Case Report:
A Subacute New Daily Persistent Headache in a Young Man

Subacute new daily persistent headache can be challenging to diagnose.

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A 29-year-old male without significant past medical history presented with a subacute headache. He was initially seen one week prior in the emergency room for a headache lasting four weeks, which began while working in an oil field. He described it as pressure behind his left eye radiating from the left frontotemporal region to the top of his head and sometimes to his occiput. It was severe in nature, nonpulsatile, with associated phonophobia, photophobia, nausea, and decreased left facial sensation. He was taking 2400mg of ibuprofen daily until the pain became unbearable.

He presented to his PCP who treated him for what he thought was sinusitis with antibiotics and steroids. The steroids initially helped but later the headache returned with the same quality and severity. The patient was evaluated two times in the ED, subsequently, and both times a phenergan and Depakote cocktail resolved the pain temporarily. On the second ED visit, a LP and MRI/MRA were performed and were normal.

The patient returned seven days later with the same quality headache but was also noted to have horizontal diplopia on exam. He was given amitriptyline and sumatriptan and asked to follow up with his neurologist. Two months later he presented to the ED with horizontal diplopia and no resolution to his headache. His physical exam was significant for left gaze palsy in his left eye, left eye ptosis, and numbness on the left side of his face. No other focal weakness or paresthesias were present.

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Repeat MRI showed an extra-axial left cavernous sinus mass extending into the foramen ovale measuring 19 x 6 mm, possibly consistent with meningioma or lymphoma. Subsequently, a LP was performed and showed monocytes and rare lymphocytes with no atypical or malignant cells. Five days later, he underwent a CT-directed needle biopsy of his left foramen ovale mass. The final pathologic diagnosis was non-diagnostic due to a poor specimen sample.

His symptoms continued to worsen, despite being treated with multiple medications including steroids. He was readmitted four days later, and underwent image-guided left-sided craniotomy for an excisional biopsy of his cavernous sinus lesion. The pathology was reported as meningioma WHO grade 1. Biopsy of his dura of the sphenoid wing was reported as showing no neoplasm or

malignancy. Intraoperatively, the lesion appeared to be en plaque meningioma spread along the surface of the dura; the dural biopsies did not confirm this.

Approximately 20 percent of the mass was resected. He will receive radiation via cyber knife for the remainder of the mass. His ptosis has improved, as well as his diplopia and facial sensation. He experiences significant headache, partially controlled with Lyrica and opiates.

DISCUSSION

Meningiomas are the most common extra-axial brain and spinal tumors, although lymphomas, sarcomas, metastatic tumors, schwannomas, hemangiopericytomas and inflammatory masses also occur adjacent to the brain and spinal cord.

WHO classification of meningiomas includes 13 morphological types and three grades: Grade I: Meningioma Grade II: Atypical Meningioma Grade III: Anaplastic Meningioma. The patient in this case had a Grade I meningioma which comprise 20 to 25 percent of all intracranial tumors. There exists a female to male ratio of 2:1 in adults, and nearly 10:1 are found in the spinal cord versus the brain. Most are benign, roughly 80 percent, but a subset is aggressive with high grade histology, high recurrence rates, and substantial morbidity and mortality.[1]

Locations of meningiomas are usually along the cerebral convexity and, to a lesser extent, in the parasagittal region, sphenoid wing, parasellar region and spinal canal. Loss of chromosome 22 is associated with multiple meningiomas. This can be seen in patients with NF 2 who also have loss of chromosome 22. Meningiomas involving the cavernous sinus may start in the sinus, or grow into it as part of a larger tumor involving the medial sphenoid wing, orbit, other areas of the middle fossa, clivus, or petrous bone as seen in this case. Multiple cranial nerve deficits (II, III, IV, V, VI) can be involved, causing a variety of abnormalities.[4]

Our patient developed diplopia, external opthalmoparesis, ptosis, and hemi-facial numbness; explained by involvement of cranial nerves III, VI, V and VII. Generally, the extent of surgical resection and histological grade represent the most important prognostic variables. The extent of the tumor is defined by enhanced MRI. Calcifications can be seen on CT or GRE. If a major surgical procedure is planned, angiography is needed to define the tumor vasculature, with the option to embolize supplying blood vessels including the internal carotid and meningeal vessels.[3] The 10-year recurrence rate for total resection (Simpson grade 1) is 9 percent, whereas for subtotal resection the rate is 40 percent. There is very limited data on recurrence by location in the cavernous sinus; most of the data suggests high rate or recurrence, probably from difficulty of

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complete resection.[1] Our patient's meningioma had the characteristic "en plaque" appearance seen intraoperatively. This is a pattern of diffuse carpet-like tumor spread along the dural surface, which does not affect prognosis.[5] The decision regarding treatment is often difficult because the symptoms may be mild or non-progressive. The natural history in some patients may be one of minimal or no growth for long periods of time.[4] There is risk of significant cranial nerve morbidity with surgical treatment, and the long-term results of new surgical treatments and radiation therapy modalities are unknown.

Gamma knife surgery is a safe and effective treatment, but high dose radiation could lead to modifications of the vascular wall leading to unintentional occlusion of the affected vessel.[6] At the present time, surgery is indicated in younger patients with worsening symptoms.[2] Radiation therapy is used when there is re-growth following subtotal removal and in older patients with worsening symptoms. Patients of any age with non-progressive or mild symptoms are observed. Given the age of this patient and the subtotal resection (20 percent), he will receive stereotactic radiation. ■

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