Neurological Case Reports

Case Series
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Stroke like Migraine Attack after Radiation Therapy - A Classical Reversible Syndrome with an Unusual Unfavorable Outcome

Vera Montes, Catia Carmona, Sandra Sousa and Fernando Pita*

Department of Neurology, Cascais Hospital Dr. Jose de Almeida, Portugal

Abstract

Stroke like Migraine Attack after Radiation Therapy (SMART) syndrome is a rare and late complication of brain radiotherapy. It is characterized by episodes, usually reversible, of headache associated with focal neurological deficits, with typical changes in brain magnetic resonance imaging. We report here two cases of SMART syndrome in long term survivors of high grade brain tumor. Our two patients suffered multiple episodes of attacks characterized by headache, aphasia, and weakness or sensory disturbance. In both patients, the course of disease was unfavorable, and eventually leads to severe and permanent neurological damage. Our present two cases allow us to broaden the clinical spectrum of SMART syndrome, demonstrating that it may be presented as a reversible and benign form to an eventually irreversible form with a severe course and permanent disability. Although extremely rare, SMART syndrome should always be considered in patients with headache, recurrent neurological deficits and a history of cranial irradiation.

Keywords: Stroke-like migraine attack; Primary brain tumor; Radiotherapy; Headache

Introduction

Stroke like Migraine Attack after Radiation Therapy (SMART) syndrome was first described in 4 children by Shuper et al. [1] in 1995. It is a rare complication of brain radiotherapy that can appear between 2 and 10 years after radiotherapy. It is typically manifested by paroxysmal episodes of headache with migraine characteristics, focal neurological deficits, usually self limiting, and epileptic seizures [2]. Reversibility of symptoms is typical in SMART syndrome; however, rare cases with incomplete neurological recovery have been described [3]. The exact pathophysiology of SMART syndrome is unknown, proposing two mechanisms: 1) radiation-induced cortical hyper excitability, similar to the mechanism in hemiplegic migraine; 2) vascular instability with endothelial dysfunction and blood brain barrier disruption. As the clinical presentation is not specific, brain Magnetic Resonance Imaging (MRI) is imperative for diagnosis, usually with typical findings as leptomeningeal enhancement pattern after gadolinium with gyriform cortical hyper signal [4]. There is no effective treatment, and the proposed therapies (antiplatelet therapy, anticonvulsants and calcium channel blockers) have no proven benefit [5].

Case Series

Case 1

We examine a 47 years old man with a history of several hospitalizations in the last six years due to episodes of acute onset headache, aphasia and right hemiparesis, with full recovery within 48 h. Brain MRI showed signs of microangiopathic ischemic leukoencephalopathy. Complete vascular study that did not identify a specific etiology related to cerebrovascular disease. He also performed electroencephalograms, which did not reveal paroxysmal activity. The patient was discharged from these hospitalizations, always with full recovery, under antiplatelet, statin and antiepileptic therapy. His past medical history is significant for brain radiotherapy for central nervous system lymphoma at 13 years of age. Two months after last hospitalization he was admitted again for an episode of frontal headache associated with language impairment and right hemiparesis. Brain CT showed no acute parenchymal lesions and the electroencephalogram showed no paroxysmal activity. The transcranial Doppler was normal. Brain MRI with gadolinium showed bilateral frontal and parietal subcortical hyper signal on T2-weighted images. He was discharged 10 days later on aspirin and levetiracetam, with discrete right hemiparesis and non fluent aphasia. Despite antiepileptic

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Correspondence:
Vera Montes, Department of Neurology, Cascais Hospital Dr. Jose de Almeida, Av Brigadeiro Victor Novais Goncalves, 2755-009 Alcabideche Cascais, Lisbon, Portugal, Tel: 00351919626037; E-mail: montesveraluciasantos@gmail.com

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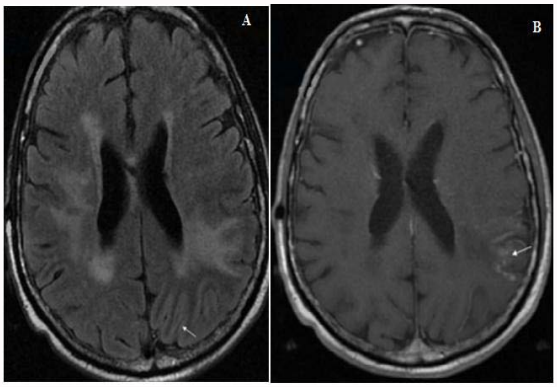

Figure 1: Brain magnetic resonance imaging A). Axial FLAIR T2-weighted sequence reveals left temporal gyriform cortical hyperintensity (arrow). B). Axial T1 weighted sequence with gadolinium shows leptomeningeal enhancement after gadolinium contrast (arrow).

treatment, the patient continued to experience paroxysmal headache events associated with language impairment and right hemiparesis, never returning to his baseline state.

Case 2

A 55 year old man with a history of cerebral astrocytoma at 38 years of age, who underwent brain radiotherapy, developed acute onset of altered mental status and language dysfunction. Over the past 5 years, he was experiencing frequent episodes of transient headache with migraine characteristics associated with aphasia, sensory disorder and altered mental status, interpreted as seizures. On neurological examination, he was confused and aphasic. There were no changes in the analytical study and arterial blood gas analysis. The cerebrospinal fluid study showed no changes and the electroencephalogram was normal. Contrast enhanced brain CT showed no acute parenchymal lesions or intracranial vessel caliber changes. Transcranial Doppler was normal, showing no suggestive changes of reversible vasoconstriction syndrome. Nevertheless, in the face of acute headache, neurological focal deficits and fluctuating state of consciousness of unidentified etiology, he began therapy with nimodipine, levetiracetam, and aspirin. Brain MRI showed left temporal gyriform cortical hyperintensity at T2 FLAIR and leptomeningeal enhancement after gadolinium contrast (Figure 1). He was left on antiplatelet and antiepileptic therapy. The reevaluation at 6 months, showed a slight improvement in the language, maintaining, however, little fluent speech and difficulty in naming and repeating. To date, he hasn't had any more episodes of transient neurological dysfunction.

Discussion

In the cases presented, the diagnosis of SMART syndrome was established after a long period of diagnostic investigation and follow-up. In both cases, the brain MRI study revealed characteristic alterations that allowed this diagnostic hypothesis to be proposed. SMART syndrome, although extremely rare, should always be considered in patients with paroxysmal phenomena characterized by headache, hemispheric dysfunction or epileptic seizures, with a remote history of cranial irradiation and typical imaging changes. The clinical spectrum of this entity is wide, ranging from typically benign reversible to severe and irreversible rare forms, as in the presented cases. Therefore, in order to establish the diagnosis of SMART syndrome, it is essential to exclude recurrence or progression of the tumor, stroke, reversible vasoconstriction syndrome and reversible posterior encephalopathy syndrome, due to overlapping of the clinical and imaging characteristics. The previous history of radiotherapy is a fundamental aspect in the diagnosis, allowing avoiding unnecessary invasive and therapeutic diagnostic measures.

References

1. Shuper A, Packer RJ, Vezina LG, Nicholson HS, Lafond D. Complicated migraine like episodes in children following cranial irradiation and chemotherapy. Neurology. 1995;45(10):1837-40.
2. Bartleson JD, Krecke KN, O´Neill BP, Brown PD. Reversible stroke like migraine attacks in patients with previous radiation therapy. Neuro Oncol. 2003;5(2):121-7.
3. Bompaire F, Zinchenko L, Lahutte M, Mokhtari K, Psimaras D, Gaultier C, et al. SMART syndrome: Classic transient symptoms leading to an unusual unfavorable outcome. Rev Neurol (Paris). 2017;173(1-2):67-73.
4. Pruit A, Dalmau J, Detre J, Alavi A, Rosenfeld MR. Episodic neurologic dysfunction with migraine and reversible imaging findings after radiation. Neurology. 2006;67(4):676-8.
5. Black DF, Morris JM, Lindell EP, Krecke KN, Worrell GA, Bartleson JD, et al. Stroke-like migraine attacks after radiation therapy (SMART) syndrome is not always completely reversible: A case series. AJNR Am J Neuroradiol. 2013;34(12):2298-303.