WYBURN MASON SYNDROME: A CASE REPORT

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PURPOSE

To report a case of Wyburn Mason Syndrome.

METHODS

Case report of a 58 year old female

RESULTS

58-year-old, white, female complaining of painless vision loss and hyperemic eyelid in left eye (OS).

Past ophthalmologic history was unremarkable.

Best corrected visual acuity was 20/20 OD and 20/50 OS. Anterior segment biomicroscopy was normal in right eye, and showed multiple arteriovenous

malformations in upper eyelid, conjunctiva and sclera on OS (photo 1). Fundus examination was unremarkable on OD and showed arteriovenous malformations, enlarged

and tortuous retinal vessels in OS (photo 2). Brain CT scan showed arteriovenous malformations.

CONCLUSION

Wyburn Mason Syndrome, also referred as Bonnet-Dechaume-Blanc syndrome or racemose angioma (RA) is a rare, congenital nonhereditary and unilateral phakomatosis. Patients affected by this condition have multiple arteriovenous malformations (AVM), predominantly in face and brain. WMS has a pleomorphic presentation, without the need to have facial, ocular and cranial arteriovenous malformations to characterize it. RA can be divided into 3 groups: I: has interposition of an abnormal capillary plexus between the major vessels; II: has arteriovenous communication without the interposition of the capillary or arteriolar elements; III: has more

extensive and complex arteriovenous communications (as originally described by Wyburn-Mason). The finding of retinal changes as a first alteration implies in Brain study given the high number of patients who have retinal involvement associated with intracranial disease. Patients generally have a good visual prognosis, unless vitreous hemorrhage occurs and may present with field loss or compressive disease in cases of large volume AVMs. Central nervous system lesions should be followed up due to the possibility of surgical resection or radiotherapy in life-threatening cases.

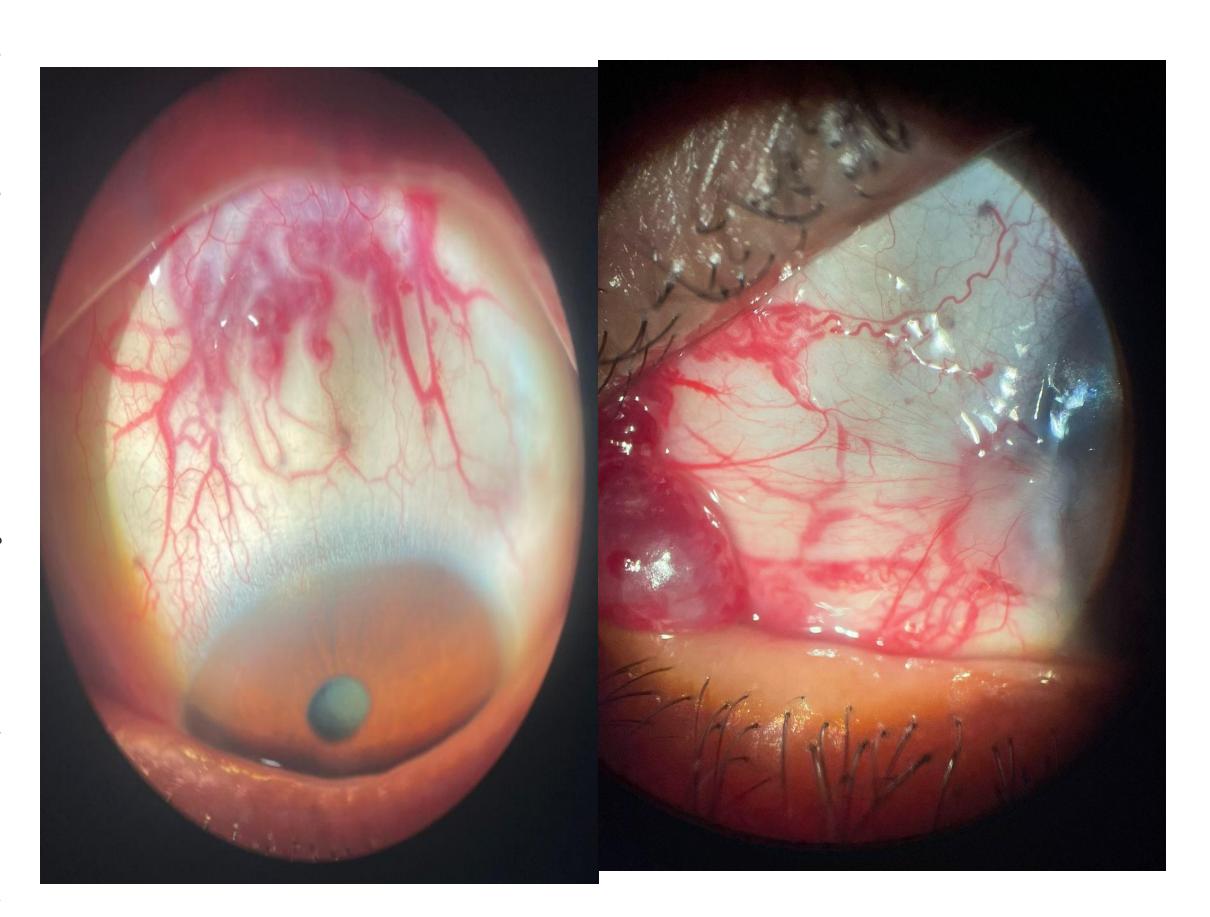


Photo 1 – arteriovenous malformations in anterior segment

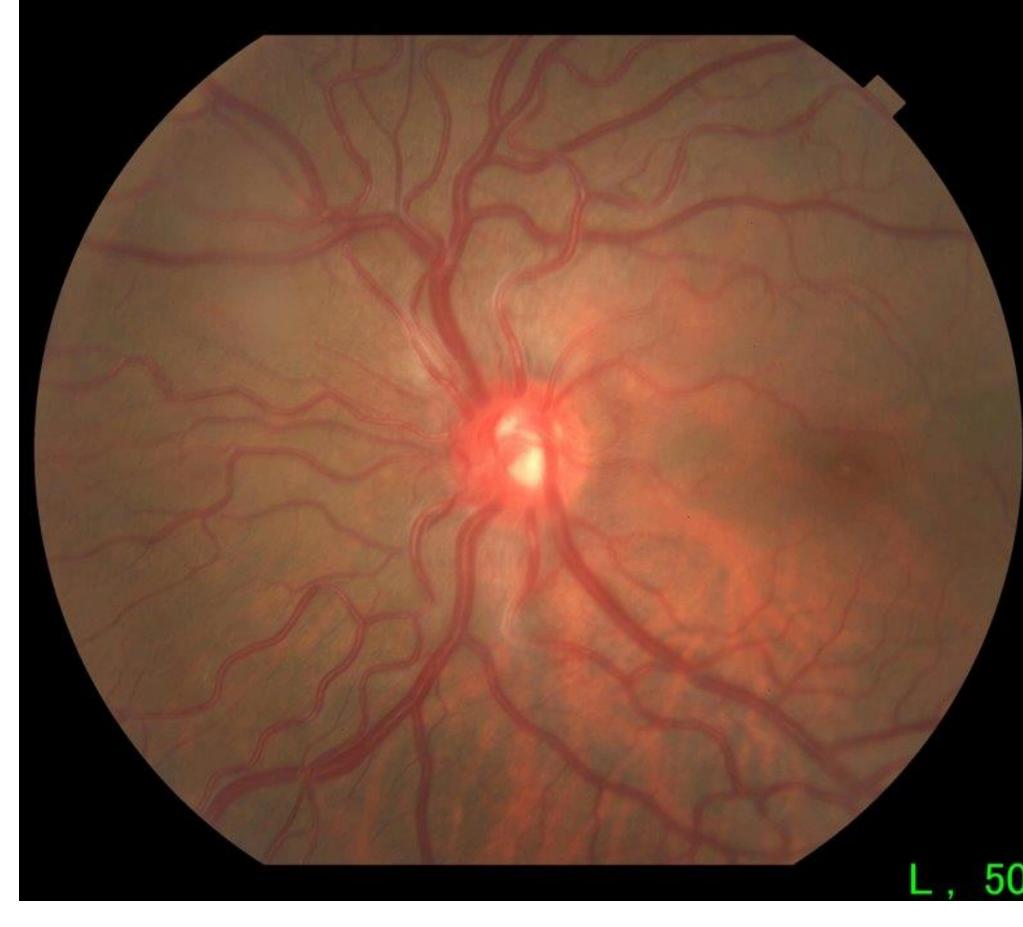


Photo 2 – multiple arteriovenous malformations and enlarged retinal vessels