Common Mesenchymal Neoplasms in and around the CNS

David Meredith, MD, PhD

Case-Based Questions (please see page 3 for answers)

1.	The best predictor of clinical behavior in leptomeningeal solitary fibrous tumors (SFTs) is:	
	a.	Absence of *STAT6* fusion event
	b.	Brain invasion
	c.	Cellularity
	d.	Mitotic rate

2.	Poorly differentiated chordomas can be best distinguished from conventional chordomas by immunohistochemistry for which of the following?	
	a.	Brachyury
	b.	INI-1
	c.	Pan-keratin
	d.	S100

3.	Which genomic alteration in meningioma is most correlated with aggressive behavior?	
	a.	*CDKN2A/B* deletion
	b.	Deletion of 1q
	c.	Deletion of 22q
	d.	Gain of chromosome 4

Scroll to Page 3 for answers

Question 1 Correct answer and rationale: **D) Mitotic rate**

A) As much as 1/3 of meningeal SFTs may show no detectable *NAB2-STAT6* rearrangement. While this finding is more commonly observed in malignant SFTs, tumors of all grades may lack the characteristic fusion product, and, therefore, fusion status is not a reliable grading criterion.

B) Unlike in meningiomas, brain invasion is not considered a risk factor for more aggressive behavior in SFTs.

C) Despite being included as a grading criterion for SFTs in the 2016 CNS WHO, tumor cellularity alone has *not* been reliably shown to affect progression-free or overall survival and was dropped in the current 2021 CNS WHO.

D) In both soft tissue and CNS locations, a mitotic rate greater than 4/10 high power fields has repeatedly been shown to increase risk of local recurrence and distant metastasis, leading to dramatic reduction in survival rates.

Question 2 Correct answer and rationale: **B) INI-1**

The immunoprofile for poorly differentiated chordoma shows considerable overlap with conventional chordoma with uniform keratin and brachyury positivity and variable S100 staining; however, poorly differentiated chordoma is defined by its loss of expression for INI-1, which is not seen in conventional chordoma.

Question 3 Correct answer and rationale: **A) *CDKN2A/B* deletion**

Focal deletions of *CDKN2A/B* are highly correlated with anaplastic meningioma, WHO grade 3 and are rarely encountered in lower histologic grade tumors. Deletion of 22q (including *NF2)* is exceedingly common in meningiomas of all grades, and deletion of 1q and gains of chromosome 4 have no known prognostic value.