Let's start by defining some terms and background information. Gangliocytomas are uncommon benign tumors of the pituitary gland, often associated with hypersecretion of pituitary hormones. They are usually non-functioning, meaning they do not produce hormones in detectable quantities. However, they can be found in association with other pituitary tumors, such as adenomas, and in some cases, they can be associated with endocrinological conditions like gigantism or acromegaly.

In the case described, the patient was a 43-year-old male with a non-secreting pituitary adenoma with suprasellar expansion. The patient’s father had died from an aortic aneurysm at 68 years of age, and the patient’s mother had a history of high blood pressure who had undergone surgery for nodal tuberculosis at 28 years of age. The patient consulted for severe occipital headache, and an MRI of the brain revealed a round lesion in the right side of the pituitary gland, 10 mm at largest diameter, with elevation of the sellar diaphragm. The lesion was hyperintense on T2 images, showed no contrast enhancement, and was consistent with a pituitary adenoma. No clinical signs of pituitary dysfunction were found. Physical examination was unremarkable, except for grade 2 obesity (body mass index, 35 kg/m²).

Basal measurements of adrenocorticotropic hormone (ACTH) were normal, except for basal cortisol levels of 30 μg/dL and basal ACTH levels of 72 pg/mL. Plasma cortisol levels were normal, and plasma cortisol levels after dexamethasone 1 mg were less than 2 μg/dL. Complete surgical resection of the tumor was performed through a transsphenoidal approach. The pathological report described a neuroepithelial tumor consisting of mature ganglion cells arranged in groups of bipolar neurons, with stroma of non-neoplastic glial cells and reticulin fibers. At immunohistochemistry, ganglion cells were positive for synaptophysin (Fig. 1) and chromogranin. Glial cells were stained with glial fibrillary protein. No adenomatous proliferation was found. After surgery, the patient remained asymptomatic and with no radiographic evidence of residual tumor or relapse, and had no clinical signs of hypercorticoidism.

Gangliocytomas are uncommon lesions in sella turcica which are usually associated with hypersecretion of pituitary hormones (mainly GH and, to a lesser extent, prolactin and ACTH), but endocrinologically non-functioning tumors, such as the one found in this patient, may also occur. Gangliocytoma coexists with pituitary adenoma in 0.52% of sella turcica lesions. The majority of cases have been reported in women.

The origin of these tumors is controversial, and different hypotheses have been proposed. The first theory is that pituitary adenoma results from paracrine or endocrine stimulation by the hypothalamic hormone releasing pituitary hormones, secreted by heterotopic and intrasellar ganglion cells on adenohipophyseal cells (although this would not explain why an adenoma occurs, instead of hyperplasia). The second theory postulates that gangliocytomas are formed due to neuronal differentiation of cells from a poorly granulated adenoma, but does not explain all types of gangliocytomas.

Let's conclude by summarizing the findings. The patient described suffered from a non-functioning gangliocytoma coexisting with a pituitary adenoma, presenting with severe occipital headache. The tumor was resected successfully with no clinical signs of pituitary dysfunction or hypercorticoidism. Immunohistochemical analysis confirmed the presence of ganglion cells and glial cells, with positive staining for synaptophysin and chromogranin.

Figure 1 The image shows immunohistochemical positivity of ganglion cells for synaptophysin, thus confirming the neural origin of proliferating cells with no atypia.
tumors. The third theory suggests an embryonic origin containing cells with intermediate characteristics between ganglion and adenohypophyseal cells. This is the most likely hypothesis because it provides the best explanation for the characteristics of both mixed tumors and pure ganglion cell tumors.

These lesions cannot be distinguished from an adenoma based on clinical, biochemical, and radiographic findings. Diagnosis is therefore made after surgery based on histological examination. Immunohistochemistry with neuronal and glial markers (mainly synaptophysin and specific neuronal enolase) and antibodies against pituitary hormones confirms diagnosis. Ultrastructural studies are also helpful.

Management should be the same as for an adenoma.

References


Paula Andújar-Plata, José Manuel Cabezas-Agrícola, María Teresa Rivero-Luis, Eugenio Pérez-Becerra, Alfredo García-Allut, Felipe Casanueva