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AVM presenting with seizure and hemorrhage

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Abstract

Background: Cerebral arteriovenous malformations (AVMs) have been long thought to be a congenital anomaly of vasculogenesis in which arteries and veins form direct communications forming a vascular nidus without an intervening capillary bed or neural tissue. Scattered case reports have described that AVMs may form de novo suggesting that they can become an acquired lesion.

Case report: Our case, a 32-year-old, right-handed, male presented with new onset seizure with history of previous episode of intracerebral hematoma which was confirmed by CT scan of brain for which surgical evacuation was performed 8 years back. Post-operative recovery was uneventful and no episode of seizure was noticed at that time. Five years later, he developed new onset of seizure which was partially controlled by conventional Antiepileptic drugs.

Conclusion: This case was initially a case of congenital small cerebral AVM that bleeds and subsequently developed new onset of seizure which became intractable. It indicates that cerebral AVM is a congenital vascular lesion.

Keywords: arterio-venous malformation, seizure, congenital, de novo, surgical excision

Introduction

AVM is a vascular malformation characterized by arterio-venous shunt through a collection of tortuous vessels without an intervening capillary bed.[1] Cerebral AVMs are presented to be congenital vascular lesions that results from abnormal vascular communication forming during embryogenesis[2]. The pathogenesis of AVM is still unknown which is traditionally thought to be congenital in origin whereas some have suggested a more dynamic developmental process [1]. The prevalence of cerebral AVM is 0.06 to 0.14% with an incidence of 1 in 100000 population. It is common (80%) at the end of the 4th decade with only 20% occurrence before 20 years of age [2]. In majority of cases AVMs remain asymptomatic and silent till it ruptures. It can present with intracerebral hemorrhage known as hemorrhagic stroke, seizure, moderate to severe headache, loss of vision, aphasia/dysphasia or motor weakness which is usually related to mass effect or ischemic still phenomenon [3].

A bleeding cerebral AVM is a life-threatening condition and require medical attention. The prognosis depends on whether the AVM is discovered before or after rupture. More than 90% of cases who bleed survive the event. Most AVMs are delineated with CT scan or MRI of brain but for any type of treatment for AVM, an angiogram may be needed for better evaluation and to identify the type of AVM.

Case Report

A 32-year-old right-handed, normotensive, gentleman presented with repeated episodes of convulsion for 3 years which was tonic-clonic in nature starting from right upper limb with secondary generalization (complex partial seizure). The convulsion lasted for 2 to 3 minutes with no pre-ictal aura but post ictal headache and weakness for few hours. Initially the attacks occurred one to two times per month. Gradually the frequency increased to four to five times per month despite usual antiepileptic drugs. The patient also suffered from recurrent attacks of severe left hemi cranial headache radiating to left jaw associated with conjunctival injection, lacrimation and nasal congestion once to twice a day It was not associated with nausea or vomiting. There was no history of visual impairment, blurring of vision or double vision.

The patient had a history of spontaneous left frontal intracerebral hemorrhage 8 years back for which he underwent surgical intervention and recovered. After one month of his operation, he went back to his usual job. On examination- the patient was conscious and alert but, Anxious & ill looking. The patient had a healthy, well-healed, non-tender curvilinear scar on his left frontal region. Nervous system examination revealed, Mild cognitive impairment with Mini Mental State Examination (MMSE) score of 24/30 with normal cranial nerve function. Motor and sensory examination of limbs also revealed normal functionality.

Based on history and clinical findings, a cerebro-vascular pathology was suspected and a brain MRI and cerebral DSA were obtained subsequently. MRI of brain with and without contrast revealed an irregular mixed intensity lesion in left frontal region with perilesional edema. T_2WI showed multiple flow voids and inhomogeneous enhancement in the center of the lesion in post contrast film. Cerebral digital subtraction angiography (DSA) showed an arteriovenous malformation (AVM) in the left frontal region being feeded by both left middle cerebral artery and anterior cerebral

artery with a single large venous drainage to superior sagital sinus.

The patient was diagnosed as a case of left frontal cortical AVM. He underwent craniotomy and complete excision of AVM was performed. Feeding arteries were cauterized meticulously with low voltage bipolar diathermy. The nidus of the AVM was properly skeletonized and excised. Hemostasis was ensured and wound was closed in layers. Histopathological diagnosis was also an arteriovenous malformation. Post-operative events were uneventful. Patient had a complete recovery with no convulsion and no focal deficit after surgery.

Pre-Operative MRI

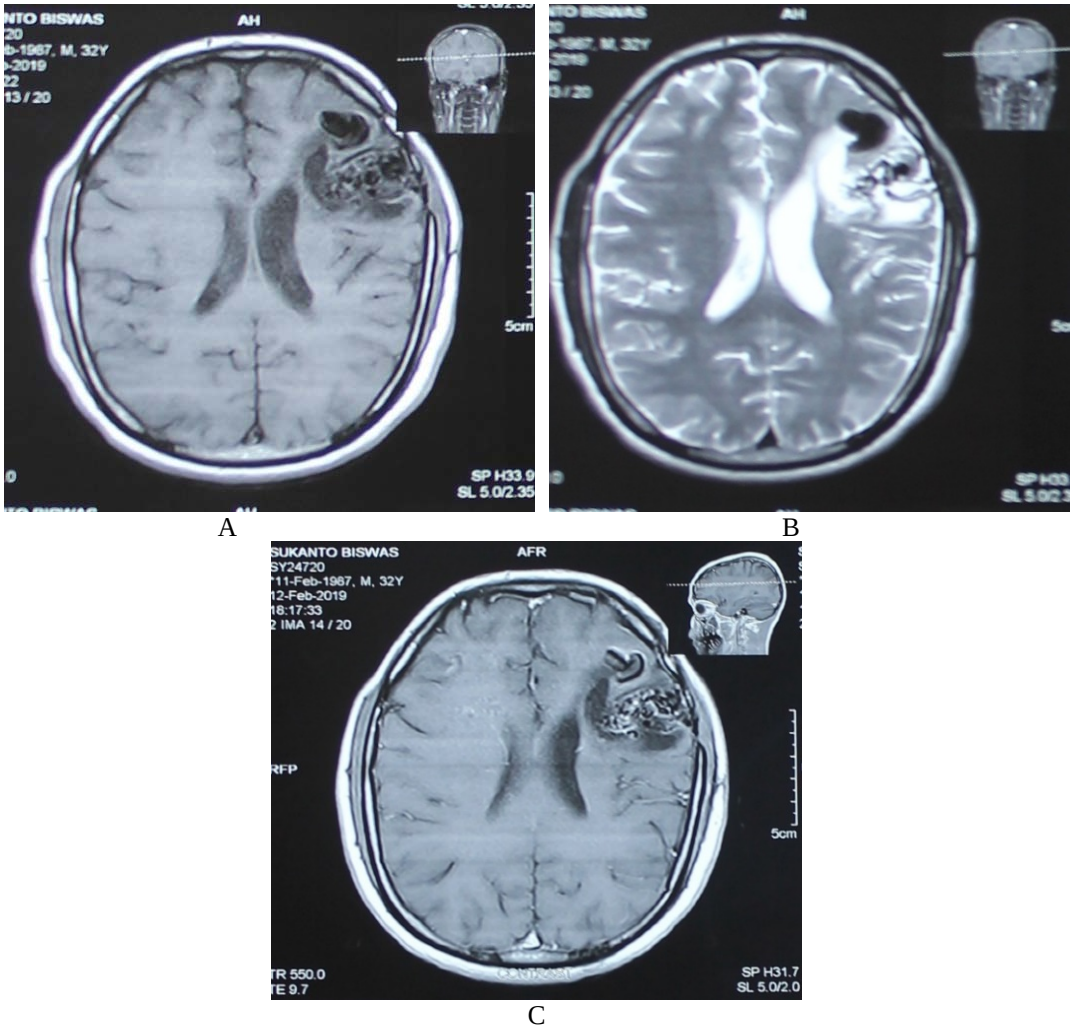


Fig 1: T1WI (A) T2WI (B) and post contrast film (C) showing a left frontal cortical AVM

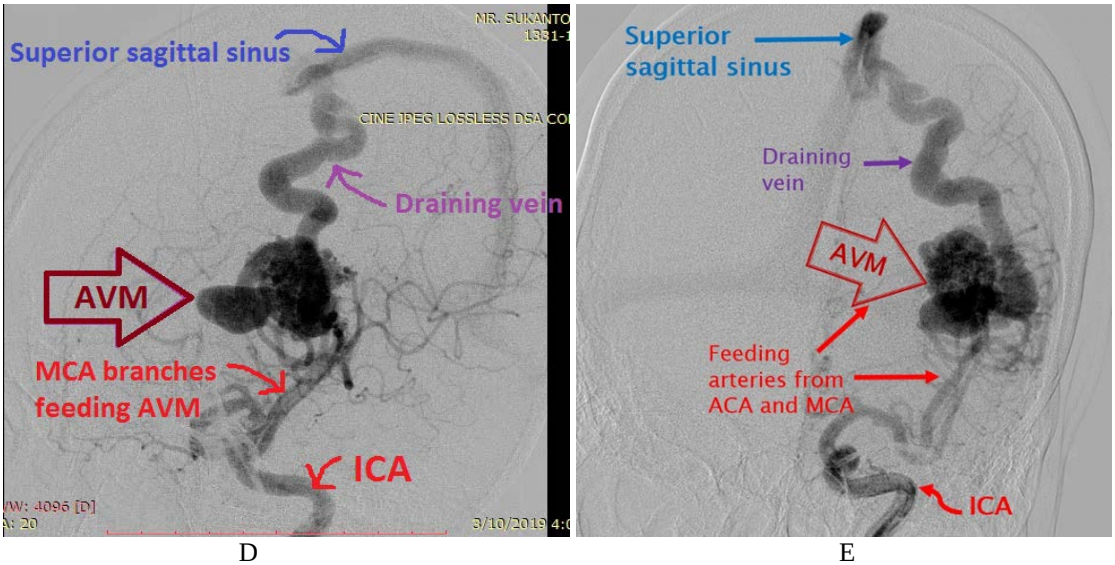


Fig 2: DSA left oblique view (D) and AP view (E) showing a left frontal cortical AVM

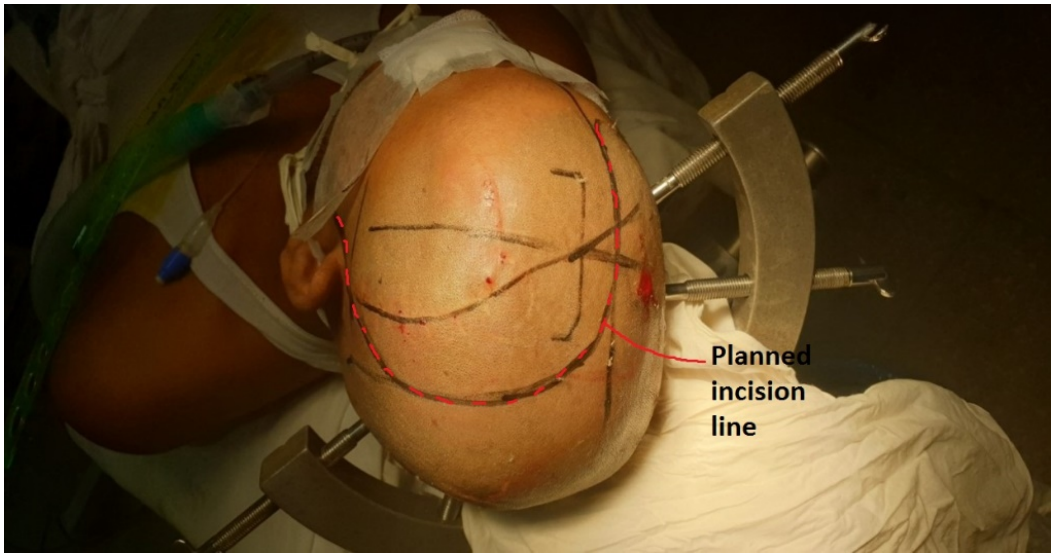
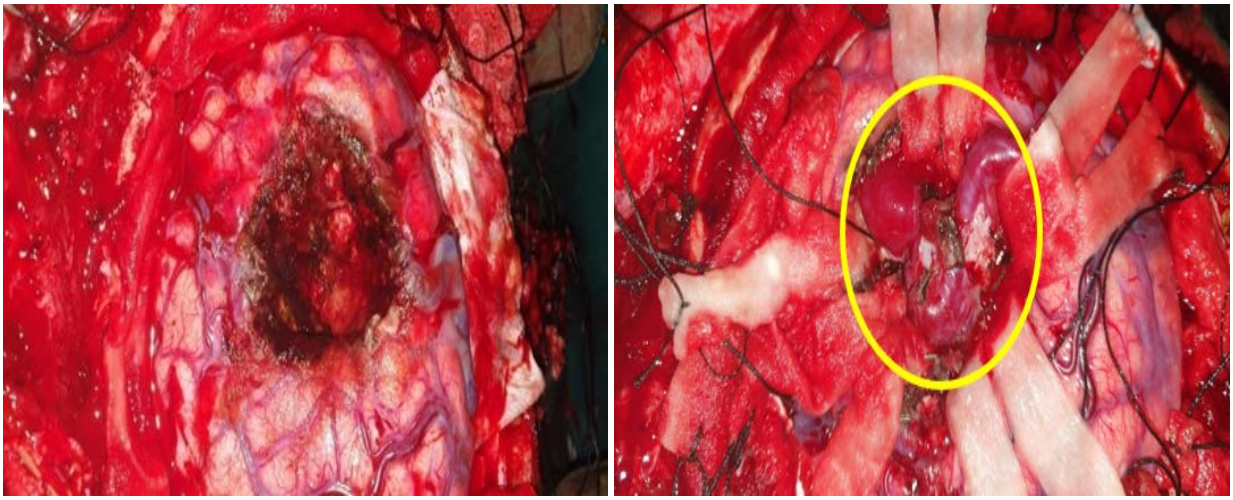


Fig 3: (F) Patient position and scalp marking before surgery.



G H

Fig 4: Per-operative view of AVM before (G) and after (H) surgical excision

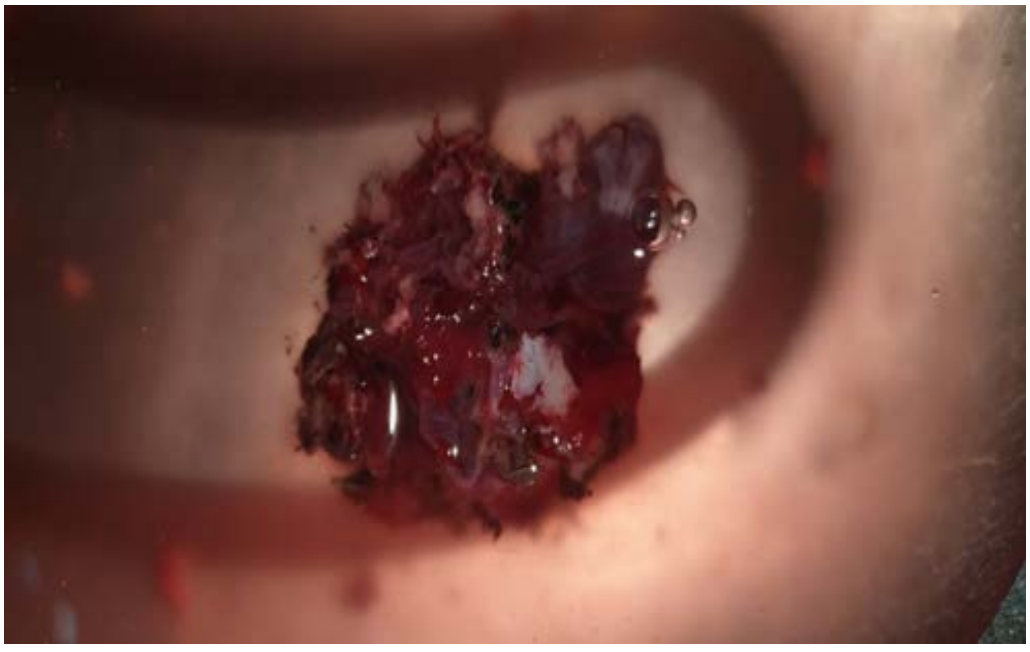


Fig 5: The AVM itself after complete excision

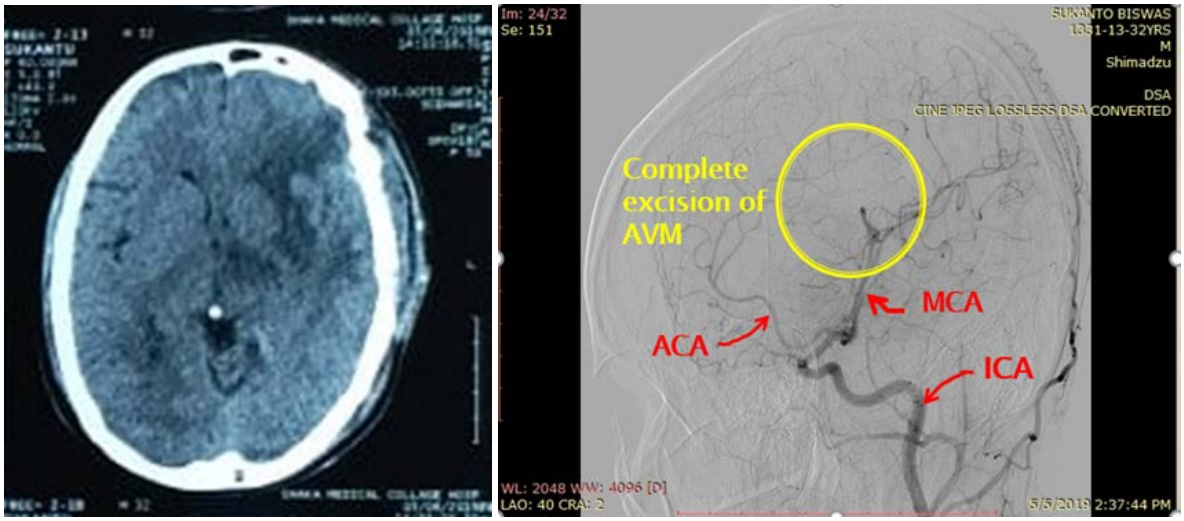


Fig 6: Post-operative CT scan of brain (J) and cerebral DSA (K) showing complete removal of the AVM.

Discussion

Cerebral arteriovenous malformations (AVMs) are abnormal vascular connections that form between arteries and veins, without an intervening capillary bed or neuronal tissues [3, 13, 14]. The incidence of cerebral AVMs in the general population is less than 1%. AVMs may be clinically silent or present with hemorrhages, often in the second or third decade of life.[4] It has been suggested that cerebral AVMs are primarily congenital, originating at or before the 40- to 80-mm embryo length stage and may be related to a primary abnormality of primordial capillary or venous formation.[5] AVMs appear to be dynamic lesions that have the potential to develop and grow with time. It is possible that acquired insults can provide the catalyst for the formation of AVMs later in life [1]. However, the dynamic nature of cerebral AVMs is attested to by multiple recent case reports demonstrating spontaneous progression and resolution [10, 11, 12]. The symptomatic presentation of AVMs in adults before the age of 40, in addition to the *de novo* AVMs reported in children, support the concept of the temporal vulnerability of vascular elements to a physiologic or environmental trigger [1]. These triggers, which can be mechanical, inflammatory, thrombogenic, ischemic/ hypoxic, or hormonal, generally lead to hemodynamic stress [1, 6]. Disturbances of the venous drainage system may contribute to the formation of cerebral AVMs. Venous stenosis, occlusion, or agenesis during embryology or chronic venous hypertension during childhood and adulthood can result in tissue hypoxia and drive the impetus for angiogenesis [3]. Genetic etiologies for cerebral AVMs are also currently being explored. AVMs have been known to occur in the context of certain genetic disorders, such as hereditary hemorrhagic telangiectasia (HHT), Wyburn–Mason syndrome, and Sturge–Weber syndrome [7].

Current study diagnosed the AVM by Magnetic resonance imaging (MRI), Computed Tomography (CT) scan, Digital subtraction angiography (DSA) of brain. DSA is more sensitive to diagnose AVM and able to determine the location well than MRI. CTA can diagnose the AVM well. Simple CT can scan able to detect hemorrhage easily. Magnetic resonance Angiogram (MRA) revealed best features of AVM [8]. In reported cases of de novo AVMs, lesions usually developed 8 years (3–17) after initial imaging, with a mean age at diagnosis of approximately 18 years (6–32). In contrast, the mean age of patients diagnosed with congenital AVMs is 33 years [9]. In our patient, the lesion developed 8 years after the initial MRI. Initial scans revealed no vascular abnormality. Subsequent imaging revealed lesions specific enough for the diagnosis of a de novo AVM.

Conclusion

This rare case report on congenital AVM in adult was initially presented as intracerebral hematoma (ICH) and treated surgically. Subsequently, he developed intractable seizure with cognitive impairment which was diagnosed as AVM and managed by excision of AVM. So, during managing a case of ICH in young patients, an underlying cerebral AVM should be kept in mind and an MRI of brain and cerebral DSA should be done for confirmation of diagnosis before surgical intervention.

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