Case 18235

Eurorad

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

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Section: Neuroradiology
Area of Interest: CNS Neuroradiology brain
Imaging Technique: MR
Special Focus: Cancer Case Type: Clinical Cases
Authors: Christina Bougia, Thomas Benekos, Persefoni Margariti, Anastasia Zikou
Patient: 52 years, male

Clinical History:

A 52-year-old man with history of metastatic testicular seminoma and adjuvant brain irradiation therapy (21 years ago) with recurrent seizures during the last 11 years presented with aphasia, altered mental status of consciousness and hemiparesis of the right upper and right lower limb, with referred symptoms' initiation 6 days ago.

Imaging Findings:

Cortical thickening of the parieto-temporo–occipital regions of the left cerebral hemisphere associated with mild abnormal high signal intensity of the affected cortex on T2 and FLAIR images (Figures 1, 2), intense gyriform post-gadolinium enhancement and coexistent leptomeningeal enhancement (Figure 3). Hyperperfusion of the affected areas was also seen on MR perfusion images (Figure 4). Follow-up brain MRI after 6 months demonstrated complete resolution of the cortical oedema, normal perfusion and absence of gadolinium enhancement of the affected areas (Figures 5, 6).

Discussion:

SMART syndrome is a rare delayed complication following cranial irradiation therapy in patients with central nervous system malignancies or brain metastatic disease. The syndrome typically presents as recurrent complex migraine attacks with reversible, unilateral focal neurologic signs such as hemiparesis, seizures and aphasia [1, 2]. Patients typically present with SMART syndrome between 1 to 37 years after cranial irradiation therapy. The exact mechanisms of SMART syndrome remain unknown; however, it has been associated with cerebral hyperexcitability, intrinsic impairment of the autoregulatory mechanisms, vascular endothelial damage and impaired blood-brain barrier integrity as a consequence of irradiation damage [2]. Development of the SMART syndrome has been correlated to a radiation dose of at least 50 Gy [4]. The diagnosis of SMART syndrome involves the combination of both clinical semiology and brain MRI specific findings. Imaging hallmarks of SMART syndrome include; prominent unilateral cortical thickening (defined as gyriform abnormal high signal intensity on T2 and FLAIR sequences) associated with mild mass effect due to oedema, intense post-gadolinium gyral enhancement and hyperperfusion of the affected areas [3, 4, 5]. The characteristic gyriform cortical enhancement typically develops within 2–7 days and usually resolves in 13–35 days [1]. All these imaging findings are usually confined to the irradiated areas and preferentially in the posterior areas of the brain parenchyma. Typically the subcortical and deep white matter structures lack involvement [1]. Restricted diffusion or mild leptomeningeal enhancement may or may not be seen [3, 4]. Imaging findings may appear 2–7 days after symptoms' onset and typically resolve after 2–5 weeks[3].
As a permanent sequelae of SMART syndrome, cortical laminar necrosis may be seen [3]. Quite common accessory imaging findings include multiple magnetic susceptibility foci compatible with cavernous malformations (>5 mm) and/or microbleeds (<5 mm) as well as imaging findings consistent with leukoencephalopathy, both as sequelae of prior radiotherapy [2, 3].

Differential Diagnosis List: SMART Syndrome, Ischaemic Stroke, Post-ictal Changes , Posterior Reversible Encephalopathy Syndrome (PRES), Peri-ictal Pseudoprogression, Nonconvulsive Status Epilepticus

Final Diagnosis: SMART Syndrome

References:

Dominguez M, Malani R (2021) Stroke-Like Migraine Attacks After Radiation Therapy (SMART) Syndrome: A Comprehensive Review. Curr Pain Headache Rep 25:33 (PMID 33761013)
Singh TD, Hajeb M, Rabinstein AA, Kunchok AC, Pittock SJ, Krecke KN, Bartleson JD, Black DF (2021) SMART syndrome: retrospective review of a rare delayed complication of radiation. Eur J Neurol 28(4):1316-1323 (PMID 33159349)
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Kerklaan JP, Lycklama á Nijeholt GJ, Wiggenraad RG, Berghuis B, Postma TJ, Taphoorn MJ (2011) SMART syndrome: a late reversible complication after radiation therapy for brain tumours. J Neurol 258:1098-1104 (PMID 21373901)
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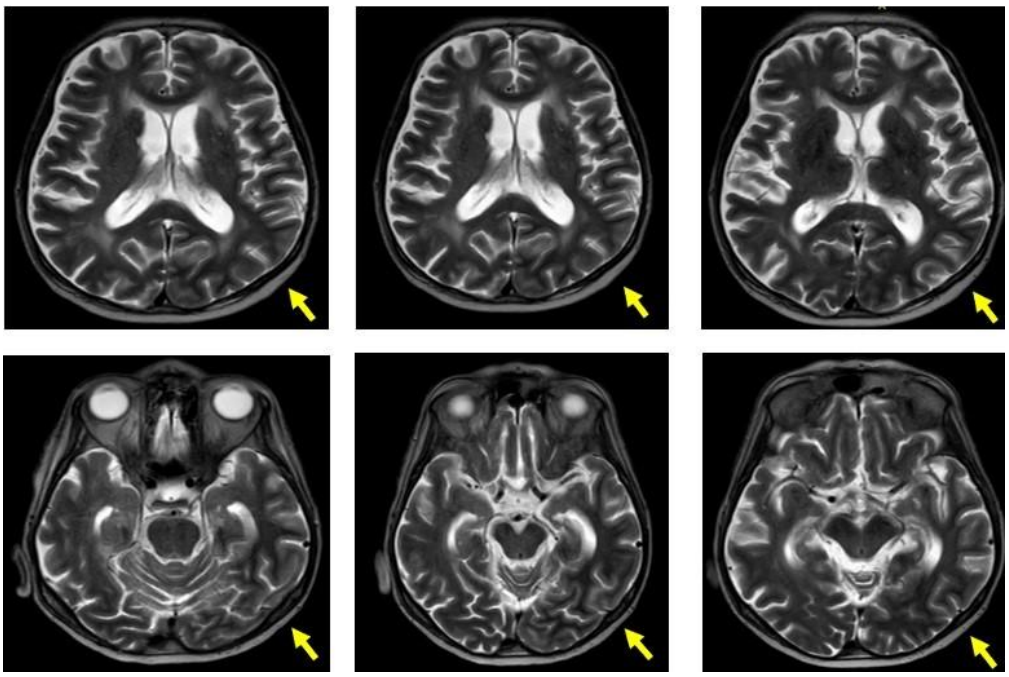
Figure 1

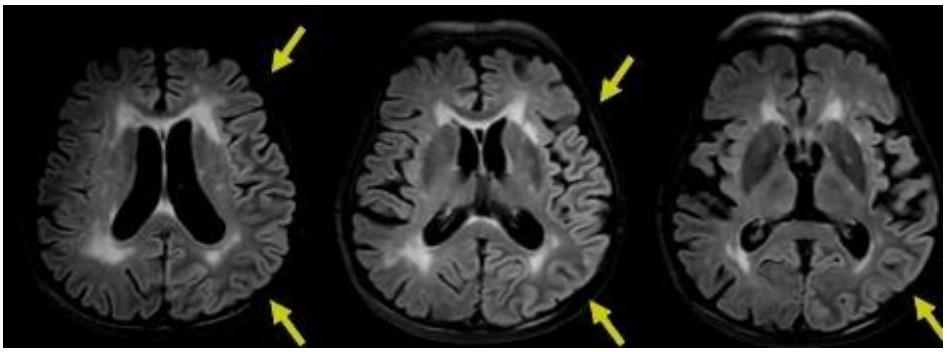
a



Description: Axial T2 weighted images showing mild thickening and high intensity signal of the cortex of the left parieto-temporo–occipital cerebral lobes (arrows) **Origin:** Department of Radiology, University Hospital of Ioannina, Greece

Figure 2

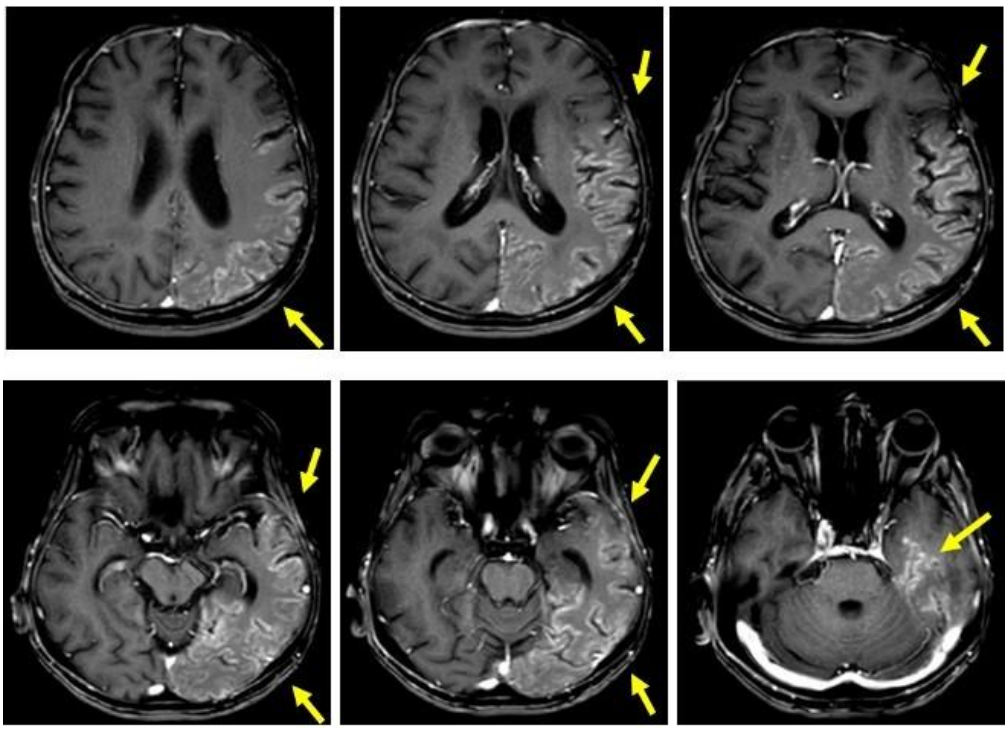
a



Description: Axial FLAIR images showing thickening and high intensity signal of the cortex of the left parieto-temporo–occipital cerebral lobes (arrows) **Origin:** Department of Radiology, University Hospital of Ioannina, Greece

Figure 3

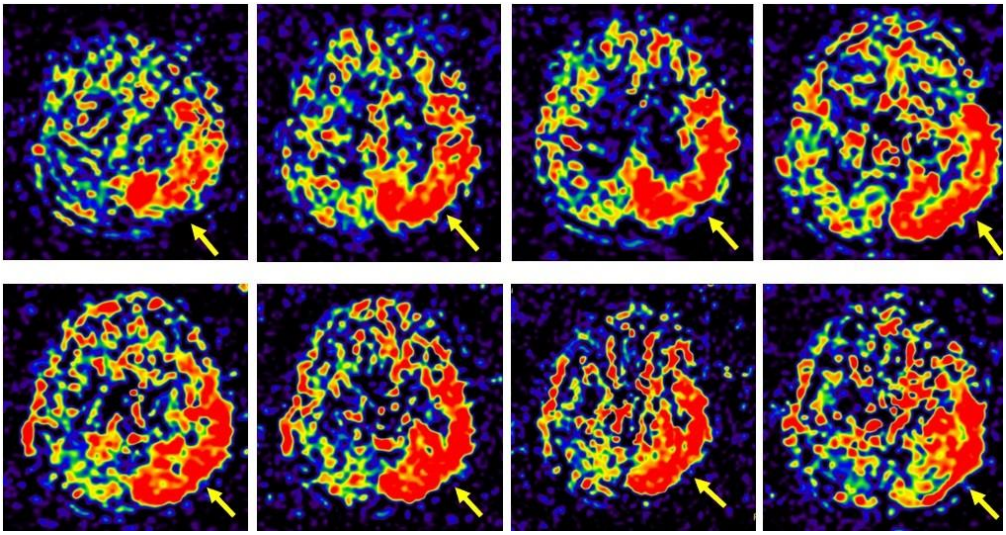
a



Description: Axial T1 weighted images after the injection of gadolinium showing intense gyriform leptomeningeal enhancement of the affected areas (arrows) **Origin:** Department of Radiology, University Hospital of Ioannina, Greece

Figure 4

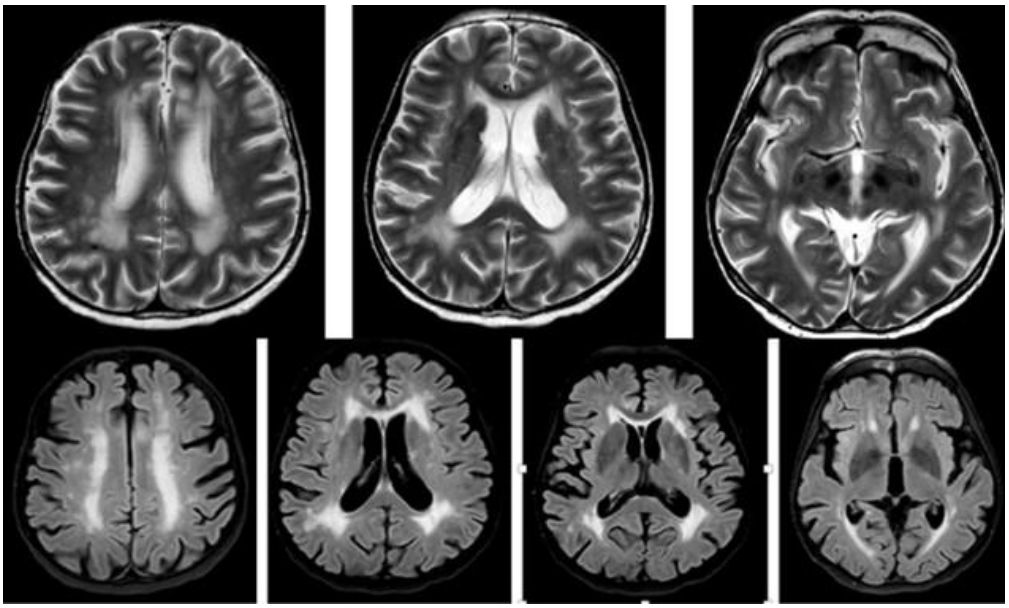
a



Description: Arterial Spin labeling sequences displaying increased perfusion of the left parieto-temporo–occipital regions that are affected (arrows) **Origin:** Department of Radiology, University Hospital of Ioannina, Greece

Figure 5

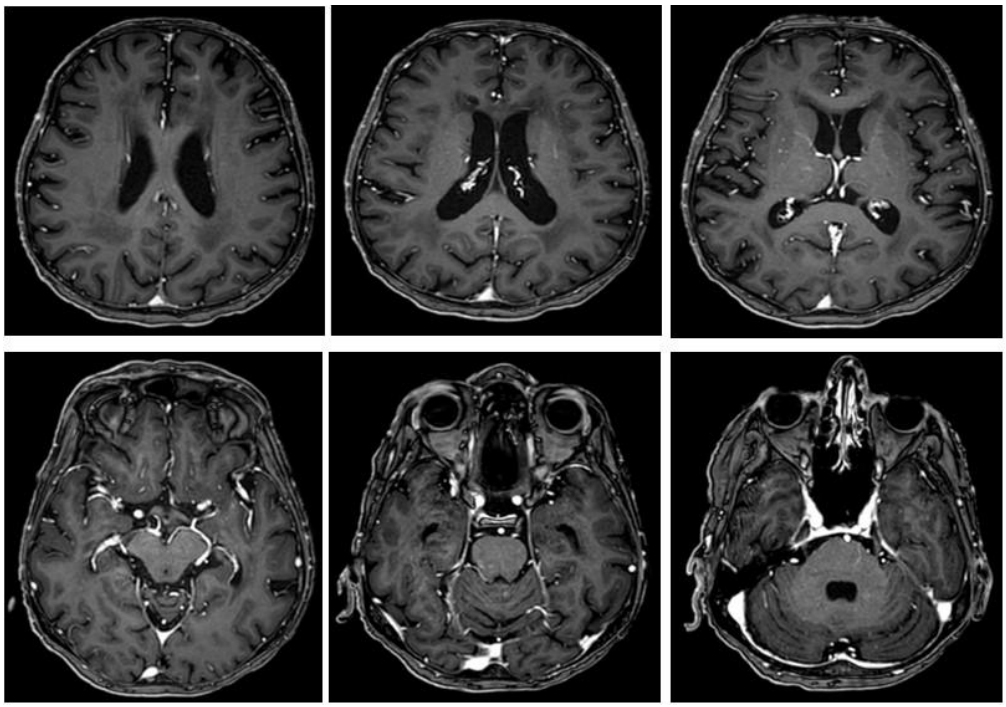
a



Description: Axial T2 weighted and FLAIR images demonstrate complete resolution of the cortical oedema at 6 months follow up **Origin:** Department of Radiology, University Hospital of Ioannina, Greece

Figure 6

a



Description: Axial T1 weighted images with intravascular gadolinium showing complete resolution of the leptomeningeal enhancement of the previously affected areas **Origin:** Department of Radiology, University Hospital of Ioannina, Greece