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CASE REPORT

Even Mild Proptosis may be Pointing Towards an Underlying Massive Pial Arterio-Venous Malformation with Hydrocephalus - a case report & review of the literature

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Abstract

Cerebral arterio-venous malformations tend to present most commonly with neurological manifestations like headache, intracranial hemorrhage, seizures, or focal neurologic deficit. Intracranial vascular malformations with aggressive imaging features manifesting only with mild unilateral proptosis are very rare with sparse literature available on it. This report stresses upon the fact that even mild to moderate proptosis may be the primary and solitary presenting feature of a large intracranial extra-orbital pial arterio-venous malformation (AVM). Another feature highlighted in this case of unruptured pial arterio-venous malformation is the presence of hydrocephalus which is secondary to extrinsic compression of the cerebral aqueduct by dilated and tortuous draining vein.

Key Words

Unilateral Proptosis, Cerebral Arterio-Venous Malformation, Hydrocephalus

Introduction

Proptosis is defined as an abnormal protrusion of the eyeball with respect to the orbit. [1] Its etiology is myriad and includes endocrinal, inflammatory, infectious, vascular, neoplastic, and metastatic pathologies of orbital as well as extra-orbital origin. The most common cause of both unilateral and bilateral proptosis is Grave's disease. [2] In cases with the unilateral presentation, one should think of orbital pseudotumor, orbital cellulitis, cavernous sinus thrombosis, or intraorbital neoplasm.

Case Report

A 28-year-old married female presented to the Ophthalmology department with a gradually increasing painless forward protrusion of the right eye for the last one month. There was no e/o trauma, headache, redness, loss of vision and diplopia. The patient was non-hypertensive and non-diabetic. She was not pregnant and there was no obvious history of intake of any drug or oral contraceptives.

On examination, proptosis of 2 mm was seen in the right eye on Hertel exophthalmometry. Proptosis was non-pulsatile and was not associated with thrill or bruit. No obvious chemosis or eye redness was seen.Visual acuity, intraocular pressure, colour vision, eye movements and pupillary reactions were normal in both eyes. Fundoscopy revealed normal optic disc and no abnormal vasculature. No sign of thyroid or systemic disease were seen.

The patient was referred to the department of Radiodiagnosis for evaluation of proptosis. Computed tomography revealed an ill-marginated hyperdense lesion with multiple specks of calcification in right temporo-parietal region with mass effect upon the supratentorial

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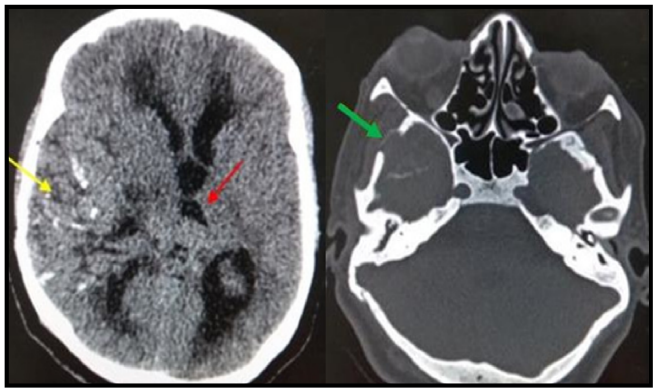

Fig. 1a and 1b: Non-contrast CT axial image of the brain showing an ill-marginated hyperdense mass lesion having multiple specks of calcification in right temporo-parietal region [Yellow arrow] causing midline shift of 4.8 mm towards left side and mass effect upon right lateral ventricle and third ventricle resulting in obstructive hydrocephalus. [Red arrow]. Non-contrast CT axial image of the brain in bone window at level of the base of skull showing linear calcification fociin lesion of right temporal lobealong with cortical thinning and pressure erosion of right greater wing of sphenoid bone. [Green arrow] Right proptosis is also evident in this image.

ventricular system and brainstem suggesting sub-falcine and descending transtentorial herniation. (*Fig 1a)*

Mild proptosis of 2.0 mm was noted on the right side (distance from the inter-zygomatic line to the anterior surface of the globe was 23.1 mm). No intraorbital, retro-orbital or intraocular extension of the lesion was noted. On bone window, cortical thinning, remodelling, scalloping of right greater wing of sphenoid (*Fig 1b*), right parietal and temporal bones were noted.

Diagnosis was confirmed with Contrast MR and MRA. A large diffuse type of pial temporo-parietal AVM on right side wasnoted as a tangle of serpiginous flow voidson T2 W images with intense enhancement on post-contrast T1 W scans (*Fig 2* respectively). On MRA, Lesion appeared to have feeder arteries from branches of the right-sided MCA and PCA (*Fig 3*) with drainage into tortuous and dilated VOG, right ICV, basal vein, straight sinus, sigmoid sinus, SSS and right superficial cortical veins. Right CS & right SOV were also prominent resulting in mild proptosis.

Mild obstructive hydrocephalus was seen owing to aqueduct compression by a prominent vein of Galen with V/H ratio of 34%. No e/o parenchymal hemorrhage, IVH and SAH was seen.

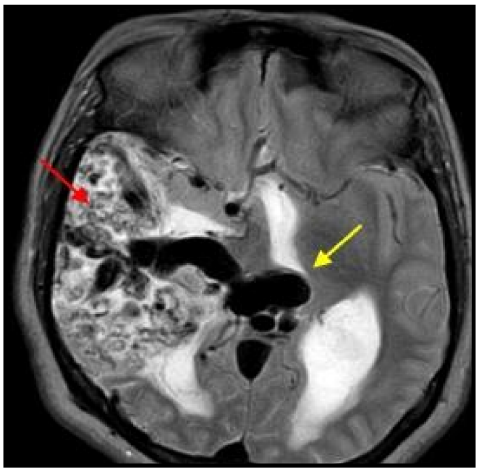

Fig. 2 Pial AVM seen on 1.5-T T2-weighted MR axial image of the brain as a conglomerate of tortuous flow voids representing AVM in right temporal lobe [Red arrow]with feeders from right sided MCA, PCAand drainage into Vein of Galen and straight sinus. Mechanical obstruction of the third ventricle is noted at level of cerebral aqueduct by dilated and tortuous Vein of Galen. [Yellow arrow]

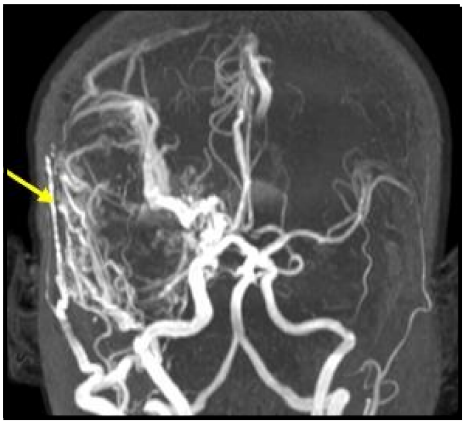

Fig. 3 MR angiogram of the brain in coronal view reveals large right temporoparietalAVM [Yellow arrow]

Considering the size, location, and venous drainage of the AVM, it was classified as grade 5 AVM as per the Spetzler-Martin Grading system. [3]

Discussion

Pial brain AVMs are defined as an abnormal conglomerate of dilated arteries and veins within the brain parenchyma without the normal intervening capillary bed, resulting in arterio-venous shunts. [4] The most common symptoms are hemorrhage, seizures, headaches, and focal

neurological deficits.

Ocular symptoms are very rare in cerebral AVMs, unlike dural arteriovenous fistulae (DAVFs) and carotico cavernous fistula (CCFs) which usually present with conjunctival chemosis, painful exophthalmos and reduced vision. Unilateral proptosis was the primary and sole clinical presentation in our case despite there being no extension of pial AVM into orbit or presence of orbital AVM, dural AVF and AVM directly in relation to cavernous sinus.

Orbital drainage of cerebral AVM is rarely reported in the literature. Volpe NJ et al in a study on 100 adult patients conducted over 4 years with cerebral AVMs found only three patients with orbital drainage. [5]

The proptosis associated with temporoparietal AVM, in this case, can be explained by its hemodynamically complex venous drainage. Apart from draining into deep & superficial cerebral venous system predominantly, some drainage of pial AVM into right cavernous sinus was also noted inflicting greater pressure on ipsilateral superior ophthalmic vein resulting in proptosis.

Another unusual finding in our patient was the presence of hydrocephalus without any evidence of intraventricular or subarachnoid hemorrhage. Geibprasert S *et al* have concluded in their 2009 article that hydrocephalus is an unusual presentation of unruptured AVMs. [6] In their study conducted for 7 years in which they could identify only 8 out of 440 patients whose primary symptoms were related to hydrocephalus from unruptured brain AVMs.

The most common cause of hydrocephalus in a patient with a pial brain AVM is the rupture of intraventricular or subarachnoid hemorrhage, with subsequent blockage of the arachnoid villi or cisterns surrounding the brainstem. A less common cause in unruptured AVMs is an overproduction of CSF, which has been reported in choroidal AVMs. [6]

In our case, imaging diagnosed mechanical obstruction of the ventricular system much before its clinical presentation with symptoms of hydrocephalus or raised ICT like headache and vomiting, etc. Marked thinning, scalloping and massive erosion of right greater wing of sphenoid, due to the pressure effect of a pulsatile AVM, also makes this case unique.

It is essential on part of reporting radiologists to keep in mind another close mimic of pial AVMs, i.e. Cerebral proliferative angiopathy as both entities are observed in young age group and share common imaging features. However, the lack of clear early venous drainage on dynamic images is the key to differentiating this disease from classic brain AVMs. [7] If high index of suspicion is not kept by the clinicians the ultimate presentation may be severe as reported in similar studies. [8, 9]

The patient was lost to follow up as she opted for further treatment at another specialized neurosurgical center. A detailed evaluation of feeders and draining veins with digital subtraction angiography is recommended in such cases to plan management strategies.

Conclusion

The role of early imaging cannot be over-emphasized even in cases with mild proptosis, considering its potential to be the sole and primary clinical presentation of cerebral AVMs.

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Conflicts of Interest

There are no conflicts of interest.

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