

Primary Central Nervous System Angiosarcoma

Angiosarcoma is an infrequent tumor among **sarcomas**, especially presenting as a **primary tumor** within the **central nervous system**, which can lead to rapid neurological deterioration and death in few months.

Literature review

Mena et al. reported in 1991 eight patients with primary angiosarcoma of the central nervous system these included five males and three females ranging in age from 2 weeks to 72 years (mean 38 years). Of the eight neoplasms, six were located in the cerebral hemispheres and one was in the meninges; the site was unknown in the other. All patients underwent surgical resection. Five of the eight patients died, four within 4 months after surgery and one after 30 months. Two of the remaining three patients were 17 and 27 years old at the time of diagnosis and were alive at follow-up review 39 and 102 months after surgery, respectively. One patient was lost to follow-up monitoring. Microscopically, all eight tumors demonstrated a well-differentiated pattern with irregular vascular channels and intraluminal papillae; in addition, four showed poorly differentiated solid areas. Immunohistochemical staining of neoplastic cells to factor VIII-related antigen and Ulex europaeus agglutinin I was performed in five tumors and was focally positive in four. No correlation could be shown between the histological features and the growth and biological behavior of the tumors ¹⁾

Case reports

Valera-Melé et al. presented a 41-year old man with a right frontal enhancing hemorrhagic lesion. Surgery was performed with histopathological findings suggesting a primary central nervous system angiosarcoma. He was discharged uneventfully and received adjuvant **chemotherapy** and **radiotherapy**. At 5 months, the follow-up MRI showed two lesions with an **acute subdural hematoma**, suggesting a relapse. Surgery was again conducted finding tumoral membranes attached to the internal layer of the dura mater around the right hemisphere. The patient died a few days later due to the recurrence of the subdural hematoma. This case report illustrates a rare and lethal complication of an unusual tumor. The literature reviewed shows that gross-total resection with adjuvant radiotherapy seems to be the best treatment of choice ²⁾.

Gao M, Li P, Tan C, Liu J, Tie X, Pang C, Guo Z, Lin Y. **Primary Central Nervous System Angiosarcoma**. World Neurosurg. 2019 Dec;132:41-46. doi: 10.1016/j.wneu.2019.08.128. Epub 2019 Aug 27. PubMed PMID: 31470162 ³⁾.

report a case of intracranial angiosarcoma in a Caucasian male and present a review of the imaging features in the recent literature. The tumor mostly presents as a well-demarcated, heterogeneous,

moderately to strongly enhancing lesion with signs of intratumoral bleeding and surrounding vasogenic edema. The differential imaging features of common hemorrhagic intracranial tumors are discussed ⁴⁾.

Two cases of primary angiosarcoma of the brain are well characterized by imaging, histopathology, and immunohistochemistry. Case 1: The first patient was a 35-year-old woman who developed exophthalmos. Subtotal resection of a left extra-axial retro-orbital mass was performed.

Case 2: our second patient was a 47-year-old man who presented with acute visual loss, word-finding difficulty, and subtle memory loss. A heterogeneously-enhancing left sphenoid wing mass was removed. We also review the literature aiming at developing a rational approach to diagnosis and treatment, given the rarity of this entity.

Gross total resection is the standard of care for primary angiosarcoma of the brain. Adjuvant radiation and chemotherapy are playing increasingly recognized roles in the therapy of these rare tumors ⁵⁾.

A case of 17-year-old man had [pineal region](#) angiosarcoma and a [hepatic angiosarcoma](#). The patient's initial symptom was headache and a CT scan revealed a high-density mass in the pineal region with [obstructive hydrocephalus](#). After the radiation therapy, the tumor disappeared completely on the CT scan. One year later, he developed [abdominal pain](#), and a CT scan and angiogram revealed multiple angioma-like lesions in the [liver](#). The mass in the pineal region showed concomitant regrowth, and finally, the patient died of abdominal hemorrhage. At autopsy, the pineal tumor and hepatic tumor were both [angiosarcomas](#), although it was uncertain which was the original tumor ⁶⁾.

References

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