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Gamma Knife Radiosurgery in Patients with Persistent Acromegaly or Cushing's Disease: Long-Term Risk of Hypopituitarism

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For patient with a recurrent or residual acromegaly or Cushing's disease (CD) after resection, Gamma knife radiosurgery (GKRS) is often used. Hypopituitarism is the most common adverse effect after GKRS treatment. The paucity of studies with long-term follow up has hampered understanding of the latent risks of hypopituitarism in patients with a Acromegaly or CD. We report the long-term risks of hypopituitarism for patients treated with GKRS for Acromegaly or CD. Methods: From a prospectively created, IRB approved database, we identified all patients with a Acromegaly or CD treated with GKRS at the University of Virginia from 1989 to 2008. Only patients with a minimum endocrine follow up of 60 months were included. The median follow-up is 159.5 months (60.1-278). Thorough radiological and endocrine assessments were performed immediately before GKRS and at regular follow-up intervals. New onset of hypopituitarism was defined as pituitary hormone deficits after GKRS requiring corresponding hormone replacement. Results: 60 patients with either Acromegaly or CD were included. Median tumor volume at time of GKRS was 1.3 cm3 (0.3-13.4), median margin dose was 25 Gy (6-30). GKRS induced new pituitary deficiency occurred in 58.3% (n=35) of patients. Growth Hormone deficiency was most common (28.3%, n=17). The actuarial overall rates of hypopituitarism at 3, 5, and 10 years were 10%, 21.7%, and 53.3%, respectively. The median time to hypopituitarism was 61 months after GKRS (range, 12-160). Cavernous sinus invasion of the tumor was found to correlate with the occurrence of a new or progressive hypopituitarism after GKRS (p=0.018). Conclusions: Delayed hypopituitarism increases as a function of time after radiosurgery. Hormone axes appear to vary in terms of radiosensitivity. Patients with adenoma in the cavernous sinus are more prone to develop loss of pituitary function after GKRS.

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Subependymal Giant Cell Astrocytoma (SEGA) in Adults without Tuberous Sclerosis

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Subependymal giant cell astrocytoma (SEGA) is typically seen in children with tuberous sclerosis (TS), who present with headaches and seizures, and characteristic clinical and cutaneous manifestations of TS. Surgical resection, CSF diversion (for hydrocephalus), radiotherapy, and chemotherapy with a mammalian target of rapamycin (mTOR) inhibitor are treatment options. SEGA can occur in adults without TS, raising diagnostic and therapeutic challenges. A 53 year old man presented with headaches and diplopia. An exophytic, enhancing mass in the left lateral ventricle was resected, confirming SEGA. There was no recurrence on MRI 13 months later. He previously had an "astrocytoma" involving the left frontal horn resected at age 19. Pathology review was SEGA. He had no clinical or cutaneous findings of TS, and no family history of TS. An identical twin was well. A 66 year old man presented with "weakness", due to diuretic-induced hypokalemia. CT showed a hypodense mass from the right caudate head deforming the frontal horn. MRI showed an exophytic, enhancing mass from the caudate head into the frontal horn. A stereotactic biopsy confirmed SEGA. He had no clinical or cutaneous findings of TS, and negative family history. His mother had a meningioma resected at age 66. Observation is planned. These 2 adult patients had SEGA without clear clinical findings or family history of TS. SEGA should be in the differential diagnosis of tumors involving the lateral ventricle.

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Presentation of Intradural Extramedullary Plasmacytoma with Intratumoral Hemorrhage: Case Report and Review of Literature

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Plasmacytomas are solitary tumours characterized by neoplastic proliferation of plasma cells and can be found isolated or in associated with multiple myeloma. Plasmacytomas uncommonly occur intracranially, and dural plasmacytomas without involvement of the calvarium are exceedingly rare. Reported cases indicate durally-based plasmacytomas mimick the appearance of meningioma, lymphoma or sarcoma of the dura. The authors report a case of a 77-year-old male with known multiple myeloma who presented with a 3-week history of confusion, speech impediment, and right sided weakness. A non-contrast CT scan revealed a dense extra-axial mass in the left frontal lobe with initial concerns of an extra-axial hemorrhage. A subsequent MRI demonstrated a contrast enhancing mass with a broad-based dural tail and no underlying calvarial lesion. Differential diagnosis included meningioma or intracranial plasmacytoma. The patient underwent surgical resection and was found to have intratumoural hemorrhage, with pathology confirming plasmacytoma. In the published literature, there are only 20 prior reports of dural plasmacytomas (with and without primary calvarial infiltration), of which only five previous cases reported associated intratumoural hemorrhage. Our case, along with this literature, suggests that new onset of focal neurologic deficits in patients with a history of multiple myeloma merits careful investigation, and that intracranial plasmacytoma should be considered on the differential diagnosis even when imaging reveals masses consistent with hemorrhage or meningioma.