Atlas of Science
another view on science
http://atlasofscience.org

Gangliocytomas of the seller region: present and future

Gangliocytomas are well-differentiated tumors formed by irregular groups of large multipolar neurons, often with atypical characteristics. They are benign tumors and a complete tumor resection is considered curative. Their localization in the sellar region is extremely rare (Fig. 1).

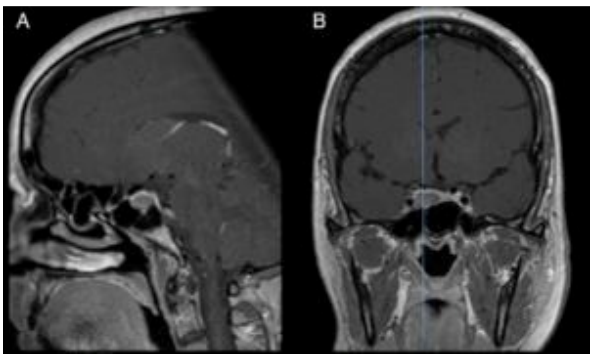

Fig. 1. Sagittal (A) and coronal (B) T1-weighted cerebral MRI after gadolinium administration showing a sellar gangliocytoma coupled with a pituitary adenoma. The radiological features are similar to an isolated pituitary adenoma. The coronal view (B) shows an invasion of the right cavernous sinus.

We performed a systematic review of the literature to collect all the cases reported up to now and to summarize the most recent literature evidences on the subject.

111 cases are reported in literature and 85% are associated with pituitary adenomas, benign and slow-growing tumors arising from pituitary cells, which also represent the most frequent tumors of the sellar region.

A female prevalence is evident, with a ratio of 2.2 : 1 for isolated sellar gangliocytomas (ISG) and a ratio of 4 :1 for sellar gangliocytomas couples with pituitary adenomas (SGPA). The mean age at diagnosis is 44 years.

Interestingly 84% of cases present endocrinological manifestation even if they have only ISG (symptoms of hyperprolactinemia such as disturbance of the menstrual cycle and galactorrhea in 44% of cases, acromegaly in 37% of cases and Cushing's disease in 19%). With ISG visual disturbances were present in 47% of cases at presentation.

In front of SGPA, patient present most frequently acromegalic manifestations (67%), followed by visual manifestations in 29%.

Immunohistochemistry showed a higher prevalence of mixed GH-PRL adenomas in association with sellar gangliocytomas (43%), followed by GH adenomas (33%). The distribution of pituitary adenomas associated with sellar gangliocytoma according to their immunohistochemistry is illustrated in Figure 2. The gangliocytoma component was most frequently positive for GHRH.

Radiologically, a pure sellar localization was found in 23% of cases and a suprasellar extension in 31% of cases of ISG. A more extensive infiltration was present in 38% of cases. The results were similar with SGPA.

A complete resection was obtained in 67% of ISG and 56% of SGPA. Most of SGPA were operated through an endoscopic trans-sphenoidal approach. For ISG a similar number of cases were operated though a transnasal or a transcranial approach.

The follow-up period was variable. No major complications were reported in the postoperative period. Endocrine remission was reported in 81% of cases of SGPA.

In conclusion, sellar gangliocytomas are rare lesions and an association with pituitary adenomas should always be searched. Mixed GH PRL adenomas are most frequently isolated and they generally provoke a GH-hypersecretion syndrome.

The female prevalence has not been investigated up to now but we can argue that female patents may present more evident clinical presentation. A hormonal dependence of intrasellar gangliocytomas should be investigated.

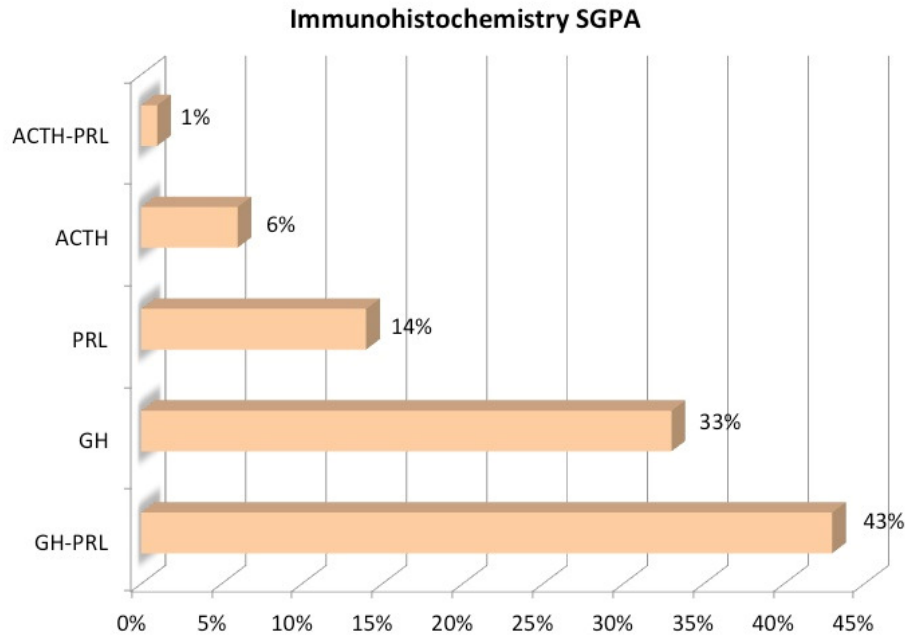

Fig. 2. The pituitary adenomas coupled with seller gangliocytomas are classified according to the immunohistochemical analysis. Most frequently the pituitary adenoma was a mixed GH-PRL adenoma or a GH-adenoma. Other histotypes were rare.

Even when isolated, sellar gangliocytomas may present with hormonal hypersecretion syndromes: ganglionic cells may secrete pituitary or hypothalamic hormones but evidences are still scarce.

Also the pathogenesis of sellar gangliocytomas and their association with pituitary adenoma merit to be further investigated.

Collision lesions seem to have a behavior similar to isolated adenoma but a more important aggressiveness should be excluded at a molecular level. Incompletely resected tumors might show limited response to hormone-targeted therapies or radiotherapy. Further studies will allow advancements in clarifying these aspects and in improving the future management of these rare entities.

Cossu Giulia, Daniel Roy Thomas, Messerer Mahmoud
Department of Neuroscience, Section of Neurosurgery, University Hospital of Lausanne, Switzerland

Publication

Gangliocytomas of the sellar region: A challenging diagnosis.
Cossu G, Daniel RT, Messerer M
Clin Neurol Neurosurg. 2016 Oct