Hemangiopericytoma Misdiagnosed for Anaplastic Ependymoma

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INTRODUCTION
Anaplastic ependymoma is a grade III malignant tumor of the central nervous system (CNS). This type of tumor arises from ependymal cells lining the ventricles. Hemangiopericytoma is a neoplastic proliferation of the dura classified as a mesenchymal neoplasm. Both tumors have high rates of recurrence and can have metastasis. Here, we describe a case of misdiagnosis of hemangiopericytoma.

METHODS
Patient records were collected and depersonalized to exclude patient identification. Subsequently, all files relating to the patient’s care were surveyed – including various pathological specimens over more than a decade.

RESULTS
A 38 yo male initially presented with focal neurological deficit due to a large intracranial tumor which was subsequently surgically resected. Initial histology was felt to be positive for GFAP and vimentin and negative for EMA, cytokeratin, and CD34. This specimen reportedly showed areas of perivascular pseudorosettes as well. At the time, these findings were considered consistent with malignant ependymoma. Over time the patient developed multiple intracranial recurrences as well as one distant metastasis to the right L2 pedicle. Neuropathological re-evaluation of these recurrent tumors as well as of the initial tissue suggested an alternative diagnosis of hemangiopericytoma. More recently biopsied tissue demonstrates high cellularity with large staghorn vascular spaces, possible weak positivity for CD34 staining (immunohistochemistry) but strong positivity for reticulin staining (EM).

DISCUSSION
According to the initial pathology the tumor was positive for GFAP (glial marker) and had pseudorosettes. However, subsequent re-evaluation at recurrence of new and original tissue showed GFAP staining of gliotic brain tissue only at the periphery of the tumor and no pseudorosettes. Clinically, delayed distant metastases are more commonly reported with meningeal hemangiopericytoma compared to ependymoma.

CONCLUSION
It is critical to recognize similarities and differences between neoplastic processes to maximize outcome. This report demonstrates the critical need to remain open to alternative diagnosis despite having a final diagnosis.

REFERENCES

DISCLOSURES
I DO NOT have any financial or organizational relationships with commercial interests or other entities. I hereby certify that to the best of my knowledge, no aspect of my current personal or professional circumstances places me in the position of having a conflict of interest with my duties, responsibilities and exercise of independent judgement as an Officer, Member of the Board of Directors, Nominee for Office, Educational Presenter and/or a representative of AANS/SF/NA.

Figure 1: A: GFAP positivity at the periphery of the tumor staining normal brain. B: Staghorn vascular spaces surrounded by a highly cellular spindle tumor.

Figure 2: Staining with reticulum shows encircling individual tumor cells or clusters of tumor cells.

Figure 3: A: H&E staining from L2 metastasis similar to original tumor. B: Positive Vimentin staining indicating mesenchymal tumor.

Figure 4: A: Original brain tumor at diagnosis with diffuse homogenous enhancement. B: Distant recurrence in the right L2 pedicle 10 years after first diagnosis.