



Western Australian Coding Rule

0725/01 SMART Syndrome

Q.

How is SMART syndrome coded (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, hemisensory 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; or 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.)

Conventions used in the ICD-10-AM Tabular List/Convention 9.5.3 is applicable i.e. the clinical diagnosis 'SMART syndrome' clearly infers an exclusive causation relationship. Because the complication/adverse effect is due to radiotherapy, ACS 1902 *Adverse effects of drugs and radiotherapy* is applicable.

When the manifestation(s) of SMART syndrome aren't specified, assign G43.8 *Other migraine* via Alphabetic Index:



Migraine (idiopathic)

...

- specified NEC G43.8

DECISION

Apply ACS 1902 *Adverse effects of drugs and radiotherapy* and ACS 0005 *Syndromes*.

For SMART syndrome with multiple manifestations, assign:

Codes for the patient's manifestations (that meet ACS 0001 and ACS 0002)

U91 *Syndrome, not elsewhere classified*

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*

U73.89 *Other specified activity, not elsewhere classified*

For SMART syndrome with a single manifestation, assign:

A code for the patient's manifestation (that meets ACS 0001 or 0002)

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*

U73.89 *Other specified activity, not elsewhere classified*

It is not necessary to assign U91 *Syndrome, not elsewhere classified* in accordance with ACS 0005 *Syndromes/Directive 1*, as this only applies when there are multiple manifestations.

This WA Coding Rule 0725/01 SMART Syndrome supersedes WA Coding Rule 0719/22 SMART Syndrome.

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

Effective 1 July 2025, ICD-10-AM/ACHI/ACS 13th Ed.

As per the Patient Activity Data Policy (MP 0164/21) Western Australian Coding Rules must be followed.



Western Australian Coding Rule

0719/22 SMART Syndrome

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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.*

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*.

Effective 1 July 2019, ICD-10-AM/ACHI/ACS 11th Ed.