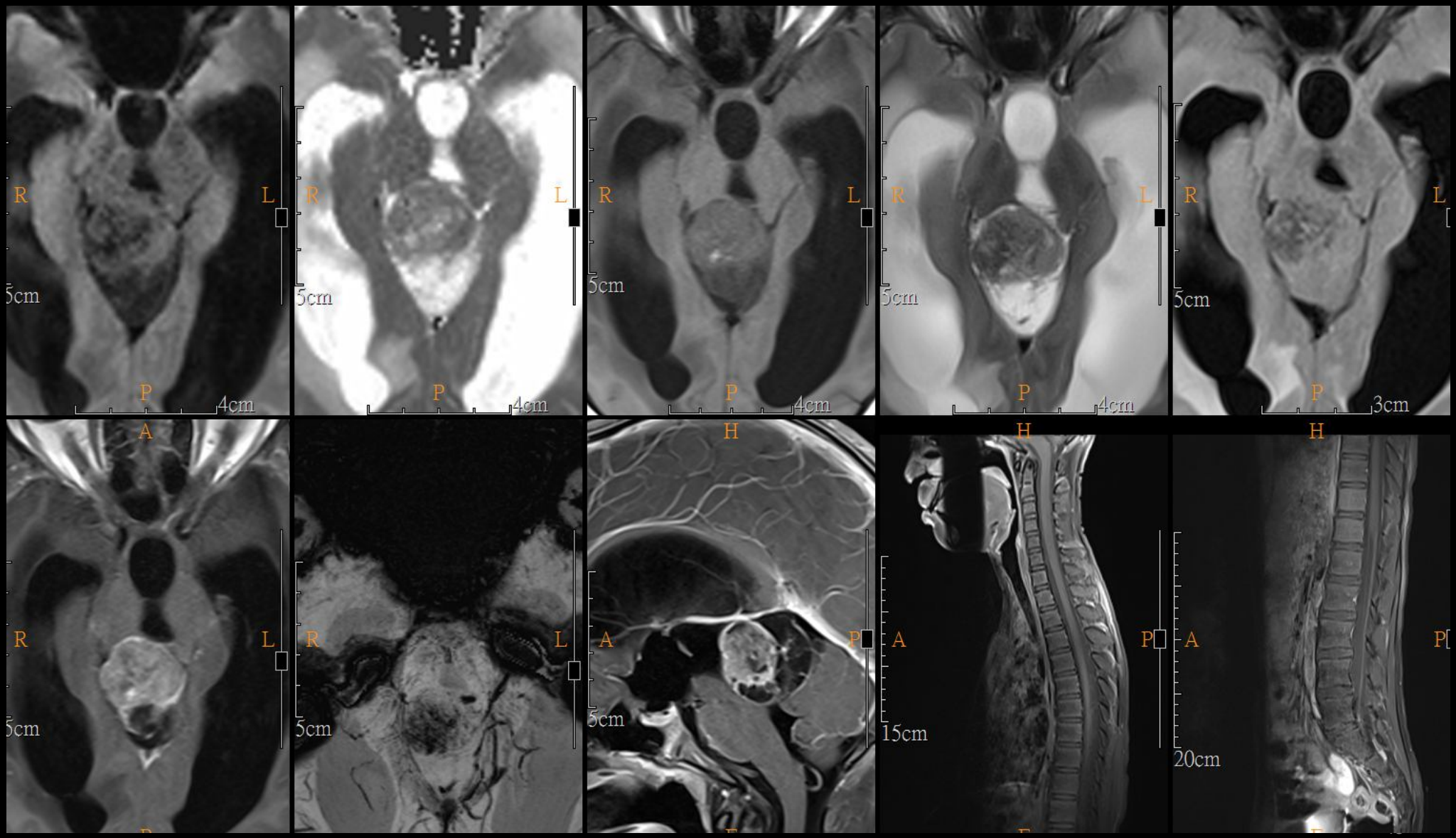
Case 3

Patient Profile

- 12 years old, male
- Chief complaint
  - Headache for 3 weeks
  - Slow walking for 4 days
  - Unsteady gait for 2 days
- Past history
  - None

Image

- 2018-10-20 MRI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Differential Diagnosis

- Germinoma
- Other malignant germ cell tumor
- Teratoma
- Pineocytoma
- Pineoblastoma

Clinical Course

- Underwent supracerebellar infratentorial approach for near total removal of tumor

- Pathology
  - Glioblastoma (WHO grade IV), H3 K27M-mutant
  - High-grade nuclei and abundant eosinophilic cytoplasm with focal rhabdoid features.
  - Positive for GFAP and variably positive for NSE, focally positive for EMA, whereas they are negative for CK (AE1/AE3), SMA, and synaptophysin.
  - Ki-67 proliferation index is variable and up to 50%.
  - INI-1 staining is equivocal that focal region shows loss of staining.
  - Further immunohistochemistry study shows the tumor cells do not express IDH-1 R132H mutant protein

Discussion

Pineal Region Glioblastoma

Pineal Region Glioblastoma

- Pineal region tumors most frequently comprise three categories: germ cell, parenchymal cell, and glial tumors
- They make up less than 1% of intracranial tumors in adults
- Most pineal gliomas are low-grade astrocytomas
- Glioblastoma multiforme (GBM) tumors are exceedingly rare tumors in the pineal region, with approximately 65 cases reported
- The clinical and radiologic characteristics of pineal GBM does not differentiate it from other malignancies of this region; thus, surgical biopsy is generally required for definitive diagnosis
- Symptoms are related to increased intracranial pressure/hydrocephalus and ophthalmologic symptoms (especially Parinaud syndrome)

Pineal Region Glioblastoma

- Heterogeneously enhancing mass with a necrotic component involving the pineal region, and infiltration into the surrounding structures, such as the midbrain and thalamus.
- Pineal region GBMs do not show a higher rate of leptomeningeal or ependymal dissemination compared to other high-grade pineal tumors
- **Differential Diagnosis:**
  - Germ cell tumors (most common)
  - Pineal parenchymal tumors
  - Astrocytoma of the pineal gland
  - Pineal metastasis

frontiers in Oncology

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Check for updates

Pineal Region Glioblastoma, a Case Report and Literature Review

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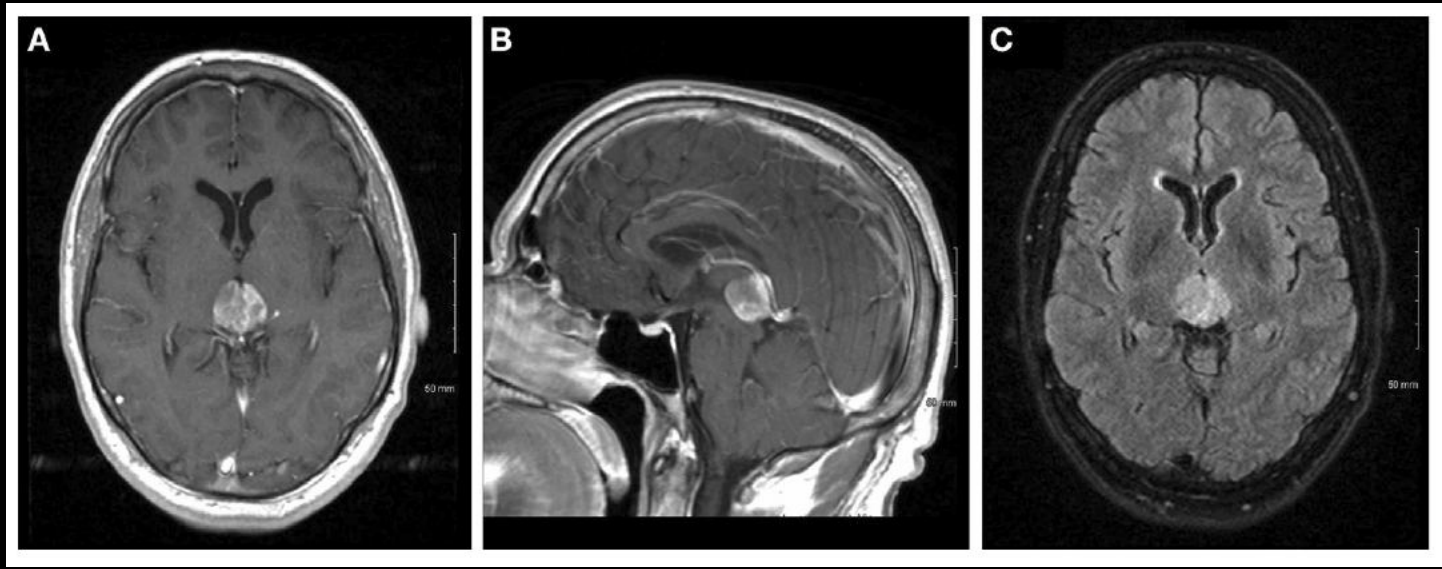
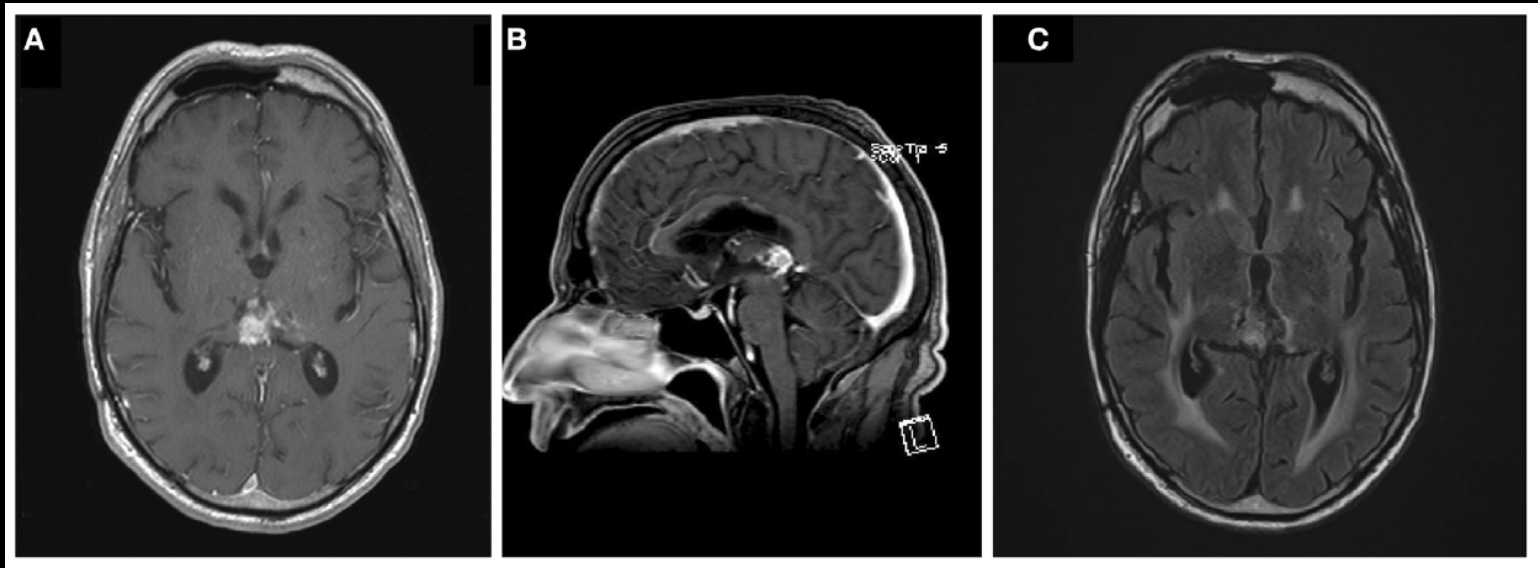
				enhancement		brain RT	
Pople et al. (14)	6	F	HA, N/V, diplopia, decreased visual acuity,	CT and MRI: HCP, enhancing mass	Yes	Shunt, resection, local RT,	4 months
Amini et al. (16)	40	M	HA, N/V, diplopia, blurry vision	CT: obstructive HCP, strong enhancement, punctuate	Yes	Endoscopic TVB, resection,	5 months
Amini et al. (16)	43	M	Ha, disequilibrium, decreased level of mental status	MRI: heterogeneously enhancing, HCP	Yes	TVB, resection, whole brain RT, chemo	7 months



Initial MRI



Follow up MRI after 166weeks

Treatment Option

- A tri-modal treatment approach of surgical resection followed by chemotherapy and radiotherapy yielded a median survival time of 12 months (range, 4–24 months)
- Those patients who received radiotherapy and chemotherapy without resection had the highest median survival duration of 20 months
- Difficulty of achieving a gross total resection in tumors ?
- Increased the risk of leptomeningeal spread ?

Treatment Option

- *O*-6-methylguanine-DNA methyltransferase (MGMT) promoter methylation predicts for better outcomes for patients with GBM in other locations in the brain
- In pineal region GBM?
- IDH mutation?