Disclosures

I have no financial relationships / conflicts of interest to disclose
Clinical History

57 year-old male who presents with headache and altered mental status
- History of NSCLC status post pneumonectomy and whole brain XRT for brain metastases, 8 yrs prior to presentation

Neurologic exam:
- Follow commands
- Left facial droop, left hemiplegia, and left sided neglect with right gaze preference
Radiologic Studies

- Head CT: Old lacunar infarct but no acute hemorrhage

- Brain MRI:
  - Diffuse right hemisphere cortical expansion with abnormal T2/FLAIR signal and associated restricted diffusion.
  - Diffuse leptomeningeal enhancement in the right cerebral hemisphere
Brain MRI. Left: Axial T1 Post contrast, Right: Axial Flair T2
A biopsy was performed of the “Right frontal lesion”
trichrome
GFAP
Diagnosis

Brain tissue with vasculopathy and severe gliosis, see note.

– Changes of capillary fibrosis and endothelial reactive atypia are likely secondary to prior radiation therapy.

– Given the clinical history and neuroimaging characteristics, the entity known as SMART syndrome (stroke-like migraines after radiation therapy) should be considered
Patient follow-up

- 6 months post biopsy, the patient shows significant improvement
  - Cognitive
  - Motor function
- No longer using a wheelchair; walks well with a walker, transitioning to a cane
- Follow-up MRI demonstrated radiation related changes
Final Diagnosis:

SMART SYNDROME
SMART Syndrome

- Episodes of reversible neurological dysfunction
  - Migraine headache + aura
  - Stroke-like deficits (aphasia, hemiparesis, hemisensory deficits, homonymous hemianopsia)

- Remote history of external beam cranial XRT (dose > 50 Gy)

- Transient, diffuse unilateral cortical gadolinium enhancement on MRI within previous radiation field
Pathophysicsology – SMART-ER?

Pathologic substrate for SMART has not been well-described

– Gliosis

Pathophysiology poorly understood
Hypothesis: Cerebral hyperexcitability with impaired autoregulation, perhaps due to endothelial damage

Pruitt A et al Neurology 2006
References


