

Acromegaly associated with gangliocytoma

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Acromegaly has an incidence of 3 cases per million people per annum, the majority of which are caused by a pituitary somatotroph adenoma. Acromegaly secondary to growth hormone-releasing hormone (GHRH)-secreting neuroendocrine tumours is responsible for less than 2% of cases. We report a case of acromegaly associated with intrasellar GHRH-positive gangliocytoma.

A 56 year old man was observed to have features of acromegaly when he presented to a surgical service for varicose vein ligation. IGF-1 was 1336 ng/ml. MR imaging revealed a mass lesion of the pituitary with stalk deviation to the right. Growth hormone (GH) failed to suppress after oral glucose tolerance testing (GH nadir 9.2 ng/ml), and mean GH was 10.3 ng/ml on five-point GH day curve. Trans-sphenoidal debulking was performed and mean GH dropped to 7.5 ng/ml after the operation. GH was not suppressed following OGTT post-operatively and IGF-1 remained elevated at 889.0 ng/ml. Insulin tolerance test revealed cortisol deficiency and he was also gonadotrophin and TSH deficient. He was commenced on replacement hydrocortisone, testosterone and thyroxine and octreotide LAR 20mg for control of GH hypersecretion. Mean GH of 1.14 ng/ml was achieved at an octreotide dose of 30 mg every four weeks.

The histology report from the debulking procedure was unusual. Adenomatous tissue was identified, which was positive for GH and prolactin on immunohistochemistry staining. Ganglion cells were interspersed with the pituitary tissue and were identified by positive synaptophysin and neurofilament stains. The ganglion cells were also positive for GHRH.

Gangliocytomas are benign tumours of neuronal tissue and are rarely located in the sella. Gangliocytomas located outside the sella are often positive for hypothalamic releasing factors but are not associated with pituitary adenoma formation. There is ongoing controversy about the origin of gangliocytomas associated with pituitary adenomas – perhaps the gangliocytoma secretes a hypothalamic releasing factor which promotes hyperplasia and adenomatous change in the surrounding pituitary, or perhaps poorly-differentiated adenomas slowly undergo cellular differentiation into neuronal tissue.

In our case the duration of acromegaly prior to diagnosis was uncertain and no hyperplasia was identified in the tissue surrounding the adenoma, thus neither of the above theories to explain the genesis of these tumours can be excluded.