A possible new etiology for ophthalmic varicose veins: disseminated intracranial hyperostosis

Félix J. de Paz¹, Josep M. Potau², Isabel San José¹, Marta González¹, Mercedes Barbosa¹, Natividad García¹

¹ Department of Anatomy and Radiology, University of Valladolid, Spain, ² Unit of Human Anatomy and Embryology, University of Barcelona, Spain

SUMMARY

We wanted to see if cranial anatomical alterations could influence the vascularization of the eyeball, particularly in the production of ophthalmic varicose veins. Orbital dissection of 85 years old woman (who suffered from Alzheimer's disease and respiratory failure).

We observed, when performing a horizontal section of the skull to access the roof of the orbit, that the intracranial surface of the anterior and middle cranial fossae was partially covered by bony outgrowths, with the optical aperture and superior orbital fissure included. Also, when removing the roof of the orbits and beginning to extract the orbital fat we saw a dark, bilateral mass in the upper part of the orbital cavity corresponding to very dilated vessels. Bone alteration of the internal face of the skull was a diffuse intracranial hyperostosis and the dark dilated masses were superior ophthalmic varicose veins.

Our hypothesis is that the origin of these ophthalmic varicose veins was the narrowing of the superior orbital fissure due to excessive bone proliferation. Intracranial hyperostosis produces a difficulty in draining orbital venous blood and, consequently, venous dilation. For this reason, we should consider this in the differential diagnosis.

Key words: Endocranial hyperostosis - Ophthalmic

varices etiology

INTRODUCTION

Our purpose was to see if cranial anatomical alterations could influence the vascularization of the eyeball, particularly in the production of ophthalmic varicose veins.

Intracranial hyperostosis is a relatively frequent finding in people of advanced age. It consists of a proliferation of the inner table of the supratentorial skull. It has a benign character. It presents a symmetrical distribution with respect to the middle line, and it involves a deformation of the inner contour of the cranial cavity with many bumps (Hershkovitz et al., 1999).

It was first described in 1719 by Morgagni (Cuesta et al., 2010). The etiology is unknown. The more consistent statistical association seems to be postmenopausal, as it is often seen in women over the age of 60 years. The aetiopathogenic endocrine-metabolic theories are becoming increasingly important. This probably explains why this alteration was found in the illustrious skull of the castrato Farinelli, who died in 1782 and whose remains were exhumed for study in 2006 (Belcastro et al., 2011).

The varicose ophthalmic veins are uncommon intraorbital masses. They are considered congenital venous malformations: primary (Menon et al., 2004) Or secondary orbital trauma, or important hemodynamic changes (Weill et al., 1998). They consist of abnormal venous vessels of low flow and low pressure that have direct communication

Corresponding author: Félix Jesús de Paz Fernández. University of Valladolid, Faculty of Medicine, Dept. of Anatomy and Embryology, C/ Ramón y Cajal, nº 5, 47005 Valladolid, Spain. E-mail: de_paz@med.uva.es

Submitted: 13 March, 2018. Accepted: 10 April, 2018.

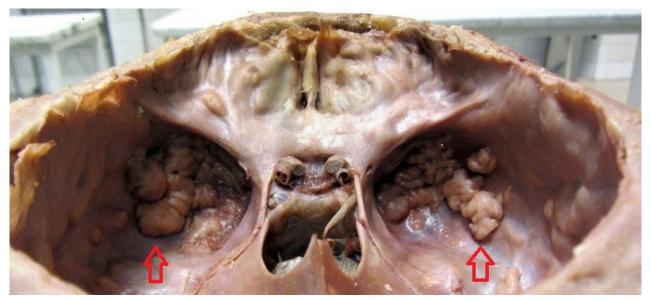


Fig 1. Diffuse intracranial hyperostosis. Posterosuperior view of the anterior and middle cranial fossae, where we can see the diffuse intracranial hyperostosis (red arrows).

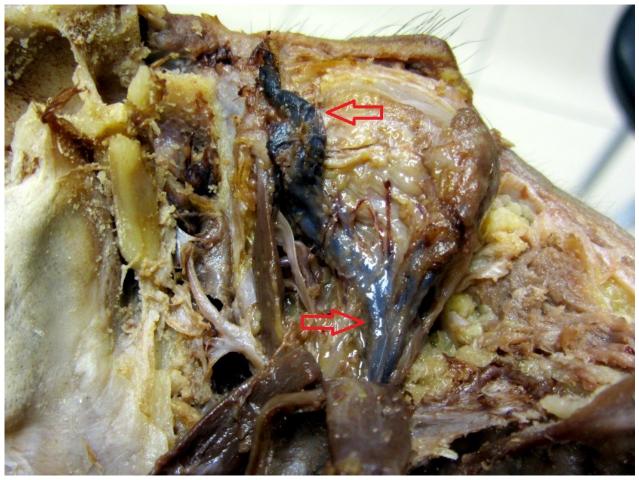


Fig 2. Orbital varix. Top view of the right orbit, after removing fat and cutting and then rejecting the frontal nerve, muscle levator palpebrae superioris and muscle rectus superior, where we can observe the dilated superior ophthalmic vein (red arrows).

with the systemic venous system (Islam et al., 2004). They can be formed by a single vein or a plexo, and most frequently affect young people independently of sex (Weill et al., 1998). Orbital veins have no valves, making them susceptible to

changes in venous pressure. Their diagnosis is based on clinical observations: they cause intermittent exophthalmos, related to an increase in systemic venous pressure, and on radiological techniques: CT, MRI, ECHO-Doppler and Arteriography.

There is no consensus on which treatment is most appropriate, although it is clear that the treatment should be as conservative as possible (Weill et al., 1998; Islam et al., 2004). Among the options we have surgery, embolization, injection of sclerosing agents and electrotrombosis (Weill et al., 1998).

As possible complications, varicose veins may cause thrombosis and hemorrhage. They can even produce an orbital compartment syndrome with intense pain, loss of visual acuity, nausea and vomiting, although, fortunately, this is infrequent (Menon et al., 2004; Islam et al., 2004).

MATERIALS AND METHODS

Orbital dissection of an 85-year-old woman (who suffered from Alzheimer's disease and respiratory failure).

RESULTS

We observed, when performing a horizontal section of the skull to access the roof of the orbit, that the intracranial surface of the anterior and middle cranial fossae was partially covered by bony outgrowths (Fig. 1), with the optical aperture and superior orbital fissure included.

Also, when removing the roof of the orbits and beginning to extract the orbital fat we saw a dark, bilateral mass in the upper part of the orbital cavity corresponding to very dilated vessels (Fig. 2).

Conclusions: Alteration of the internal face of the skull was a diffuse intracranial hyperostosis and the dark dilated masses were superior oph-thalmic varicose veins. Our hypothesis is that the origin of the ophthalmic varicose veins was the narrowing of superior orbital fissure due to excessive bone proliferation. Intracranial hyperostosis produces a difficulty in draining orbital venous blood and, consequently, venous dilation. For this reason, we should consider this in the differential diagnosis.

ACKNOWLEDGEMENTS

The authors would like to thank Isabel Borderas for English grammatical revision of the manuscript.

REFERENCES

- BELCASTRO MG, TODERO A, FORNACIARI G, MAR-IOTTI VJ (2011) Hyperostosis frontalis interna (HFI) and castration: the case of the famous singer Farinelli (1705-1782). *J Anat*, 219(5): 632-637.
- CUESTA M, POTAU JM, CAMPILLO D (2010) Hiperostosis endocraneales supratentoriales. *Paleopatología-AEP*, 7: 1-11.
- HERSHKOVITZ I, GREENWALD C, ROTHSCHILD BM, LATIMER B, DUTOUR O, JELLEMA LM, WISH-BARATZ S (1999) Hyperostosis frontalis interna: an anthropological perspective. *Am J Phys Anthropol*,

109(3): 303-325.

- ISLAM N, MIRESKANDARI K, BURTON BJ, ROSE GE (2004) Orbital varices, cranial defects and encephaloceles: an unrecognized association. *Ophtalmology*, 111: 1244-1247.
- MENON SV, SHOME D, MAHESH L, SUBRAMANIAN N, KRISHNAKUMAR S, BISWAS J, NORONHA OV (2004) Thrombosed orbital varix a correlation between imaging studies and histopathology. *Orbit*, 23: 13-18.
- WEILL A, COGNARD C, CASTAINGS L, ROBERT G, MORET J (1998) Embolization of an orbital varix after surgical exposure. *Am J Neuroradiol*, 1: 921-923.