SMART syndrome mimicking glioblastoma recurrence 6 years after surgical excision and radiotherapy: A Case Report

Authors:

- POON Wui Chung, LEE Wing Yan Michael, PANG Kai Yuen Institution:
- Department of Neurosurgery, Pamela Youde Nethersole Eastern Hospital, Hong Kong.





Background

Stroke-like migraine attacks after radiation therapy (SMART) syndrome

- a rare, reversible syndrome that occurs in patients having received radiotherapy for intracranial neoplasms.

- occurs quite a few years after intracranial irradiation.

- patients typically present with headache, seizures, or other focal neurological deficits mimicking disease recurrence or sub-acute stroke.

The exact mechanism of SMART syndrome: unknown.

Preliminary investigations suggested post-radiation neuronal dysfunction may be the underlying mechanism.¹

Case illustration



- 35-year-old gentleman, table tennis coach
- Admitted to our unit with malignant and vascular right parieto-occipital tumour in MRI presenting with left sided headache in 2014.
- Craniotomy and excision of brain tumour in 2014.
 - Pathology: glioblastoma multiforme, negative for MGMT.
- Gliolan fluorescence guided excision 2 weeks after the first operation due to residual tumor in post-operative MRI.
- Post-operative chemoradiotherapy and adjuvant temozolomide were completed in 2015.
- Patient survived till today 6 years after the operation at the age of 41, and worked as a table tennis coach still.
- He presented to us in May 2020 with headache, fever and transient left upper limb weakness. His symptoms resolved in weeks, and he was able still to work.



MRI in May 2020 - gyriform enhancement pattern over right cerebrum, worrisome of tumour recurrence



MRI in Aug 2020 - resolved abnormal gyriform signal change and swelling in the right cerebrum, no tumour recurrence

Discussion

	SMART syndrome	Stroke	Recurrent tumour
Clinical features	 History of brain irradiation for cancer Partial/complete reversal of symptoms 	 Cerebrovascular risk factors such as atrial fibrillation, carotid stenosis, hypertension, hyperlipidaemia Acute symptoms 	- History of brain irradiation for cancer - Progressive symptoms
Typical MRI features	Increased T2 hyperintensity and gyral enhancement in the previously radiated area	 Intravascular thrombus Vascular hyperintensity on FLAIR Vascular territory in MRI finding 	- Well circumscribed lesion - Increased perfusion in the affected area
Short-term follow-up MRI	Partial/complete reversal of MRI findings	Persistence of the abnormal signal	Persistence of the abnormal signal

Conclusion

- The occurrence of SMART syndrome as a rare delayed complication of brain tumour irradiation may be potentially misdiagnosed as tumour recurrence or stroke.
- Given its self-limiting nature, early recognition of this syndrome is paramount.
- It has a favourable response to conservative treatment with complete or partial reversal of their radiological findings within months.
- Proper recognition of this syndrome can prevent invasive diagnostic techniques such as brain biopsy or vascular imaging.²
- There is **no proven method for treatment of SMART syndrome**.³ Some authors suggest steroid pulse therapy may provide speedy recovery from and diagnosis of SMART syndrome, and it should be considered before invasive investigations. ⁴

~The End~

References:

¹ Farid K, Meissner WG, Samier-Foubert A, et al. Normal cerebrovascular reactivity in Stroke-like Migraine Attacks after Radiation Therapy syndrome. Clin Nucl Med. 2010 Aug;35(8):583-5. doi: 10.1097/RLU.0b013e3181e4db6f. PMID: 20631504. ²Daniel April, MD,1 Neil Lall, MD,2 Andrew Steven, MD1. Stroke-Like Migraine Attacks After Radiation Therapy

Syndrome. Ochsner Journal 20:6–9, 2020 DOI: 10.31486/toj.19.0090

³Zheng Q, Yang L, Tan LM, Qin LX, Wang CY, Zhang HN. Stroke-like Migraine Attacks after Radiation Therapy

Syndrome. Chin Med J 2015;128:2097-101.

4 Wenting Jia, Ryuta Saito, Masayuki Kanamori, Naoya Iwabuchi, Masaki Iwasaki, Teiji Tominaga. SMART (stroke-like migraine attacks after radiation therapy) syndrome responded to steroid pulse therapy: Report of a case and review of the literature. eNeurologicalSci 12 (2018) 1–4