Western Australian Coding Rule

0719/22 SMART Syndrome

Q.

What is the correct way to code SMART syndrome – Stroke-like migraine attacks after radiation therapy?

A.

SMART Syndrome is a rare, usually *reversible*, *delayed, complication* of cerebral radiation therapy for CNS malignancy.

There is a male prevalence and patients who receive cranial irradiation at a younger age tend to develop the syndrome sooner. Little is known about the mechanism causing this disorder.

Patients present years after radiotherapy with prolonged *stroke-like* episodes. The associated neurological dysfunction may include; visuo-spatial deficits, confusion, hemi-sensory deficits, hemiparesis, aphasia and seizures. The attacks are usually associated with headaches and preceded by a migraine-like aura.

Radiographic investigations show typical changes. MRI of the brain shows transient, diffuse, unilateral, cortical enhancement of the cerebral gyri, sparing the white matter. The changes are within a previous radiation field.

SMART Syndrome is a diagnosis of exclusion. There must be no evidence of; residual or recurrent neoplasm, cerebral infarction, delayed direct effects of radiation, a known seizure or migraine disorder.

The neurological symptoms can last several weeks but it is typically a *self-limiting syndrome with full recovery*. However, recently in the American Journal of Neuroradiology researchers have retrospectively reviewed several cases that have had permanent neurologic and imaging sequelae. The diagnostic criteria are still evolving. (Armstrong and DiMario n.d.; Armstrong, Gillan and Dimario 2013; Black et al. 2013; Koffel 2011; Weerakkody and Gaillard n.d.)

DECISION

SMART syndrome should be coded to:

G43.8 *Other migraine*
U91 Syndrome, not elsewhere classified
Y84.2 *Radiological procedure and radiotherapy as the cause of abnormal reaction of the patient, or of later complication, without mention of misadventure at the time of the procedure*
Y92.2- *Place of occurrence*, *health service area*
U73.8 *Injury or poisoning while engaged in other specified activity.*

Government of **Western Australia**
Department of **Health**

If there are other residual deficits that meet ACS 0002 *Additional diagnoses*, codes for those should also be assigned after G43.8 *Other migraine*.

This WA Coding Rule 0719/22 *SMART Syndrome* supersedes WA Coding Rule 0318/68 *SMART Syndrome*.

This advice has been modified to correspond with an update in ICD-10-AM/ACHI/ACS Eleventh Edition.

[Effective 01 July 2019, ICD-10-AM/ACHI/ACS 11th Ed.]

Government of Western Australia
Department of Health

Western Australian Coding Rule

0318/68 SMART Syndrome

Q.

What code/s should be assigned for SMART syndrome – Stroke-like migraine attacks after radiation therapy?

A.

SMART Syndrome is a rare, usually *reversible*, *delayed, complication* of cerebral radiation therapy for CNS malignancy.

There is a male prevalence and patients who receive cranial irradiation at a younger age tend to develop the syndrome sooner. Little is known about the mechanism causing this disorder.

Patients present years after radiotherapy with prolonged *stroke-like* episodes. The associated neurological dysfunction may include: visuo-spatial deficits, confusion, hemi-sensory deficits, hemiparesis, aphasia and seizures. The attacks are usually associated with headaches and preceded by a migraine-like aura.

Radiographic investigations show typical changes. MRI of the brain shows transient, diffuse, unilateral, cortical enhancement of the cerebral gyri, sparing the white matter. The changes are within a previous radiation field.

SMART Syndrome is a diagnosis of exclusion. There must be no evidence of; residual or recurrent neoplasm, cerebral infarction, delayed direct effects of radiation, a known seizure or migraine disorder.

The neurological symptoms can last several weeks but it is typically a *self-limiting syndrome with full recovery*. However, recently in the American Journal of Neuroradiology researchers have retrospectively reviewed several cases that have had permanent neurologic and imaging sequelae. The diagnostic criteria are still evolving. (Armstrong and DiMario n.d.; Armstrong, Gillan and Dimario 2013; Black et al. 2013; Koffel 2011; Weerakkody and Gaillard n.d.)

DECISION

SMART syndrome should be coded to:

G43.8	***Other migraine***
Y84.2	***Radiological procedure and radiotherapy as the cause of abnormal reaction, or of later complication, without mention of unintentional events at the time of the procedure***
Y92.2-	***Place of occurrence, health service area***
U73.8	***Other specified activity***

Government of **Western Australia**
Department of **Health**

If there are other residual deficits that meet ACS 0002 *Additional diagnoses*, codes for those should also be assigned after G43.8 *Other migraine*.

This WA Coding Rule 0318/68 *SMART Syndrome* supersedes WA Coding Rule 1013/03 *SMART Syndrome*.

This advice has been modified to correspond with an update in ICD-10-AM/ACHI/ACS Tenth Edition.

[Effective 01 Jul 2017, ICD-10-AM/ACHI/ACS 10th Ed.]

Government of Western Australia
Department of Health

Western Australian Coding Rule

1013/03 SMART Syndrome

Q.

What is the correct way to code SMART syndrome – Stroke-like migraine attacks after radiation therapy?

A.

SMART Syndrome is a rare, usually *reversible*, *delayed, complication* of cerebral radiation therapy for CNS malignancy.

There is a male prevalence and patients who receive cranial irradiation at a younger age tend to develop the syndrome sooner. Little is known about the mechanism causing this disorder.

Patients present years after radiotherapy with prolonged *stroke-like* episodes. The associated neurological dysfunction may include; visuo-spatial deficits, confusion, hemi-sensory deficits, hemiparesis, aphasia and seizures. The attacks are usually associated with headaches and preceded by a migraine-like aura.

Radiographic investigations show typical changes. MRI of the brain shows transient, diffuse, unilateral, cortical enhancement of the cerebral gyri, sparing the white matter. The changes are within a previous radiation field.

SMART Syndrome is a diagnosis of exclusion. There must be no evidence of; residual or recurrent neoplasm, cerebral infarction, delayed direct effects of radiation, a known seizure or migraine disorder.

The neurological symptoms can last several weeks but it is typically a *self-limiting syndrome with full recovery*. However, recently in the American Journal of Neuroradiology researchers have retrospectively reviewed several cases that have had permanent neurologic and imaging sequelae. The diagnostic criteria are still evolving. (Armstrong and DiMario n.d.; Armstrong, Gillan and Dimario 2013; Black et al. 2013; Koffel 2011; Weerakkody and Gaillard n.d.)

DECISION

SMART syndrome should be coded to:

G43.8 *Other migraine*
Y84.2 *Radiological procedure and radiotherapy as the cause of abnormal reaction of the patient, or of later complication, without mention of misadventure at the time of the procedure*
Y92.22 *Health service area*
U73.8 *Injury or poisoning while engaged in other specified activity.*

Government of **Western Australia**
Department of **Health**

If there are other residual deficits that meet ACS 0002 *Additional diagnoses*, codes for those should also be assigned after G43.8 *Other migraine*.

[Effective 18 Oct 2013, ICD-10-AM/ACHI/ACS 8th Ed.]