Case 14978

SMART syndrome (stroke-like migraine attacks after radiation-therapy)
Published on 22.09.2017

DOI: 10.1594/EURORAD/CASE.14978
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Diagnostic procedure
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: Digital radiography
Imaging Technique: MR
Special Focus: Seizure disorders Case Type: Clinical Cases
Authors: Laura Koren, Amaya Hilario, Patricia Martin, Elena Salvador, Ana Ramos
Patient: 32 years, female

Clinical History:

32 year-old woman was admitted to the hospital with headache, right homonymous hemianopsia and weakness in the right upper limb. She had a history of pineal germinoma treated with surgery and whole-brain radiotherapy 24 years before. An EEG done at presentation demonstrated left hemisphere slowing with no epileptogenic activity.

Imaging Findings:

MRI showed thickening, increased T2 signal and gyriform contrast enhancement of the parieto-occipital left cortex. Diffusion-weighted image demonstrated T2 shine through without real evidence of restricted diffusion on de ADC map.

Ancient ischaemic stroke in the right MCA territory of cryptogenic aetiology and sequelae of frontal bilateral external ventricular shunt were also present.

There was no follow-up MRI but the patient received corticosteroid treatment and her symptoms improved when she was discharged.

Discussion:

SMART syndrome is a delayed and recurrent complication of whole-brain radiation therapy that demonstrates characteristic imaging findings. Development of the SMART syndrome has been related to a radiation dose of at least 50 Gy and it can occur even 35 years after radiotherapy [1-3].

The pathophysiology of the SMART syndrome is poorly understood. Several theories have been proposed including neuronal and endothelial damage/dysfunction, vascular instability, and vasospasm leading to hypoxia/ischaemia[2]. A specific vulnerability of the parieto-occipital cortex for radiation or chemotherapy, similar to that observed in PRES, may explain why the imaging findings are preferentially observed in this region [2-4].

Attacks are typically sub-acute in onset and involve stroke-like deficits such as homonymous hemianopsia, hemiplegia, aphasia, migraine headaches and/or seizures [1]. This entity has been classically described as a benign
condition that resolves without sequelae but more recent publications refer incomplete recovery with residual neurologic deficits in up to 45\% of cases [1].

MR imaging findings include thickening, increased T2 signal and gyriform contrast enhancement of the affected cortex. Diffusion-weighted abnormalities are minimal and primarily demonstrated T2 shine through without real evidence of restricted diffusion.

The findings are usually unilateral with prominent involvement of the temporo-parietal and occipital lobes and relative sparing of the frontal lobes [1, 3]. The affected areas are beyond the bounds of a single vascular territory, which helps to distinguish SMART syndrome from ischaemic stroke.

MRI findings are usually transient, beginning a few days after the onset of symptoms and then disappearing. However, some patients present cortical laminar necrosis in the affected areas as permanent sequelae [1]. Post-ictal MR imaging demonstrate findings similar to SMART. The absence of headache, less significant neurologic impairment and more rapid clinical recovery are differentiating aspects. Meningeal enhancement is frequent in peri-ictal changes and differs from the cortical enhancement seen in SMART [1].

The differential diagnosis includes post-ictal changes, infections, vascular diseases such as ischaemia, venous sinus thrombosis and arteriovenous fistulas, and posterior reversible encephalopathy [3, 5].

To sum up, SMART syndrome is a rare delayed complication of whole-brain irradiation characterised by episodes of reversible neurologic deficits, migraine-like headaches and seizures. MR imaging findings include a unilaterally increased T2 signal, thickening and gyral enhancement of temporal, parietal or occipital cortices that usually disappears as symptoms resolve. Recognition of this syndrome is important as it can help avoid invasive testing such as cerebral angiography or brain biopsy.

**Differential Diagnosis List:** SMART syndrome, Ischaemic stroke, Post-ictal changes, Posterior reversible encephalopathy, Arteriovenous fistula, Venous sinus thrombosis

**Final Diagnosis:** SMART syndrome

**References:**


Description: Axial T2-FLAIR MRI images demonstrating increased signal intensity in the left parieto-occipital cortex. Ancient ischaemic stroke in the right middle cerebral artery territory and sequelae of frontal bilateral external ventricular shunt are also present. Origin: Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
Description: Axial T2-FLAIR MRI images demonstrating increased signal intensity in the left parieto-occipital cortex. Ancient ischaemic stroke in the right middle cerebral artery territory is also present.

Origin: Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
**Description:** Axial T2-FLAIR MRI images demonstrating increased signal intensity in the left parieto-occipital cortex. Ancient ischaemic stroke in the right middle cerebral artery territory is also present.

**Origin:** Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
Description: Axial T2-FLAIR MRI images demonstrating increased signal intensity in the left parieto-occipital cortex. Ancient ischaemic stroke in the right middle cerebral artery territory is also present.

Origin: Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
Description: Diffusion weighted image shows hyperintensity in left parieto-occipital brain region without convincing evidence of restricted diffusion in ADC map. Origin: Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
Description: T1-weighted image of the brain following intravenous administration of gadolinium. Gyriform enhancement of the left posterior parieto-occipital cortex matching the T2 signal abnormality.

Origin: Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.
Description: Axial T2-weighted image shows thickening and increased signal of the affected cortex (star) in comparison with the normal one (arrow). **Origin:** Koren L, Department of Radiology, Hospital Universitario 12 de Octubre, Madrid, Spain.