

P-017 - PERSISTENT CRANIOPHARYNGEAL CANAL ASSOCIATED TO MORNING-GLORY SYNDROME

P. García Feijoo¹, J. Zamorano Fernández¹, M. Sáez Alegre² and C. Pérez López¹

¹Hospital Universitario La Paz, Madrid, Spain; ²Lehigh Valley Health Network Allentown, Pennsylvania, USA.

Resumen

Introduction: A persistent craniopharyngeal duct represents a rare condition (0.42% of general population) where the main concerning symptom are recurrent bacterial meningitis, caused by the abnormal communication between the floor of the sella and the roof of the nasopharynx. On the other hand, the Morning-Glory syndrome (MGS) is another rare condition (2.6 per 100.000), characterized by a congenital optic disc disturbance where a funnel-shaped optic nerve can be appreciated on MRI. The syndrome is mostly unilateral and includes an enlarged disc situated centrally within an excavated area, a dense microvascular network in the peripapillary capillary layer and a whitish glial tissue present in the center of the optic disc. These findings lead to visual acuity defects, strabismus, and myopia. Abnormalities of the face and the central nervous system are described, including basal encephalocele. The physiopathology is not fully understood, but some authors advocate for a partial development of the lamina cribrosa, and incomplete closure of the posterior scleral wall due to a primary mesenchymal abnormality. A primary neuroectodermal dysgenesis can be related to the central gliosis and the abnormal vascular pattern observed.

Case report: We aim to present a pediatric case of persistent crano-pharyngeal canal associated to MGS, consisting of a 1-year-old kid with strabismus who underwent a complete funduscopic exam, OCT, and cranial MRI. A thin-slices skull base CT was further performed to assess these findings. An asymptomatic persistent craniopharyngeal duct and an atretic occipital encephalocele were detected. Non-surgical management was decided after ruling out bacterial meningitis presence and strict instructions about “red flags” for infection were given to parents.

Discussion: A case of persistent craniopharyngeal duct without basal encephalocele associated to MGS, which, to our best known, is an undescribed association, is added to the medical literature.