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A Rare Case of Unilateral Av Malformation Lateral Aspect of the Eye Ball Causing Esotropia in Right Eye

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Abstract: *We present a case of 21yrs male presented to opd for defective vision left eye. On examination, pulsatile swelling noted over lateral aspect of right eye with esotropia, left eye normal. The swelling was ignored since birth, pulsatile with defect over the temporal bone right side. Vision in right eye – no perception of light, left eye 6/18 ,no improvement with pin hole. Ultrasound, colour Doppler, CT & CECT findings have confirmed the presence of periocular AV malformation present over the lateral aspect of right eye with normal left eye. The case was diagnosed as unilateral periocular AV malformation causing esotropia in the right eye*

Keywords: *Pulsatile swelling, Esotropia, Ultrasound, Color Doppler, AV malformation, CECT*

I. Introduction:

Arteriovenous malformation (AVM) is an abnormal connection between arteries and veins, bypassing the capillary system. This vascular anomaly is widely known because of its occurrence in the central nervous system, but can appear in any location. Although many AVMs are asymptomatic, they can cause intense pain or bleeding. AVMs are usually congenital and belong to the RASopathies[1]. In a normal functioning human body, arteries carry blood away from the heart to the lungs or the rest of the body, where the blood passes through capillaries, and veins return the blood to heart. An AVM interferes with this process by forming a direct connection of the arteries and vein. AVMs are often associated with the brain and spinal cord, they can develop in any part of the body[4].

II. Case Report:

A 21years male presented to our OPD for defective vision in the left eye from 1year. On examination we found pulsatile swelling over the lateral aspect of right eye with esotropia and normal left eye for external examination. On enquiring about the history of the swelling, he said it was congenital and gradually progressed over years shifting the globe medially for which they never seeked any medical consultation. The swelling have distorted the external morphology of the right eye and is associated with defect over the temporal bone. On examination visual acuity RE- No perception of light, left eye 6/18 no improvement with pin hole. Slit lamp biomicroscopy showed pulsatile right globe and eye in esotropia with no pupillary reaction. Left eye anterior segment examination is normal, pupils reacting to light. Fundus right eye showed Optic atrophy and left eye showed normal fundus except for the presence of drusen at the macula. Ultrasound and Color Doppler of the orbit revealed a heterogenous lesion noted on the lateral aspect of the right orbit with vascularity, suggestive of AV Malformation. MRI brain , CT scan and CECT were done and the findings suggested AV malformation presenting as a reticulum over the right lateral aspect of the right globe pushing the eyeball medially down and out with the reticulum extending over the brain tissue . Temporal bone over the area was absent with brain showing gliosis in the region. So the diagnosis of Unilateral AV Malformation over lateral aspect of right eye causing Esotropia.

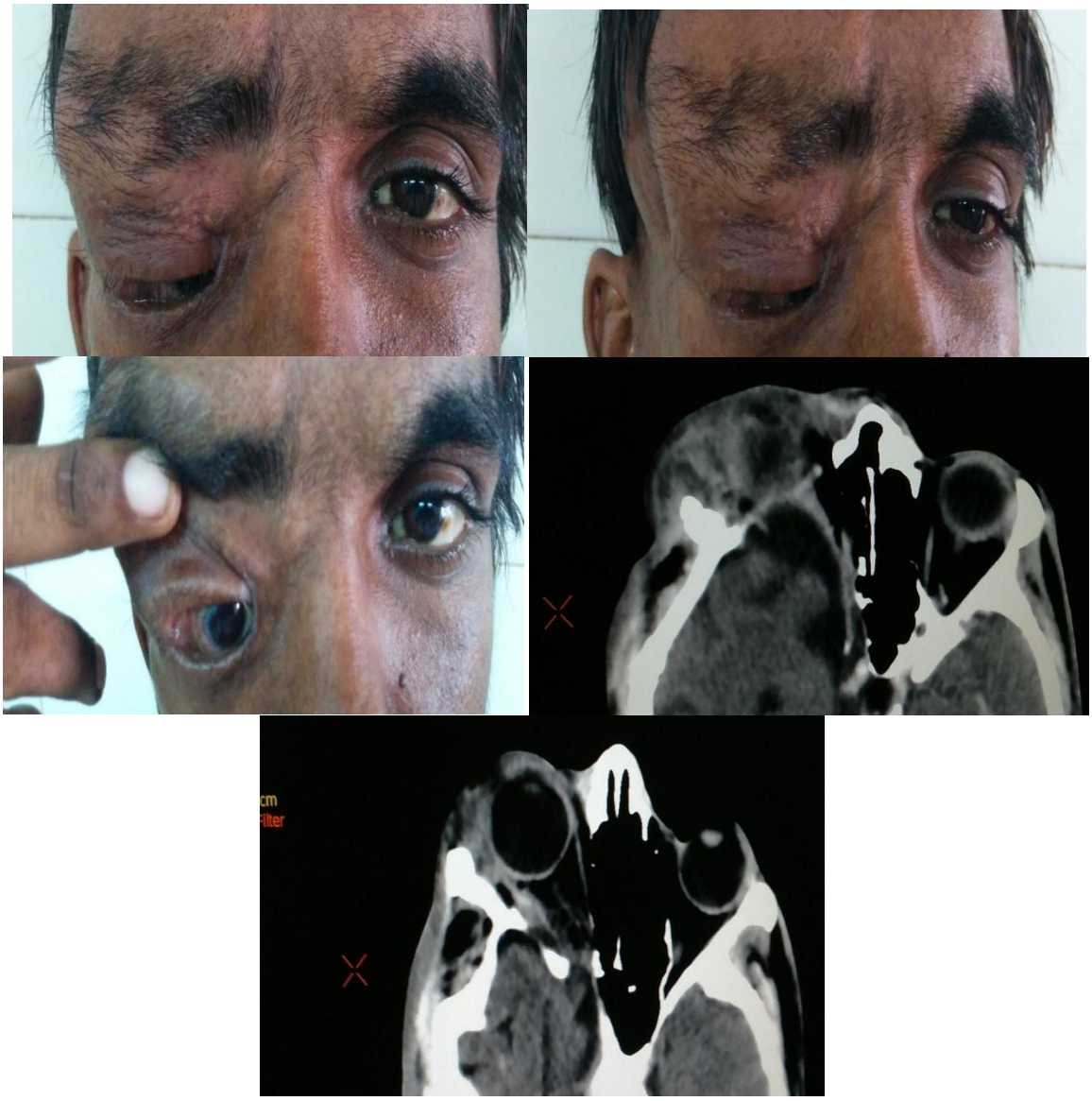
III. Discussion

Arteriovenous malformation (AVM) is an abnormal connection between arteries and veins, bypassing the capillary system[1]. Thisvascular anomaly is widely known because of its occurrence in the central nervous system, but can appear in any location. They are comprised of snarled tangles of arteries and veins. They occur in males and females of all racial or ethnic backgrounds at roughly equal rates[2]. Although many AVMs are asymptomatic, they can cause intense pain or bleeding. AVMs are usually congenital and belong to the RASopathies[3]. The RASopathies are developmental syndromes caused by germline mutations in genes that alter the Ras sub family and Mitogen-activated protein kinases that control signal transduction, including:

- Capillary malformation-AV malformation syndrome

- Autoimmune lymphoproliferative syndrome
- Cardiofaciocutaneous syndrome CFC syndrome
- Hereditary Gingival fibromatosis type 1
- Neurofibromatosis type 1
- Noonan syndrome
- Costello syndrome, Noonan-like[5]
- Legius syndrome, Noonan-like
- LEOPARD syndrome, Noonan-like

Signs and symptoms of a brain AVM may include..,Seizures,Headache or pain in one area of the head, Muscle weakness or numbness in one part of the body[6]. Some people may experience more-serious neurological signs and symptoms, depending on the location of the AVM, including, Severe headache, vision loss, Difficulty speaking, Confusion or inability to understand others, Severe unsteadiness[9]. Specific neurological symptoms that vary from person to person, depending primarily upon the location of the AVM. Such symptoms may include muscle weakness or paralysis in one part of the body; a loss of coordination (*ataxia*) that can lead to gait disturbances; *apraxia*, or difficulties carrying out tasks that require planning; dizziness; visual disturbances such as a loss of part of the visual field; an inability to control eye movement; *papilledema*) [8]; various problems using or understanding language (*aphasia*); abnormal sensations such as numbness, tingling, or spontaneous pain (*paresthesia*or *dysesthesia*); memory deficits; and mental confusion, hallucinations, or dementia. AVMs located on the frontal lobe close to the optic nerve or on the occipital lobe, the rear portion of the cerebrum where images are processed, may cause a variety of visual disturbances[7].



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