A Mixed Gangliocytoma–Plurihormonal Tumor of PIT1-Lineage of the sellar region: Case Report and Review of the Literature

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Case Presentation

A 46-year-old male presented with acromegaly and progressive visual loss due to an expansive sellar and suprasellar mass of 26 mm that compressed the optic chiasm. The patient was submitted to an endonasal endoscopic transsphenoidal surgery for tumor resection.

Histological examination revealed a PitNET ("pituitary adenoma") mostly composed of pleomorphic cells with large and chromophobic cytoplasm besides few tumor cells with eosinophilic cytoplasm. The neoplastic cells showed slightly hyperchromatic round nuclei and the chromatin was stippled (Fig. A). Noteworthy was the presence of some intermingled mature neuronal cells with evident ganglionar differentiation (Fig. B).

Reticulin stain showed loss of the usual acinar pattern. Immunohistochemistry study revealed diffuse nuclear positivity for PIT-1 and focal cytoplasmic positivity for GH, PRL and TSH in the neoplasm. The low molecular weight cytokeratin CAM5.2 was expressed in the majority of tumor cells and the Ki-67 labeling index was 2% in the hot spot. Other hormonal markers were negative. Ganglion cells stained positive for NEU-N and MAP-2 (Fig. E).

The final diagnosis was established as mixed gangliocytoma – plurihormonal tumor of PIT1-lineage.

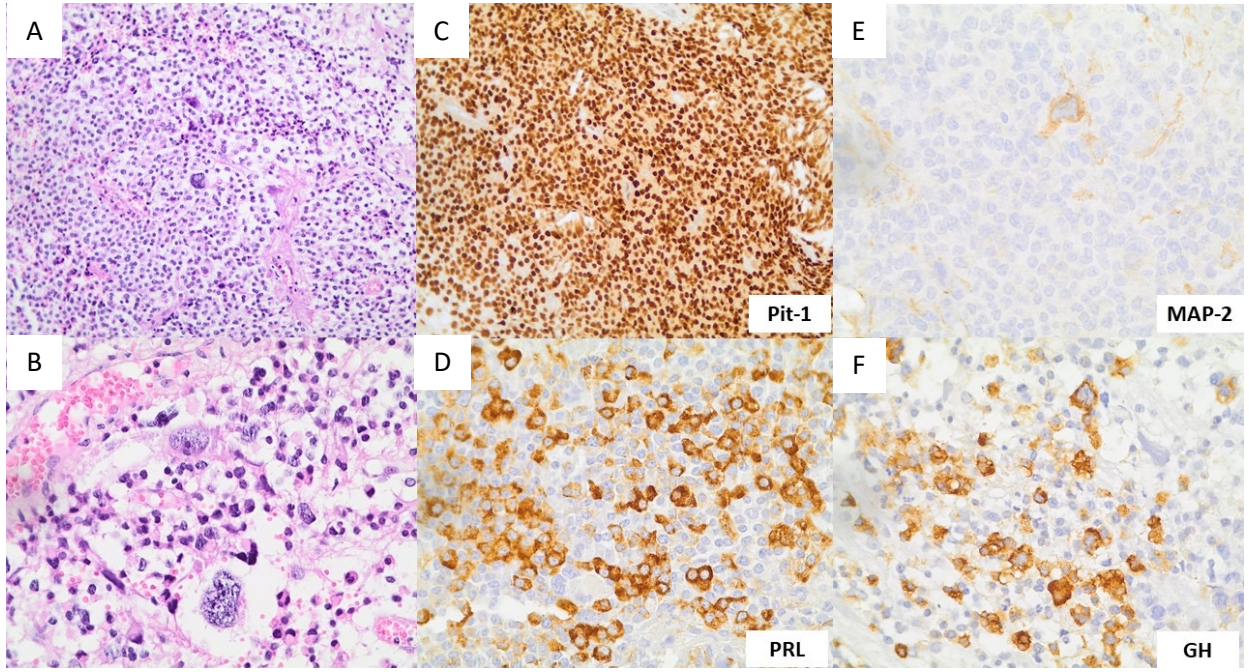
During follow-up, nineteen months later the patient underwent total thyroidectomy to remove a papillary thyroid carcinoma and 3 of 4 hyperplastic parathyroid glands.

Discussion

Mixed gangliocytomas – PitNETs are rare tumors in the hypothalamic/sellar region with biphasic morphology: neuroendocrine and gangliocytic components. In most cases, patients had acromegaly. It is still controversial whether these tumors should be considered as derived from the hypothalamus or the pituitary gland.

A proposed mechanism involves the production of GH-releasing hormone (GHRH) by the gangliocytic component, which promotes the tumor growth, including the neuroendocrine GH-secreting component.

In short, we present a mixed gangliocytoma-pituitary adenoma, a rare neoplasm of uncertain origin with poorly understood pathogenesis. In the current case, the tumor exhibited PIT1-lineage differentiation and concomitant expression of GH, PRL and TSH.



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