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STROKE LIKE MIGRAINE ATTACKS AFTER RADIATION THERAPY (SMART) VS RADIATION VASCULOPATHY

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ABSTRACT
Stroke like migraine attacks after radiotherapy (SMART) syndrome is a phenomenon of reversible neurologic dysfunction which occurs rarely years after radiotherapy for brain tumors. We present a 35years old patient who had new onset neurological dysfunction with fits twice in between a symptom free interval of one year. He had radiotherapy for grade 1 astrocytoma about 27 years ago. Tumor recurrence or growth was ruled out both times by neuroimaging. Alternate diagnosis like meningoencephalitis were also ruled out by appropriate workup. Patient was managed on antiepileptics and antiplatelets. Radiation induced vasculopathy was an important differential. The mechanism of both SMART and radiation induced vasculopathy appears similar. Having knowledge of such rare but important conditions is important for neurologist as it can save patients from unnecessary investigations.

KEY WORDS: Stroke, Migraine with aura, Radiotherapy

INTRODUCTION

Stroke-like migraine attacks after radiation therapy (SMART) is an extremely rare delayed complication of brain irradiation. Patients suffer from recurrent episodes of complicated migraine symptoms, consisting of transient neurologic deficits such as hemiparesis, aphasia, and sensory disturbances. 42 cases of SMART have been reported in the literature since it was first described in 1995.1
We present a case of young male who had episodes of recurrent neurological dysfunction with seizures.

CASE REPORT

A 35yearmale who had history of astrocytoma (grade 1) brain diagnosed about 27 years ago when he was only 8 years old. He had about 60 cycles of radiotherapy following tumor removal. The patient recovered well and was independent with activities of daily living for the next 25 years. He had an episode of sudden onset right sided weakness with dysarthria and fits two years ago. His MRI brain ruled out any tumor recurrence. He was started on dual antiplatelets, high dose statins. and was continued on antiepileptics and symptoms resolved.
He had no risk factors for ischemic stroke and he was thoroughly investigated for young stroke.MRI brain also showed large area of gliosis in left frontoparietal region as shown in figure 1.

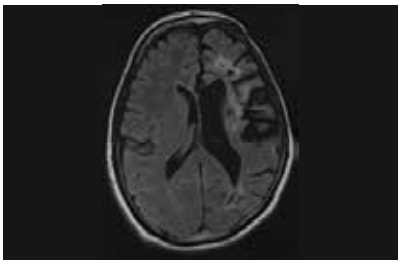

Figure 1. MRI brain showing large area of gliosis in left frontoparietal region (arrow) and left cortical atrophy.

After about a year and a half he had another episode of headache, photo phobia,along with dysphasia and new onset right sided weakness. Headache was throbbing and lasted for a day before development of right sided symptoms.He also had transient episodes of vacant spells which were treated as seizure,antiepileptics were increased and such spells were controlled. CSF routine examination and opening pressure was normal. EEG did not show any epileptiform discharges.MRI brain with contrast again ruled out tumor recurrence and

leptomeningeal disease. MRI brain DWI sequence showed subacute infarcts in left parietal parasagittal and parasylvian region as shown in figure 2.

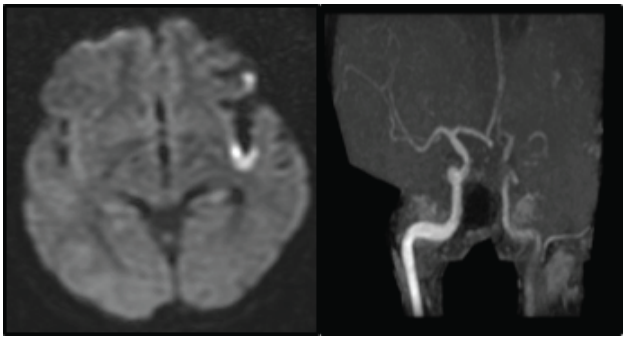

Figure 2. MRI brain DWI sequence (Right) showing area of restricted diffusion left parasylvian region(arrow).MRA brain (left)showed attenuated left ICA(arrow) and severely attenuated and thrombosed left MCA.

MRA brain showed attenuated left ICA and attenuated and thrombosed left MCA as shown in figure 2 left). Patient was thoroughly worked up for thrombophilia, His protein C, S, factor laiden, antithrombin III were negative. His ANA and vasculitis profile was negative.ECG, echocardiogram and 24 hours holter monitoring was also normal. Patient was started on dual antiplatelets and symptoms gradually improved but residual right sided weakness persisted this time.

DISCUSSION

When tumor recurrence has been ruled out in a patient with brain tumor treated with surgery and radiotherapy who presents with neurological symptoms, complications of radiation therapy such as progressive leukoencephalopathy with cognitive decline, focal radiation necrosis mimicking recurrent tumor as well as SMART should be considered.[2]

The clinical and radiological picture of this patient is compatible with the SMART syndrome. SMART syndrome involves transient, reversible neurological dysfunction which may include migrainous headache, at times preceded by aura, prolonged hemispheric neurological impairment and sometimes seizure activity1like our patient had headache, focal right sided weakness and aura like visual symptoms.His symptoms resolved after first episode but after second episode they were considered to be due to radiation induced vasculopathy as confirmed by MRA changes.

Neuroimaging studies of patients with SMART syndrome typically show focal gyral thickening of the affected cortex and gyriform contrast enhancement[2].

The clinical presentation of SMART has similarities with PRES (posterior reversible encephalopathy syndrome), including headaches, neurological deficits and seizures are seen more frequently in PRES than in SMART. The pathophysiology of PRES is combined vasoconstriction and vasodilation and has the same patterns as seen in vasculopathy. This results in blood brain barrier disruption leading to symmetric hemispheric edema and contrast enhancement on MRI.[3]

Since radiation may preferentially damage endothelial cells, the SMART syndrome might be a reversible radiation vasculopathy comparable with PRES. An alternative hypothesis is that post-radiation neuronal dysfunction is the underlying mechanism, such as in migraine or epilepsy, with impairment of the trigeminovascular system or a lowered threshold for cortical spreading depression.[2]

Radiation to the brain and neck can induce a delayed vasculopathy with accelerated atherosclerosis which increases the risk of stroke, but this increased risk also manifests years after the radiotherapy.4 It is therefore difficult to differentiate between SMART and radiation induced vasculopathy. SMART episodes are generally considered reversible while vasculopathy is usually irreversible, but there have been few case series suggesting that SMART may be irreversible5. Our patient second episode of symptoms was also not reversible and was left with residual weakness.

Although SMART syndrome is an extremely rare condition, improvements in cancer survival rates have likely resulted in a rise in its frequency as probably previously patients did not survive long enough to see such late effects of these treatments and this entity was probably unrecognized. The interval in years between radiotherapy and the diagnosis of SMART ranges from 1 to 35 years. There does not appear to be an association between SMART and a particular tumor type.[5]

SMART is an extremely rare delayed complication of brain irradiation but awareness and recognition of this disorder are important to make a rapid diagnosis and avoid aggressive interventions such as brain biopsy or cerebral angiography. Further studies are needed to determine the exact etiology of this disorder, its mechanism of pathogenesis, potential biomarkers, its treatment and also how to avoid such complications especially when post radiation survival is greater.

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Author's contribution:
Farheen Niazi; concept, data collection, data analysis, manuscript writing, manuscript review