ABSTRACT

Aneurysms are categorized as intranidal, flow-related, or unrelated to the AVM. Aneurysms were designated intranidal if they filled early during angiography, before substantial venous filling had occurred, and if they were localized within the boundaries of the AVM nidus. Though the incidence of aneurysms associated with AVMs reported in the literature has varied widely, but intranidal aneurysm are rare ones. Here, we report a case of cerebral AVM with intranidal aneurysm.

KEY WORDS:  AVM(Arterio-Venous Malformation), Aneurysm

INTRODUCTION

The association of arteriovenous malformations (AVMs) with intracranial aneurysms has been well documented in numerous reports.\(^1\)\(^,\)\(^2\) It is generally believed that aneurysms lead to a higher risk of hemorrhage as the initial presentation and also as part of the clinical course and natural history of patients with this combination of lesions.\(^3\)\(^,\)\(^4\) Aneurysms in a patient with an AVM may occur on unrelated vessels and be found coincidentally or they may be associated with the AVM on a pathophysiological basis due to an underlying vascular defect or as a result of the dynamic interaction between hemodynamic stimuli, vasoactive substances, and locally generated growth factors leading to structural and functional alterations through the process of vascular remodeling.\(^5\) There have been several attempts to categorize AVM-related aneurysms,\(^6\) but a widely accepted system of classification based on their anatomical and pathophysiological relationship to the AVM has yet to be developed and validated. Most publications have dealt with descriptive aspects of these associated lesions, whereas less emphasis has been placed on appropriate surgical and endovascular management priorities. An imperfect understanding of the relationship between AVMs and associated aneurysms has led to widely varying treatment strategies. Concern that abrupt elimination of an AVM might put aneurysms located along feeding arteries at immediate risk of distention and subsequent rupture has led some to recommend treating the aneurysm before the AVM.\(^2\) Alternatively, the reduction of flow through feeding arteries following AVM elimination has prompted others to recommend elimination of the AVM first, with the expectation that resulting hemodynamic alterations may lead to diminution or complete regression of related aneurysms.\(^7\)

CASE REPORT

Nine year old Right handed school girl from western part of Nepal was brought to our casualty with the chief complaints of sudden onset of severe headache associated with loss of consciousness 1 day prior to presentation. She also had multiple episodes of vomiting and 1 episode of seizure. There were no history of trauma, fever in the past. There were no significant past or familial history.

On examination, in casualty, all the vital parameters were within normal limits. Her systemic examinations also didn’t revealed any abnormalities. On neurological examination, her Glasgow Coma Scale was 15/15. All the cranial nerve examinations including both fundi were also normal. There were no motor or sensory deficit. All the reflexes were also normal.

She was admitted and investigated. CT Scan brain showed Left occipital hyperdense lesion consistent withintracerebralhaematoma due to vascular insult.\(^1\)\(^,\)\(^2\)
Subsequent CT angiogram showed AVM of size 1.8 cm in left Occipital area supplied by occipital branch of the left PCA and drained via superficial cortical vein into the left transverse sinus with surrounding lobar haematoma in the occipital lobe and intraventricular blood in the left lateral ventricle, associated with intranidal aneurysm of size 5x4 mm and neck of the aneurysm was 2.2 mm.

Figure 1.

Figure 2.

She was planned for elective excision of the aneurismal AVM. Left postero-parieto-occipital box flap was raised. And intra-operatively, she had tense dura with subcortical haematoma of about 20 ml. There were tangle of abnormal vessels with aneurysm in the centre. The lesion was about 2 cm of size, fed by occipital branch of the Lt PCA and drained via superficial cortical vein into the Lt transverse sinus.

Figure 3.

Figure 4.

Total excision of the aneurysmal AVM was carried out. Her post operative days were uneventful and she was discharged on 5th post operative day.

Post operative CT cerebral angiogram showed no evidence of vascular lesion and there was gliotic area in left occipital lobe.

Figure 5.
Histopathological report could have confirmed the diagnosis but unfortunately it was not sent.

**DISCUSSION**

Aneurysms are categorized as intranidal, flow-related, or unrelated to the AVM. Aneurysms were designated intranidal if they filled early during angiography, before substantial venous filling had occurred, and if they were localized within the boundaries of the AVM nidus. Simple arterial ectasias, infundibulae, venous pouches, and variceal dilations were excluded. Saccular aneurysms arising along the course of arteries that eventually supplied the AVM were classified as flow related. Flow-related aneurysms were subclassified as proximal if they were located on the supraclinoid internal carotid artery (ICA), the circle of Willis, the middle cerebral artery (MCA) up to and including the primary bifurcation, the anterior cerebral artery (ACA) up to and including the anterior communicating artery (ACoA), or the vertebrobasilar trunk. All flow-related aneurysms beyond these locations were subclassified as distal. Aneurysms that occurred on arteries with no supply to the AVM were classified as unrelated. Although the anatomical and clinical features of aneurysms associated with AVMs have been the focus of several studies, little is understood about their pathogenesis. It is generally accepted that hemodynamic factors and a hyperdynamic circulatory state account, at least in part, for their occurrence. This theory is supported by the observation that the incidence and distribution of aneurysms on feeding arteries supplying AVMs are greatly in excess of what would be expected in the absence of the AVM and that some aneurysms regress after AVM removal. The vasculature is a dynamic structure, capable of sensing biochemical and hemodynamic alterations and of changing itself through the local production of mediators that influence both structure and function. “high-flow angiopathy” consisting of intimal thickening and destruction, elastic degeneration, irregular thickening and thinning of vessel walls, and alteration of the muscular layer has been described in a rabbit model of a high-flow arteriovenous shunt. However, given that flow-related and intranidal aneurysms occur in association with AVMs of all sizes and flow rates, yet only in a minority of all AVMs, their development must be the result of complex interactions of host-specific and hemodynamic factors.

The flow-related saccular aneurysms associated with cerebral AVMs cannot be distinguished on the basis of their angiographic or histological features from those aneurysms that occur in the absence of any other vascular malformation. The question may be raised as to whether intranidal aneurysms or distal flow-related aneurysms located near the AVM nidus identified on angiography following a hemorrhage represent true aneurysms that were present prior to bleeding or “pseudoaneurysms” developing from the rupture of a thin-walled vessel. This issue is of some importance, because these aneurysms are believed to increase the risk of hemorrhage from AVMs significantly, and their identification may lead to a greater sense of urgency with respect to AVM treatment. However, if a substantial proportion of intranidal aneurysms are really pseudoaneurysms that arise as a result of hemorrhage, then it is inappropriate to extrapolate a high risk of bleeding to intranidal aneurysms found on AVMs that have not bled.

Pseudoaneurysms may be suspected if they appear on angiography or magnetic resonance imaging as vascular cavities of irregular shape within or at the margins of a hematoma. These aneurysms can only be confirmed as pseudoaneurysms on the basis of histological examination or if a comparison with previous vascular examinations confirms the aneurysm as a new angioarchitectural feature. This acquired nature secondary to a hemorrhagic event is pathognomonic. Garcia-Monaco, et al. reported finding pseudoaneurysms on angiography in 15 cases (8%) in a population of 189 patients with AVMs. None of the pseudoaneurysms was confirmed histologically. Five were treated with embolization, one was treated surgically, and eight of the remaining nine pseudoaneurysms resolved on follow-up angiography. Marks and colleagues reported on 15 patients with intranidal aneurysms, all of whom had a history of bleeding. Three patients underwent surgical excision, and in two cases the aneurysms could be located in the pathological specimens obtained. Histological evaluation demonstrated these aneurysms as thin-walled vascular structures rather than pseudoaneurysms secondary to previous hemorrhage.

The incidence of aneurysms associated with AVMs reported in the literature has varied widely, with most series reporting between 10% and 20%, up to as high as 58%. Lack of a uniform system of classification...
and nomenclature has prevented direct comparison between series. Anderson and Blackwood\textsuperscript{1} reported autopsy findings in a series of nine patients with AVMs. In five cases (55.6\%) there were associated aneurysms, all of which were localized to the circle of Willis or the MCA. No intranidal aneurysms were described.

**CONCLUSION**

Though the incidence of aneurysms associated with AVMs reported in the literature has varied widely, but intranidal aneurysm are rare ones.

**REFERENCE**