ABSTRACT

Introduction: Arteriovenous malformations (AVM) of the brain have a low prevalence but are responsible for a significant component of visual pathology in the young population. Vascular studies using selective angiography are necessary to identify arterial connections and venous drainage, and provide useful information for planning treatment.

Clinical case: A 10-year-old child with a temporoooccipital vascular malformation resulting in a left homonymous hemianopia.

Discussion: Arteriovenous malformations of the brain are lesions with a high morbidity and mortality. For the ophthalmologist, they are of special importance due to the possibility of causing a variety of visual defects, depending on their location (Arch Soc Esp Oftalmol 2007; 82: 635-640).

Key words: Homonymous hemianopia, arteriovenous malformation of the brain, angiography, arterial connections, venous drainage.

RESUMEN

Introducción: Las malformaciones arteriovenosas (MAV) son patologías de baja prevalencia pero responsable con relativa frecuencia de patología visual en la población joven. El estudio de la malformación vascular ha de realizarse mediante angiografías selectivas que marquen tanto las aferencias arteriales como los drenajes venosos, además, éstas, ofrecen información muy valiosa para orientar el tratamiento.

Caso clínico: Se presenta un caso de hemianopsia homónima izquierda instaurada en un varón de diez años de edad con malformaciones arteriovenosas cerebrales temporoooccipitales derechas.

Discusión: Las malformaciones arteriovenosas cerebrales son lesiones con una morbimortalidad elevada. Para el oftalmólogo, tienen especial importancia debido a la posibilidad de provocar, dependiendo de su localización, sintomatología visual variada.

Palabras clave: Hemianopsia homónima, malformación arteriovenosa cerebral, angiografía, aferencias arteriales, drenajes venosos.
INTRODUCTION

Arteriovenous malformations may be found in the brain, encephalic trunk and spinal chord. They are made up by anomalous glomerular capillaries connecting the arterial and venous systems and unhindered by capillary beds. Their size varies, the largest frequently appearing in the posterior half of either hemisphere (1).

They affect approximately 0.01 to 0.5 percent of the population. Although these are congenital lesions, complications are likely to appear between 10 and 40 years of age.

The most significant symptoms are hemicrania, epileptic episodes and other conditions resulting from intraparenchymal cerebral hemorrhage, appearing in 50 to 70 percent of cases, and thus the most frequent clinical condition associated with these malformations. Whenever they result in gradual neurological defects, the most frequent are hemianoptic visual defects (2).

Using imaging diagnosis, a contrast CAT scan may detect AVM before it ruptures. NMRs are even more sensitive. However, the study of vascular malformations requires performing selective arteriographies that mark both arterial afferences and venous drainage. Furthermore, these provide very valuable information when prescribing treatment.

Treatment remains controversial: the approach may be passive or else more proactive, through interventional radiology, conventional or stereotactic radiotherapy and surgical procedures, which are the most popular techniques, though not exempt from general and added neurological complications (3).

CASE REPORT

A ten-year-old male suffered in 2001 a sudden and marked cephalina associated with vomits and refractory to ibuprofen. Eye fundus examination revealed an incipient right papilledema. The CAT scan revealed a right parasagittal occipital hemorrhage (fig. 1).

Subsequently, several angiographic studies were performed, revealing two malformation nests, one on the medial temporal right lobe with 7.5 cc volume, and the other, more posterior and localized on the right occipital lobe with 8 cc volume, both lobes showing peripheral angiogenesis. The occipital AMV side presented greater flow than the anterior side.

Neurological examination revealed by confrontation a left homonymous hemianopsia and no other significant findings. The eye fundus examination, once the acute bleeding episode had subsided, was normal for both eyes.

From 2001 to 2005, the child underwent selective embolizations of his two malformation components, decreasing the blood stream in the AMV afferences as well as their size. Subsequently, Gamma Knife radiosurgery was performed (figs. 3a and 3b).
The patient has suffered no relapses as far as the hemorrhage goes, though he presented symptomatic photosensitive epileptic episodes while being treated with valproic acid.

His electroencephalogram shows interhemispheric asymmetry in the right cerebral hemisphere, compatible with a right parieto-occipital cerebral involvement and sporadic epileptiform anomalies localized in these regions.

Currently, the pharmacological dose prescribed has increased in response to new episodes and the patient has been referred by the neurology unit to assess a 4-month-old intermittent dyplopia that was originally attributed to his epileptic episodes and has not subsided despite the pharmacological increase in valproic acid.

During ophthalmologic examination, the patient registered VA of 0.6 (RE) and 0.5 (LE). Pupillary reflexes are normal. He is suffering from orthophoria with ductions and regular versions. The Cover Test revealed near exophoria of -3° -5° and far exophoria of -5° -7°. The eye fundus is apparently normal. The refractive defect reveals myopia of -0.75 diopters in both eyes. A visual field study is performed (Humphrey, 24-2 threshold test) to show a left homonymous hemianoptic defect highly congruent for both eyes, and greater defect density in the lower left quadrant and preservation of the central degrees of the visual field (figs. 4a and 4b). Optical correction is prescribed for the patient in order to improve his visual acuity and bringing his phoria under control.

**DISCUSSION**

Brain AMVs are lesions capable of generating ophthalmologic conditions due to involvement of the visual path. The retrochiasmatic optic tract is made up of fibers carrying information originated in the contralateral hemifields. This is the reason why his lesions result in contralateral homonymous hemianopsias whose specific traits vary depending on the location of injuries. The closer they are to the posterior region and near the occipital cortex, the more congruent the visual field defect. In other words, visual field defects for both eyes share more similarities and the maximum similarities are achieved whenever the lesion of the optic tract is found in the occipital lobe. Furthermore, in this cerebral

*Fig. 3: Serial angio-magnetic resonances with cuts at different angles in order to direct Gamma Knife.*
region there may be preservation of the central degrees of the visual field due to the double irrigation of the macular area. AMVs account for a small percentage of occipital injuries though, regardless their underlying cause, visual field alterations and epileptic episodes are clinical conditions common to all of them (2).

The association between occipital AMVs and visual defects remains unknown. There is no evidence indicating a correlation between the size of malformations and the severity of visual field defects (3).

Since cerebral AMVs are slow-growth congenital lesions, the resulting damages are frequently non-symptomatic and thus diagnosed only when they become obvious or else when diagnostic tests are performed for other reasons. The suggested pathogenic mechanisms for visual field defects include direct compression of the visual tract, heart attacks, hemorrhages, and coronary steal processes leading to partial ischemia caused by tissue hypoperfusion.

Although some cases of occipital AMV with secondary retrograde neuronal degeneration and sectorial papillary atrophy and pupillary defect have been described, they are nevertheless rare (4).

Finally, one should keep in mind that surgical resection and/or stereotactic radiosurgery for these lesions, with or without prior embolization, may be performed without high risk of causing new visual defects or worsening the already existing ones, and even some authors state that early interventions may improve in many cases visual defects secondary to these cerebral vascular lesions (5).

REFERENCES

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