Enhancing Sellar/Suprasellar Mass in an Adolescent

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Case Presentation

A 16-year-old boy with a history of acute lymphoblastic leukemia (ALL) in remission presented with increasing headache, ptosis, myalgias, fatigue, and numbness and pain in his lower back and sacrum. Initial workup, including CT and MRI of the brain is shown (Fig.).

Figure. Axial noncontrast CT image of the brain (A) shows a round, hyperdense non-calcified mass in the suprasellar region. Post-contrast sagittal (B) and coronal (C) T1 MRI images reveal a circumscribed sellar and suprasellar mass with diffuse homogenous enhancement.
Key imaging finding
Enhancing sellar/suprasellar mass in an adolescent

Differential diagnoses
- Craniopharyngioma
- Chiasmatic-hypothalamic glioma
- Germ cell tumor
- Recurrent leukemia
- Langerhans cell histiocytosis

Discussion
Approximately 2.5-4 per 100,000 children will be affected by a tumor of the central nervous system, which is second in incidence for childhood malignancies behind leukemia. The differential diagnosis for CNS tumors varies based upon location, age, sex, and image characteristics, as well as clinical symptoms and history of any known malignancy. The differential diagnosis for an enhancing sellar/suprasellar mass in an adolescent includes more common entities, such as craniopharyngioma, chiasmatic-hypothalamic glioma, and germ cell tumor. Pituitary macroadenomas (in children), Langerhans cell histiocytosis, and hypothalamic hamartoma are less common. Leukemia should be a consideration in a patient with a history of prior or current disease. Magnetic resonance imaging is the modality of choice for further characterization of masses of the sellar/suprasellar region with computed tomography playing a complimentary role in evaluating for calcifications.

Craniopharyngioma.
Craniopharyngioma is a low-grade (WHO grade 1) neoplasm and represents the most common sellar/suprasellar tumor in children, accounting for approximately 6-9% of CNS tumors. Common presenting symptoms include headache, visual disturbances, hormonal imbalances, and behavioral changes. There are two pathologic variants which determine the age of presentation (bimodal) and overall prognosis. The adamantinomatous variant occurs in children with a peak age of presentation between 10 and 14 years. It presents as a cystic and solid sellar/suprasellar mass with avid enhancement of solid components and calcifications (>90% of cases). The calcifications tend to be stippled and are often peripheral in location. The papillary variant occurs in middle-aged and older adults and typically presents as a solid enhancing sellar/suprasellar mass; cystic changes and calcifications are less common.

Chiasmatic-Hypothalamic Glioma.
Gliomas are common CNS tumors in children, the majority of which are low grade. Common locations in pediatric patients include the posterior fossa and optic pathways. Gliomas may occur sporadically or in association with neurofibromatosis type 1 (NF-1). Chiasmatic-hypothalamic gliomas have no sex predilection and are typically seen in children; they rarely occur in adults. Presenting symptoms include painless proptosis, visual impairment, diabetes insipidus, diencephalic syndrome, and precocious puberty. Radiographs demonstrate a classic J-shaped sella turcica, as well as enlarged optic canals. On MRI, gliomas are typically T1 isointense to mildly hypointense, T2 hyperintense, and show variable degrees of enhancement.

Germ Cell Tumors.
Intracranial germ cell tumors are fairly uncommon, accounting for approximately 0.4-3.4% of intracranial neoplasms. Boys have an incidence rate approximately twice that of girls. Germ cell tumors have a propensity for the suprasellar and pineal regions. Symptoms depend upon the aggressiveness of the tumor and its location. Common presenting symptoms include headache, hormonal imbalance, and visual disturbances. Prognosis is dependent upon the tumor type, the most common of which is a germinoma which has a peak incidence during pubescence. Prognosis for germinomas is generally good with a 10-year survival rate over 90% after radiation therapy. Nongerminomatous germ cell tumors present in slightly older aged children (between 10-21 years of age) and have a worse prognosis with cure rates of approximately 40-60%. Tumor markers are important for the diagnosis and classification of germ cell tumors.

On MRI, germinomas present as infiltrating, homogeneously enhancing masses which typically...
follow gray matter signal intensity on pre-contrast sequences. CSF seeding is common; therefore, imaging of the entire neuroaxis should be performed. A teratoma typically presents as a heterogeneous cystic and solid mass with macroscopic fat. Less common nongerminomatous tumors have nonspecific imaging and often heterogeneous characteristics. Biopsy is typically required for specific diagnosis.

Recurrence Leukemia.

Leukemic infiltration can be difficult to differentiate from more common sellar/suprasellar tumors. The presentation of recurrent leukemia as a solid homogeneous mass in the sellar region is relatively rare and is asymptomatic in most cases. When symptomatic, common presentations include headache, diabetes insipidus, and visual disturbances. Bilateral hemianopsia is the most common visual field impairment due to compression of the optic chiasm. If tumor infiltrates into the cavernous sinus, patients may present with cranial neuropathy.⁵ On MRI, leukemic infiltration may appear circumscribed or irregular, enhances avidly, may demonstrate thickening of the infundibulum, and most importantly, shows rapid growth on interval exams. Some clues which are particularly suspicious for malignancy include rapid growth with sudden onset of symptoms, especially in the setting of cranial nerve VI palsy, since it is typically well sheltered within the cavernous sinus; its involvement indicates advanced tumor infiltration.

Langerhans Cell Histiocytosis (LCH).

LCH is a rare disease of uncertain etiology which typically presents in children under two years of age, is more common in boys, and may involve any organ system. There is an inverse relationship between the severity of the disease and the patient’s age at the time of presentation. The most common form of intracranial involvement includes infiltration of the hypothalamic-pituitary axis, resulting in hypopituitarism and diabetes insipidus, with or without lytic calvarial lesions.⁶ On MRI, the infundibular stalk is thickened, slightly T2 hyperintense, and avidly enhances.⁷ There is often absence of the posterior pituitary bright spot on pre-contrast T1 sequences.

Diagnosis

Recurrent Leukemia

Summary

In summary, the differential diagnosis for CNS tumors is based upon several factors, to include location, age, sex, imaging characteristics, and patient demographics. When presented with an enhancing sellar/suprasellar mass in an adolescent, the most common etiologies include craniopharyngioma, chiasmatic-hypothalamic glioma, and germ cell tumor. Leukemia should be a consideration in a patient with a history of prior or current disease, especially if the lesion demonstrates rapid growth on subsequent imaging exams. Magnetic resonance imaging is the primary modality of choice for characterization of masses in the sellar/suprasellar region; CT plays a complimentary role in evaluating for calcifications.

The views expressed in this material are those of the author, and do not reflect the official policy or position of the U.S. Government, the Department of Defense, or the Department of the Air Force.

References