Case Report
Recurrent Retinal Artery Ischemia as a Presenting Symptom of Ophthalmic Artery Aneurysm

R.M.R. QAMAR Q. MANSOOR T. MAJEED
Department of Ophthalmology, B.V. Hospital / Quaid-i-Azam Medical College, Bahawalpur
Correspondence to Dr. Rao Muhammad Rashid Qamar, 45-46/C, Mubajir Colony, Bahigadd Road, Bahawalpur, E-mail: dr rashadgr@yahoo.com

Aneurysms most commonly cause nerve palsy, though visual loss may occur. Recurrent transient retinal ischemic attacks caused by ophthalmic artery aneurysm are rare. Ophthalmic artery aneurysms affect the sensory visual pathways by compression of the optic nerve and chiasm.

Case Report:
An 81 years old woman presented with sudden painless greying of her vision in the right eye. This resolved within 1-2 hours without any residual visual loss. She experienced similar symptoms twice in 4 months. The attacks lasted forty to sixty minutes. She had no history of hypertension, diabetes mellitus, migraine, ischemic heart disease and atrial fibrillation. Further neurological and systemic enquiry was negative.

On ocular examination, best visual acuity was 6/9 right eye and 6/60 in left eye. Slit lamp biomicroscopy, tonometry and discs appearances were normal. Right macula showed multiple drusen and dry type of ARMD. There was left disciform macular degeneration. The vascular pattern of both fundi was normal and did not reveal any embolis. Extraocular movements were normal.

Cardiovascular examination, Carotid artery palpation and auscultation were all normal.

Serum cholesterol level was normal. Thrombophilic screen was negative. Magnetic Resonance Angiogram showed aneurysm of right ophthalmic artery near the junction of carotid-ophthalmic artery.

After neurosurgical consultation, she was advised to take Tab. Aspirin 75mg daily.

Comments: Ophthalmic artery aneurysm is a saccular aneurysm. Most sacular aneurysms occur as isolated non-hereditary lesions. Women are most susceptible. The peak incidence of aneurysms occurs in the 4th-7th decades of life. 85% of aneurysms originate from the branches of internal carotid artery. Carotid – ophthalmic aneurysms are rarer than internal carotid-posterior communicating aneurysms and usually produce sensory symptoms and signs due to compression of optic nerve and chiasm. These aneurysms arise from ophthalmic artery beneath the optic nerve and compress the nerve superiorly against the dura causing unilateral visual loss with an inferior visual field defect. Very rarely thromboembolic seeding from the aneurysm to central retinal artery can also lead to transient visual loss. We therefore suggest that ophthalmic artery pathologies should also be included in the differential diagnosis of recurrent retinal ischemic attacks.

Figure 1: MR Angiogram arrow showing ophthalmic artery aneurysm.

References:
Case Report
Adult Onset Adrenal Hyperplasia

S ANWAR N SHAMI N IQBAL SHAHEENA ASIF
Department of Obstetrics & Gynaecology, Lahore Medical & Dental College/ Ghurki Trust Teaching Hospital, Lahore
Correspondence to Shaila Anwar

Congenital adrenal hyperplasia is of two types – early and late onset. The disease is characterized by biochemical markers of decreased serum cortisol, increased adrenocorticotropic hormone (ACTH) and accumulation of the precursors of the affected enzyme which in this case is 21 hydroxylase. Levels of 17 hydroxyprogesterone (17 OHP) differentiates between congenital early and late onset adrenal hyperplasia. ACTH stimulation test is sometimes used to confirm the diagnosis.

Management: Miss P, daughter of G.N. 25 years of age, resident of Lahore, presented in the Outpatient Department of Ghurki Trust Teaching Hospital, Lahore on 08.07.2005 with the complaints of secondary amenorrhoea, weight gain and increasing severity of hirsutism for the last four years.

This patient went to the doctor for the first time at 14 years of age with the complaints of primary amenorrhoea and hirsutism where she was diagnosed a case of congenital adrenal hyperplasia. Clitoroplasty was performed (no documentation was available) and she was given hormone replacement therapy after which she had regular menstruation for 6-7 years. Now, for the last four years she again had amenorrhoea accompanied by increasing weight and hirsutism.

EUA (examination under anaesthesia) was planned. The patient was catheterized; urethral and vaginal openings were seen to be separated by a thin septum. Both labia were normal looking and clitoris appeared to be normal in size. A normal sized uterus was felt and fornices were clear. Various investigations which had been carried out were:

Blood Group: O +ve
Heamoglobin: 11 gms/dl
Blood Sugar Random: 82 gms/dl
Abdomino-pelvic Ultrasonography:
Liver was normal in size and echo texture. Gallbladder was normal. Both kidneys and spleen were normal.
Anteverted uterus 7.1 x 3.4 x 4.2 cm in size with no focal lesion.
Endometrial thickness was 5.8 mm.
Both ovaries were normal looking with a dominant follicle of 19 x 14 mm in the right ovary.

Serum Cortisol: 57 µg/dl
Serum 17 hydroxyprogesterone (17 OHP): 28.2 ng/dl ↑
Serum Testosterone: 511 ng/dl ↑
Serum FSH and Estradiol were within normal limits

CT scan of the abdomen revealed bilateral adrenal hyperplasia.
A diagnosis of late onset adrenal hyperplasia was established.

Conservative management was decided, the patient was put on norethisterone 5 mgs three times a day for 10 days.

She came on the fifteenth day with menstruation. She was then put on ethinyl estradiol and cyproterone acetate (Diane 35) for the next 6 months. She was advised to come for follow up after 3 months.

Discussion
Disease frequency is estimated to occur in 1% of general population, 1-2% of Hispanic and Yogoslavs and 3-4% of Ashkenazi Jews.
When an infant is born with ambiguous genitalia, management also includes counselling the parents. It is helpful to reassure them that the child is healthy but there is a developmental anomaly of the genitalia. Menses is often delayed by up to 2 years. The age of menarche is directly related to the hormonal control of the disease, although menstrual irregularity, including oligomenorrhea and even amenorrhea may occur in spite of good control. Fertility is reduced in some cases. Such patients are genetic females and potentially fertile and must be brought up in the female role regardless of the degree of masculinization of the external genitalia, which can always be corrected.

Changes of masculinization are secondary to the elevated levels of androgens as a result of enzyme defect.

When the disorders present at birth are seen in later life, differences in clinical features must be emphasized. Reconstructive surgery is less commonly necessary in patients who are reared as females.7

Affected females are born with enlargement of clitoris and excessive fusion of the genital folds which obscure the vagina and urethra. Thickening and rugosity of the labia majora are evident and bear some resemblance to the scrotum. The uterus, fallopian tubes and vagina are always present.

The hyperandrogenic symptoms of adult onset CAH are mild and typically present and or after puberty.7

There are two corrective procedures, reduction in size of clitoris and division of labial folds to expose the urethra and vagina beneath. A clitoral reduction is best undertaken in the neonatal period. The division of fused labial folds is best when the folds are there until well after puberty.

The 21 hydroxylase gene is located on the short arm of chromosome 6.4 First trimester prenatal screening is recommended in families at risk with CAH.5 Fetal DNA is used for specific amplification for 21-hydroxylase gene called CYP 21 using PCR (polymerase chain reaction) amplification. Dexamethasone can be given, 20 mg/kg, in three doses and started up to 9 weeks of gestation. Chorionic villus sampling or amniocentesis is then done, if the fetus is unaffected, treatment is discontinued. If the fetus is an affected female, the therapy is continued.

References
7. Gorden R. R., O’Gorman F.P.J., Dewhurst C.J and Blank C.E (1960); Lancet; ii, 736