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A rare case of retinal arteriovenous malformation associated with conjunctival telangiectasia and cerebral arteriovenous malformation

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Abstract

Arterio-venous communications of the retina are rare congenital anomalies that are direct communications between artery and vein without intervening capillary bed. Retinal AVM has a widely variable spectrum of presentation. Retinal arteriovenous malformation (AVM) is known to be associated with intracranial AVM. Retinal AVM may be associated with central and branch retinal vein occlusion, neovascular glaucoma, optic atrophy, retinal exudation, and vitreous hemorrhage. Racemose aneurysm of the retina is the rarest, which may be associated with similar AVM in the ipsilateral part of the brain. Coexistence of AVM in the retina and the brain is considered to be one of the phakomatosis. A 27 year old male presented with hemiparesis, conjunctival telangiectasia and retinal AVM. Fundus examination showed convoluted, dilated, tortuous retinal vessels extending from the optic disc to the periphery of retina in left eye. Massively dilated vessels were noted in the superotemporal and inferotemporal quadrants. Computed Tomography of brain revealed linear hypodense area with gliosis in the left parietotemporal region. MRI showed evidence of AVM with multiple tiny tortuous feeder vessels from the left middle cerebral artery, forming a sparse nidus within the gliotic area. Hemiplegia occurs secondary to subarachnoid or intracranial hemorrhage, caused by the AVM. It is imperative that a detailed fundus examination should be done in any case of conjunctival telangiectasia. Imaging of the brain is advisable in a case of retinal AVM to rule out cerebral AVM. Retinal AVM may result in visual loss and requires vigilant follow up.

Keywords: Conjunctival telangiectasia, Intracranial AVM, Retinal arteriovenous malformation (AVM)

Introduction

Arterio-venous communications of the retina are rare congenital anomalies that are direct communication between an artery and a vein without an intervening capillary bed.(1) Retinal arteriovenous malformation (AVM) is known to be associated with intracranial AVM. We discuss a 27 year old male who presented with hemiparesis, conjunctival telangiectasia and retinal AVM. Neurological imaging revealed evidence of infarction in the parietooccipital region signifying a nidus of AV malformation. It is imperative that a dilated fundus examination should be done in any case of conjunctival telangiectasia to look for retinal AVM. Imaging of the brain is advisable in a case of retinal AVM to rule out cerebral AVM, which may cause significant neurological deficits.

Case Report

A 27 year old male presented with diminished vision in left eye for three months. He had right sided weakness following a cerebrovascular accident one year back. There were no seizure episodes. Anterior segment examination of the left eye revealed dilated tortuous vessels on the temporal aspect of bulbar conjunctiva suggesting conjunctival telangiectasia. (Fig. 1) There was no facial abnormality, portwine stain or palatal haemangiomas as seen in Sturge Weber syndrome. There were no features of tuberous sclerosis or Wyburn Mason syndrome. There was no significant family history.

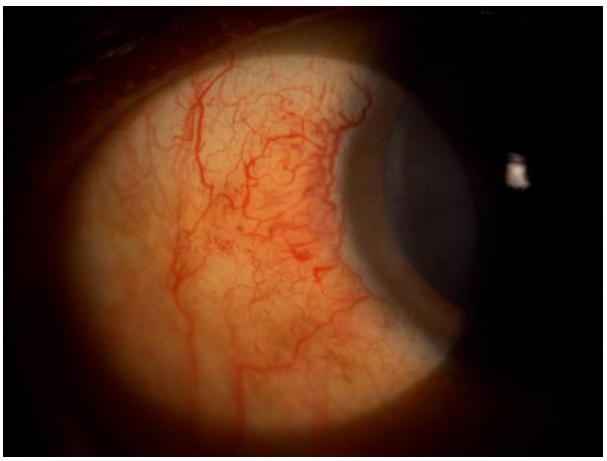

Fig. 1: Conjunctival telangiectasia of left eye

Fundus examination showed convoluted, dilated, tortuous retinal vessels extending from the optic disc to the periphery of retina in left eye. Massively dilated vessels were noted in the superotemporal and inferotemporal quadrants. (Fig. 2) There was no mass lesion in the periphery on indirect ophthalmoscopy. There was no evidence of retinal capillary haemangioma, choroidal haemangioma or racemose angioma. The optic disc and macula were normal. Visual field analysis was normal in both eyes. Fundus Fluorescein Angiography showed rapid filling of vessels with no leak in late phases. (Fig. 3) There was

no intervening capillary network. The right eye was remarkably normal.

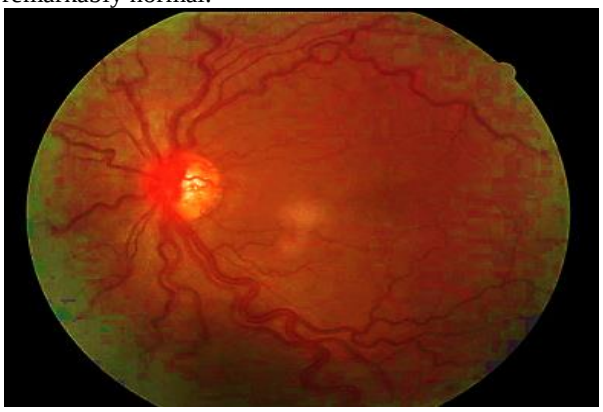

Fig. 2: Massively dilated and tortuous vessels (arteries and veins) in left eye

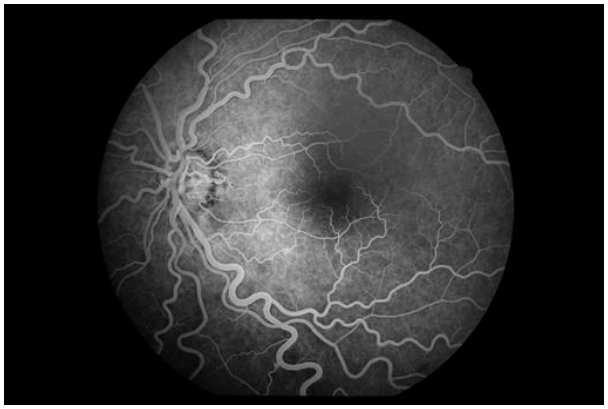

Fig. 3: Fundus Fluorescein Angiography (FFA) showing rapid filling of vessels with no leakage

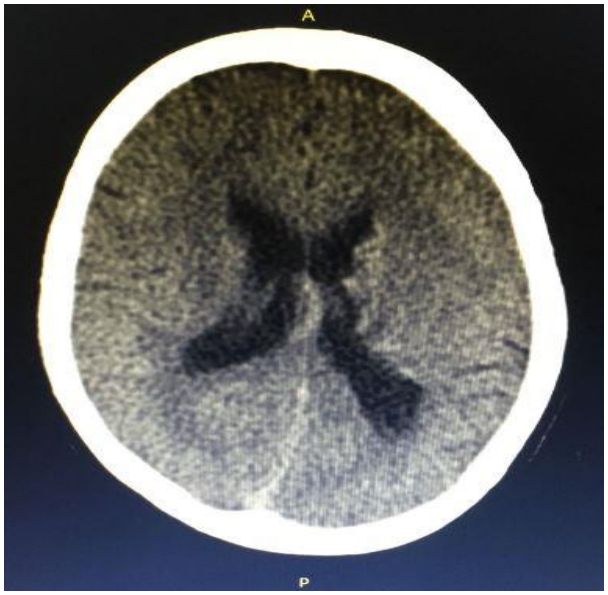

Fig. 4: CT brain showing linear hypodense area with gliotic changes in left parietotemporal region suggesting old AVM

Ultrasound abdomen was normal. CT brain revealed linear hypodense area with gliotic changes in the subcortical aspect of left parietotemporal region, extending from the occipital horn to the insular cortex. There were prominent vascular markings in the subependymal region of left lateral ventricle. There was no parenchymal mass lesion or calcification. (Fig. 4) MRI brain revealed evidence of parenchymal AVM with multiple tiny tortuous feeder vessels from the left middle cerebral artery, forming a sparse nidus within the gliotic area. Magnetic Resonance Arteriography and Venography confirmed the above findings.

The patient was explained the implications of the condition, need for immediate review in case of visual or neurological symptoms, and regular follow up.

Discussion

AVM of the retina signifies a direct communication of an artery and vein without intervening capillary bed. They are classified into three grades. Grade I is an anastomosis between a small arteriole and a venule; Grade II is an anastomosis between branch artery and a branch vein; Grade III is the diffuse marked dilatation of the whole vascular system.[(1)] Our patient most likely presented with grade III AVM, with grossly dilated tortuous vessels involving most of the retina.

Bonnet-Dechaume-Blanc syndrome (Wyburn mason syndrome), Retino-optic-mesencephalic syndrome, Neuro-retinal angiomatosis and Encephalo retinal facial angioma are syndromes which have characteristic association of facial, retinal and intracerebral arteriovenous communication.[(2)] The rarest of the retinal AVM is Racemose aneurysm, which may be associated with similar malformation in the ipsilateral part of the brain.[(2,3,4)] Involvement of retina and brain is considered to be one of the phakomatosis. It tends to have a positive family history.[(3,4)]

Arteriovenous malformation can present with a wide variability in clinical spectrum. They are usually non-progressive but they may have associated complications such as central and branch retinal vein occlusion, neovascular glaucoma, optic atrophy, retinal exudation, and vitreous hemorrhage.[(1)]

A study on arteriovenous aneurysm of retina revealed characteristic ophthalmic and neurological features. The optic disc may be obscured by tortuous and convoluted vessels. Arteries and veins are difficult to be distinguished by color and caliber. It presents a variable spectrum and only some vessels may be affected, others being spared.[(5)]

Fundus examination revealed dilated, tortuous arteries and veins which were difficult to differentiate from each other. Massively dilated vessels were seen in the inferotemporal and superotemporal arcades. Studies report that the temporal retinal vessels are most commonly involved. The associated features such as

retinal exudates, edema or hemorrhage are extremely rare.[2,5]

Telangiectases or naevi on the face, scalp or oral mucosa may be associated with Racemose aneurysm of retina.[3] In our patient, conjunctival telangiectasia was the only sign of the occult AVM in the retina. There were no facial naevi or telangiectasias.

Retinal AVM though innocuous in the majority of cases, may cause visual complaints in an occasional patient. Visual acuity is 6/9 or better in 30% of the cases and 6/12 to no perception of light in other cases.[5] Deterioration in visual acuity may be due to the slow progression of vascular loop approaching the fovea.[6] Visual loss could be due to retinal atrophy and gliosis.[2] Optic atrophy occurs as a result of abnormal tortuous vessels extending from the retina to the midbrain.[4] A patient with Bonnet-Dechaume-Blanc syndrome has been reported with blindness in the right eye and temporal hemianopia in the left eye. Congenital unilateral retinocephalic AVM may involve the visual pathway from retina and optic nerve to ipsilateral occipital cortex, leading to visual loss.[7,4]

Visual field is generally normal, except a few cases where homonymous hemianopia was observed. Intracerebral angioma was invariably associated in such patients.[5] Visual field analysis was absolutely normal in our case.

Neuroimaging in our patient revealed linear hypodense area with gliosis in the left parietotemporal region extending from the occipital horn to the insular cortex. MRI showed evidence of AVM with multiple tiny tortuous feeder vessels from the left middle cerebral artery forming a sparse nidus within the gliotic area. A study of fifty cases enlightens that the majority of the angiomata were located on either side of the sylvian fissure, corresponding to their origin from middle cerebral vessels.[5]

Hemiplegia occurs due to subarachnoid or intracranial hemorrhage, caused by the AVM. It may be sudden or more often gradual in onset, and develop over few years. It may be due to minute rupture, or decreased blood perfusion of cerebral cortex supplied by middle cerebral artery caused by rapid shunting of blood from artery to vein.[5] The intensity and propensity of intracranial AVM may be variable. If florid, it may lead to subarachnoid hemorrhage, neurological deficit and even death in some cases.[8]

Some cases of retinal Racemose hemangioma do not have intra cranial extension. A case was reported with convoluted and enlarged retinal vessels without intracranial angioma. Visual loss may occur secondary to intraretinal and macular haemorrhage, vaso-occlusive disease, neovascular glaucoma or vitreous haemorrage.[1,8]

Conclusion

Retinal AVM has a widely variable spectrum of presentation. Retinal AVM may be associated with conjunctival telangiectasia. It is mandatory to perform neuroimaging in all cases of retinal AVM to uncover occult cerebral AVM. Retinal AVM may cause visual loss and requires vigilant follow up. Intracranial AVM may result in life threatening intracranial haemorrhages and neurological deficits.

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