Neurocirugia. 2017;28(Espec Congr):201

Neurocirugía

https://www.revistaneurocirugia.com

C0349 - RUPTURED RATHKE´S CLEFT CYST WITH ASSOCIATED XANTHOMATOUS HYPOPHYSITIS

C. Barrena López, I. Arrese Regañón and R. Sarabia Herrero

Hospital Universitario Río Hortega, Valladolid, Spain.

Resumen

Objectives: Hypopituitarism in association with visual disturbances is almost always caused by a hypothalamic-pituitary mass. But, in rare instances, may be caused by an inflammatory lesion. Inflammatory lesions of the pituitary gland are rare and observed in less than 1% of surgical interventions. Moreover, asymptomatic Rathke´s cleft cysts (RCCs) are relatively common and have been found in up to 23% of unselected autopsies. Symptomatic RCCs used to be large and cause symptoms via compression of surrounding structures.

Methods: A 68-year-old woman, with a psychiatric history of potomania disorder, was admitted in our centre with panhypopituitarism, headache and bitemporal hemianopia. MRI showed a suprasellar lesion measuring 16 × 16 × 12 mm in contact with the optic chiasm. Contrast-enhanced T1W1 revealed peripheral contrast enhancement with cystic-necrotic parts. The patient underwent an endoscope transphenoidal, transplanum approach. The cyst was excised completely. Postoperative recovery was uneventful. Histopahological examination showed necrosis surrounded by accumulation of foamy cells and xanthomatous epithelioid cells. The tissue was covered on one surface by squamous epithelium. The diagnosis was chronic XGH with ruptured RCCs.

Results: XGH is an autoimmune disorder, recently described, which can occur in isolation, as a part of an autoimmune systemic disorder, or as a reactive degenerative response to an epithelial lesion (RCC). XGH inflammation confined to the hypothalamic neurohypophyseal system causes diabetes insipidus and/or panhypopituitarism. In cases of associated inflammation the RC cyst wall undergoes metaplasia changes from single-layered to stratified epithelium. Hence, as in our case, XGS can also occur in the context of RCC metaplasia.

Conclusions: We propose the ruptured of RCC as the initial pathogenic event for the development of a secondary xanthomataus hypophysitis. Radical surgery is the treatment of choice and carries an excellent prognosis with no recurrences.

See front matter © 2017 Elsevier España, S.L. Todos los derechos reservados