A 41-year-old woman who had been otherwise healthy presented with several months of worsening headaches, expressive aphasia and decreased visual acuity. No constitutional symptoms. She had no history of IV drug use, smoking, or sexual promiscuity. Physical exam revealed right sided homonymous hemianopia. She did not have gait instability or sensory deficits. Viral serology was negative for EBV and HIV. Figure 1 (A) shows CT brain revealing a 4.5 cm left occipito-parietal lobe lesion with moderate mass effect. MRI further qualified a 6 x 5 x 4.6 cm extra-axial mass arising from the left tentorium. Patient underwent uncomplicated left occipital craniotomy with complete resection achieved. Histologic examination (Figure 1, B, C, D) revealed a grade 2 spindle cell carcinoma with immunostaining positive for vimentin, desmin, PR, and Bcl-2. Staging CT of chest, abdomen and pelvis were negative for other organ involvement. Patient received adjuvant radiation therapy of 60 Gy in 30 fractions. Adjuvant chemotherapy was not pursued by patient. Nine months later, patient developed worsening headache and mental status changes. Brain MRI revealed increase mass effect with deep water edema suspicious for recurrence of tumor. Salvage craniotomy with left occipital lobectomy was performed, post neurosurgical complications included Klebsiella meningitis with bone flap infection. Bone flap debridement and IV antibiotics followed. Three months later, 12 months after initial resection, right arm weakness, spasms, and aphasia developed. Imaging was consistent with radiation necrosis which was managed with two cycles of bevacizumab and decadron taper. Outside of mild expressive aphasia, patient continues to follow with medical oncology.

Here, we examine a case of primary intracranial LMS and examine characteristics of prior presentations reported in the literature. In adults, intracranial LMS more commonly manifests as a result from hematogenous spread from primary sites such as GI, uterus, and subcutaneous tissue. Often a mimicker of meningiomas, immunohistochemistry reveals elongated tumor cells with tapering cytoplasmic processes. Staining is used to further aid in differentiation. Our case was strongly positive for vimentin and desmin, indicating that our sample was a sarcoma equipped for muscle differentiation. Currently there is no reported standard treatment for primary intracranial LMS. Surgery, chemotherapy and adjunctive radiation therapy has remained the mainstay. Chemotherapy aimed at sarcoma has included doxorubicin, ifosfamide, gemcitabine, cyclophosphamide, and doxetaxel. Based on published case studies however, chemotherapy is not often pursued. Regardless, prognosis remains poor with the longest survival being 8 years after diagnosis.

### References