI) brain revealed nidus of AVMs about 3 cm, arterial feeders were branched of the right middle cerebral artery and right posterior cerebral artery as shown in Fig. 2. The hypertrophic cortical vein drains posteriorly through transverse sinus. There was no evidence of venous sinus thrombosis, bleeding or hydrocephalus. The venous hypertension in these unruptured AVMs was postulated as a cause of intracranial hypertension. Three days after admission, the patient underwent angiography and embolization. Cerebral angiography demonstrated corticosubcortical AVMs at right temporal lobe, with 3 cm nidus size. AVMs were supplied by hypertrophic anterior temporal branch of the right middle cerebral artery (MCA) and branch from right posterior cerebral artery (PCA). Early venous drainages are the right vein of Labbe and the right basal vein of Rosenthal as shown in Fig. 3A&B&C. There was no intranidal or flow related aneurysm. Trans-arterial embolization was performed for three times using glue in one session. Post-embolization angiogram of left internal carotid artery (ICA) revealed 95% obliteration of nidus. There was an incidental arterial rupture during the procedure which was well controlled. The patient's symptom of headache resolved within two weeks after the embolization. Papilledema was improved within two weeks after treatment (Fig. 1C&D) and disappeared in 6 weeks (Fig. 1E&F). One month later, post-embolization angiography demonstrated complete obliteration of the AVMs as shown in Fig. 3D&E&F.

**Discussion**

People with unruptured AVMs mostly remain silent and may present with headache and seizure. The characteristics of headache are similar to migraine and the pain is located in hemicranium or occipital area[3,4]. From A Randommized Unruptured Brain Arteriovenous malformation (ARUBA), found more benefits for medical therapy including lower death and stroke rate are lower than interventional therapy[5]. Thus, the severe headache with papilledema is not common in unruptured AVM. The management is depend on cause because risk of treatment in unruptured AVMs is high. The cause of intracranial hypertension in this case has two assumptions. First is pseudotumor cerebri condition and incidental cerebral AVMs. Second is decompensated venous hypertension in unruptured AVMs. However, the diagnosis of pseudotumor cerebri is not correct because there was an evidence of venous hypertension with its relationship to the hypertrophic vein. Many cases in literature review were managed as pseudotumor cerebri using the medication and CSF diversion, but symptom persisted[10,11,14]. The patients thus required the surgical excision or embolization to resolve critical illness and thus resulting

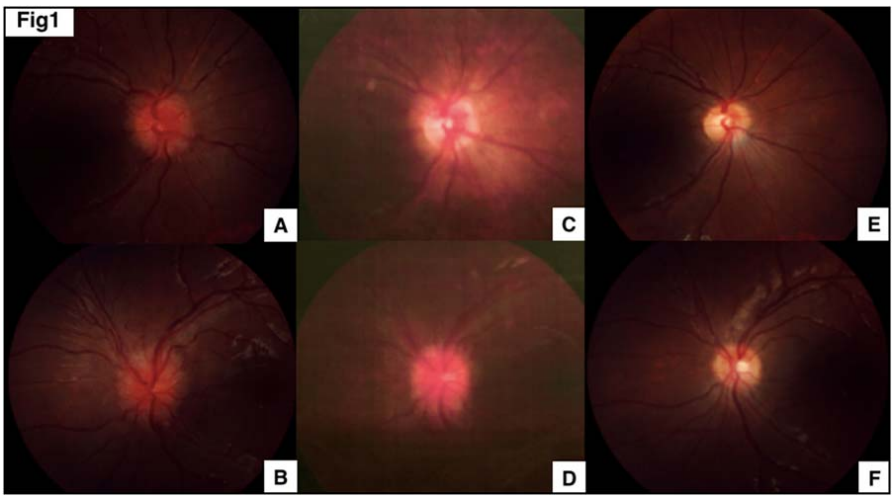
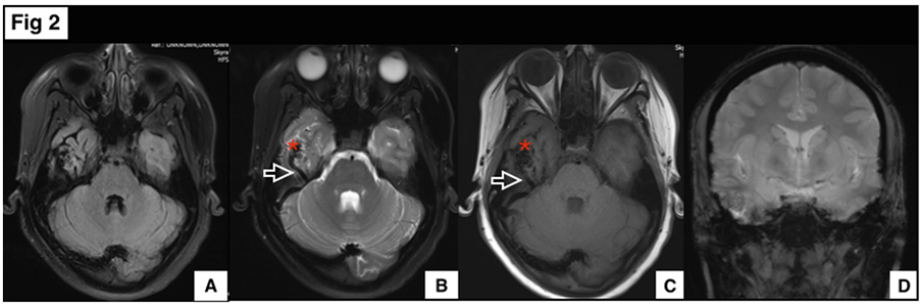
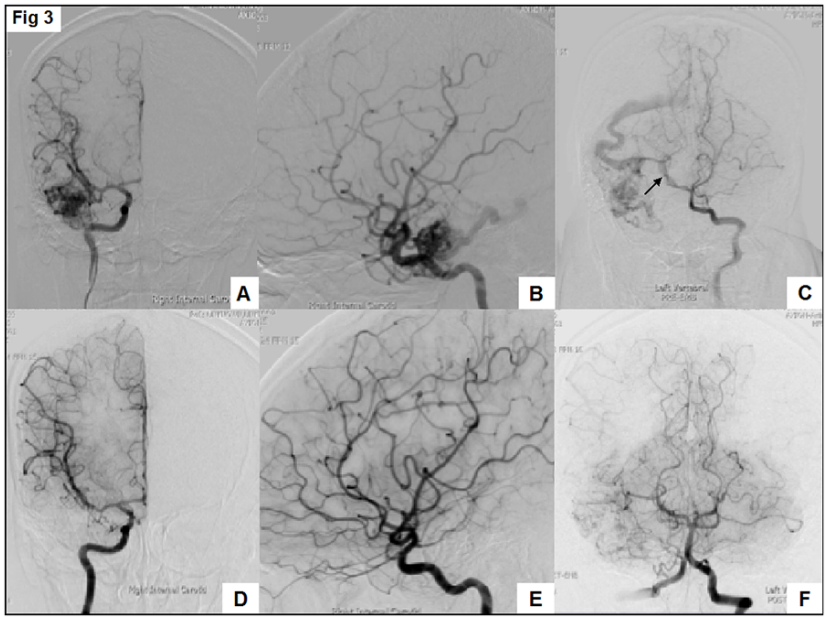



**Fig. 1** Preoperative fundoscopy reveals papilledema of both eyes **(**A&B). Two weeks after embolization, there was an improvement of papilledema (C&D**)**. Optic discs demonstrate complete recovery at six-weeks after treatment(E&F).




* = nidus of AVM, → = hypertrophy of vein of Labbe

**Fig. 2** MRI brain (A&B&C). Demonstrates a nidus of cerebral AVMs at inferior temporal area and hypertrophy of cortical vein (vein of Labbe). There were neither bleeding nor hydrocephalus. A) MRI with axial FLAIR sequence B) axial T2 sequence, C) T1 sequence of axial MRI and D) coronal MRI with gradient sequence.




→ = Posterior cerebral artery segment 2 (P2 segment)

**Fig. 3** Cerebral angiography shows nidus size of 3 cm and an arterial feeder from anterior temporal branch of right middle cerebral artery (A). Figure B is alateral view of right ICA, and an angiogram demonstrates hypertrophic Labbe vein as the draining vein. Another feeder is the branch of posterior cerebral artery. (P2 segment) and its draining vein is a basal vein of Rosenthal (C). Post embolization of cerebral angiography reveals complete obliteration of AVM. Right ICA injection in AP view (D), in lateral view (E) and left vertebral artery injection in AP view (F).

in appropriate treatment which often prolong time to improve papilledema and resulting in the complications of optic nerve such as decreased visual acuity, visual field defect as well as optic nerve atrophy. Management of intracranial hypertension in unruptured AVM should be prompt to avoid such complications. The type of treatment depends on risk and benefit.

The pathogenesis of intracranial hypertension in unruptured AVM is uncertain. There are two hypotheses. The first one is a massive overload of the cerebral venous outflow resulting in an increase cerebral blood volume and impairment of cerebrospinal fluid absorption. The second one is a high venous pressure with partial venous obstruction[10,15].

Table 1 summarizes the data of unruptured AVM. Cerebral unruptured AVM were mostly located

**Table 1.** Summary data of unruptured cerebral AVM with intracranial hypertension[6-15]

| Literature | Onset | Age | Gender | Location | Feeder | Draining vein | Treatment | Outcomes/complications |
|---|---|---|---|---|---|---|---|---|
| Kosary et al, 1973 | 1½ year | 25 | M | Rt parietal | Rt ACA | SSS | Surgery excision | 2-3 days to 1 D |
| Weisberg et al, 1977 | 15→4 year | 40 | M | Rt PO compressed lat. ventricle | Rt ATCA ACA PCA | - | LP shunt | Not mention |
| Vassilouthis, 1979 | 5 month→3 month | 11 | M | Rt Postrior frontal | Rt ACA | SSS, ISS | Surgical excision | Resolve in 1 year |
| Schiffer et al, 1984 | 10 year→2 month | 31 | F | Rt midrolandic region | Rt MCA ACA | SSS, ICV | Surgical excision | Not mention in papilledema. L upper extremity paralysis → resolve in 4 month |
| Barrow, 1988 | 2-3 year | 21 | F | Rt frontal | Rt ACA | SSS | Medication → LP shunt → excision | Resolve in 2 month |
| Barrow, 1988 | 1 year | 33 | F | Lt Parietal | Lt MCA ACA PCA | SSS Vein of galen | Medication → surgery | Post op brain edema and neuro deficit, resolve in 3 month |
| | 5-6 year | 19 | F | Rt PO | Rt MCA ACA PCA | SSS | Embolized → surgery | Resolve in 2 month |
| | 4 week→2 week | 21 | F | Rt parietal | Rt MCA | SSS | Medication → surgery 2 month after onset | Resolve in 2 month but optic atrophy both |
| Chimowitz et al, 1990 | 18 year (papilledema 2 year) | 32 | F | Lt frontal | Lt MCA ACA | SSS | Medication → Proton beam irradiation | Unchanged VA & VF |
| | 3 months | 28 | M | Rt parietal | Rt MCA ACA | SS but stenosis at Rt. TS | Embolization → excision | Resolve in 1 month → normal in 4 month |
| | 32 year | 44 | F | Rt PO | Rt MCA PCA | SSS | Medication | Resolve in 2 week but continued headache |
| | 7 hour | 24 | M | Rt PO | Rt MCA PCA ACA | SSS stenosis | Embolization → resection | Resolve in 1 m but residual bilateral optic atrophy |

ACA = Anterior Cerebral Artery, F = Female, hr = hour, ICV = Internal Cerebral Vein, ISS = Inferior Sagittal sinus, Lt = Left, LP shunt = Lumboperitoneal shunt, M = Male, MCA = Middle Cerebral Artery, m = month, PO = Parietooccipital lobe, PCA = Posterior Cerebral artery, Rt = Right, SSS = Superior Sagittal Sinus, TP = Temporoparietal lobe, TS = Transverse Sinus, VA= Visual acuity, VF = Visual field

**Table 1.** Cont.

| Literature | Onset | Age | Gender | Location | Feeder | Draining vein | Treatment | Outcomes/complications |
|---|---|---|---|---|---|---|---|---|
| Rosenfeld et al, 1991 | 1 week | 32 | M | Rt TP | Rt MCA Ant.choricdal | SSSTS | Excision | VF subside at discharge |
| Kamite Y, 1994 | ? | 14 | M | Lt. Temporal | Lt. MCA PCA | TS | Excision | Subside? |
| David et al, 1995 | 1 year | 29 | M | Lt TP | Lt MCA | - | Optic n. fenestratation | Slightly improved |
| Vorstman et al, 2002 | 2 week | 9 | F | Lt Parietal | Lt PCA | SSS | Embolization with onyx | Resolve in 12 days but Optic atrophy (decrease VA, loss color vision) |
| Present report, 2016 | 1 week | 16 | F | Rt Temporal | Rt MCA PCA | TS basal vein of Rosenthal | Embolization with glue | Resolve in 2 weeks. Normal in 1½ month |

ACA = Anterior Cerebral Artery, F = Female, hr = hour, ICV = Internal Cerebral Vein, ISS = Inferior Sagittal sinus, Lt = Left, LP shunt = Lumboperitoneal shunt, M = Male, MCA = Middle Cerebral Artery, m = month, PO = Parietooccipital lobe, PCA = Posterior Cerebral artery, Rt = Right, SSS = Superior Sagittal Sinus, TP = Temporoparietal lobe, TS = Transverse Sinus, VA= Visual acuity, VF = Visual field

in the corticosubcortical area but not deep structure or posterior fossa. Feeders were usually branches of either middle cerebral artery or posterior cerebral artery and nidus sizes varied. All draining veins had been described as "hypertrophic of cortical vein" and few cases had deep drainage. There were no intranidal or flow related aneurysm. The patient's age varied from 9 to 40 years, which are usually found in pediatric and young adults[6-15]. Most of the patients had a previous symptom of seizure or headache, and developed intracranial hypertension. Whilst the case No. 16 and our patient have intracranial hypertension as a first symptom. Risk of rupture in cerebral AVM are nidus size less than 3 cm, locating deep part or posterior fossa, draining in deep system and intranidal aneurysm or flow related aneurysm[3,4]. From literatures review, all of reported unruptured AVM have low risk of rupture[6-15]. Nowadays, unruptured cerebral AVMs with symptoms such as headache or seizure are normally treated by medication due to high risk of interventions[5]. Intracranial hypertension in unruptured AVM should be urgently treated to decrease morbidity and mortality, even in low risk of ruptured AVM. Thus, early detection and close monitoring of the intracranial hypertension must be planned in unruptured AVMs.

## Conclusion

Unruptured cerebral AVMs can be presented with intracranial hypertension. This condition should be discovered early and treated in order to reduce mortality and permanent morbidity.

## What is already known on the topic?

State of raised intracranial pressure is common in ruptured cerebral AVMs. However, in unruptured cerebral AVMs, this condition is rare. There are 16 case reports in nine literatures. From the review, the management was as the same as pseudotumor cerebri such as treating with acetazolamide, or CSF diversion. Whether patient's symptoms improve on medication or not it often leads to the definite treatments. The definite treatments include surgical removal and/or endovascular approaches.

## What this study adds?

Increase intracranial pressure with unruptured cerebral AVMs are a rare condition but can be found. This study propose the pathogenesis of venous hypertension using evidence of enlarged cortical venous drainage from angiographic finding. This condition is an urgent situation. Any delay in management could lead to patient morbidity and mortality.

## Potential conflicts of interest

None.

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## *รายงานกรณีศึกษาภาวะความดันสูงในกะโหลกในโรคหลอดเลือดขอดที่ไม่แตก*

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*รายงานกรณีศึกษาผู้ป่วยที่มีสภาวะความดันในกะโหลกสูงในโรคเส้นเลือดขอดในสมอง ซึ่งยังไม่มีการแตกของหลอดเลือดโดยพบผู้ป่วยหญิงอายุ 16 ปี น้ำหนักเกินมาตรฐานมีอาการปวดศีรษะเฉียบพลันเป็นครั้งแรก และตรวจพบจอประสาทตาบวม หลังตรวจด้วยเอกซเรย์คอมพิวเตอร์ไม่พบเลือดออกหรือภาวะน้ำคั่งในโพรงสมอง ผลเอ็กซเรย์คลื่นแม่เหล็กไฟฟ้าพบเส้นเลือดขอดในสมองบริเวณสมองส่วนกลีบขมับด้านขวา (temperal lobe) ผู้ป่วยได้รับการรักษาด้วยการอุดกาวแบบเร่งด่วน อาการปวดศีรษะลดลงจนเกือบปกติใน 2 สัปดาห์ส่วนจอประสาทตาเริ่มลดบวมตั้งแต่ 2 สัปดาห์และหายสนิทใน 6 สัปดาห์*

*จากการกรณีศึกษาผู้ป่วยที่มีภาวะเส้นเลือดขอดในสมองแม้นยังไม่มีเลือดออกควรได้รับการรักษาอย่างรวดเร็ว ไม่ว่าจะด้วยวิธีการผ่าตัดหรือการอุดหลอดเลือดด้วยกาว เพื่อลดความพิการแบบถาวรของเส้นประสาทคู่ที่ 2 เช่น ความชัดของสายตาลดลง, ลานสายตาผิดปกติและเส้นประสาทฝ่อ*