

**Mycosis Fungoides****Mycosis Fungoides and Sezary Disease of Skin, Vulva, Penis, Scrotum****C44.0-C44.9, C51.0-C51.2, C51.8-C51.9, C60.0-C60.2, C60.8-C60.9, C63.2****M-9700-9701****C44.0 Skin of lip, NOS****C44.1 Eyelid****C44.2 External ear****C44.3 Skin of other and unspecified parts of face****C44.4 Skin of scalp and neck****C44.5 Skin of trunk****C44.6 Skin of upper limb and shoulder****C44.7 Skin of lower limb and hip****C44.8 Overlapping lesion of skin****C44.9 Skin, NOS****C51.0 Labium majus****C51.1 Labium minus****C51.2 Clitoris****C51.8 Overlapping lesion of vulva****C51.9 Vulva, NOS****C60.0 Prepuce****C60.1 Glans penis****C60.2 Body of penis****C60.8 Overlapping lesion of penis****C60.9 Penis****C63.2 Scrotum, NOS**

**Note 1:** Laterality must be coded for C44.1-C44.3 and C44.5-C44.7. For codes C44.3 and C44.5, if the tumor is midline (e.g., chin), code 5 (midline) in the laterality field.

**Mycosis Fungoides****CS Tumor Size****See Standard Table****Mycosis Fungoides****CS Extension**

**Note 1:** For skin, patch indicates any size skin lesion without significant elevation or induration.

**Note 2:** For skin, plaque indicates any size skin lesion that is elevated or indurated.

**Note 3:** For skin, tumor indicates at least one 1 centimeter (cm) diameter solid or nodular lesion with evidence of depth and/or vertical growth. If a physician describes a skin lesion of less than 1 cm as a tumor, use code 550.

**Note 4:** Use code 250 when skin involvement is present but only a general location/site is mentioned (i.e., face, torso, arms, legs). Use code 300 when there is skin involvement but there is no mention of location/site.

**Note 5:** Ignore erythroderma if stated to involve less than 50% of the skin. Use codes 110-650.

**Note 6:** Physicians use more than one method to estimate percentage of skin involvement by mycosis fungoides (MF). Code percent of skin involvement as stated by the physician. If no percentage is

stated and no word such as localized or generalized is used and no stage is given, code as percent not stated.

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
100	<b>OBSOLETE DATA RETAINED V0200</b> Plaques, papules, or erythematous patches ("plaque stage"): Less than 10 % of skin surface, no tumors Limited plaques/patches MFCG Stage I	ERROR	T1	L	L
110	Patches only with less than 10% of skin surface involved	T1a	T1	L	L
120	Plaques or papules, with or without patches, with less than 10% of skin surface involved, no tumors	T1b	T1	L	L
130	Skin involvement less than 10%, not stated whether plaques, papules or patches, no tumors Limited plaques/patches	T1NOS	T1	L	L
150	Plaques, papules, or erythematous patches: % body surface not stated, no tumors	T1NOS	T1	L	L
200	<b>OBSOLETE DATA RETAINED V0200</b> Plaques, papules, or erythematous patches ("plaque stage"): Greater than or equal to 10% of skin surface, no tumors Generalized plaques/patches MFCG Stage II	ERROR	T2	L	L
210	Patches only with 10% or more of skin surface involved	T2a	T2	L	L
220	Plaques or papules, with or without	T2b	T2	L	L

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
220 cont'd	patches, with 10% or more of skin surface involved, no tumors	T2b	T2	L	L
230	Skin involvement 10% or more, not stated whether plaques, papules or patches, no tumors Generalized plaques/patches Stated as T2, NOS	T2NOS	T2	L	L
250	<b>OBSOLETE DATA CONVERTED V0203</b> ; See code 150 Plaques, papules, or erythematous patches: % body surface not stated, no tumors	ERROR	ERROR	ERROR	ERROR
300	Skin involvement, NOS: Extent not stated, no tumors Localized, NOS	T1NOS	T1	L	L
500	<b>OBSOLETE DATA RETAINED V0200</b> One or more tumors (tumor stage) Cutaneous tumors	ERROR	T3	RE	RE
550	Skin lesion described as tumor less than 1 cm	T2NOS	T3	RE	RE
600	One or more tumors equal to 1 cm or greater Cutaneous tumor, size not stated	T3	T3	RE	RE
650	<b>OBSOLETE DATA CONVERTED V0203</b> ; See code 740 Stated as T3 with no other information on extension	ERROR	ERROR	ERROR	ERROR
700	<b>OBSOLETE DATA RETAINED V0200</b> Generalized erythroderma (greater than 50% of body involved with diffuse	ERROR	T4	RE	RE

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
700 cont'd	redness) Sezary syndrome/Sezary disease MFCG Stage III	ERROR	T4	RE	RE
730	Generalized erythroderma (Greater than 50% less than 80% body surface involved with diffuse redness)	T3	T4	RE	RE
740	Stated as T3 with no other information on extension	T3	T3	RE	RE
750	Generalized erythroderma (Equal to or greater than or equal to 80% body surface involved with diffuse redness)	T4	T4	RE	RE
800	Stated as T4 with no other information on extension	T4	T4	RE	RE
950	No evidence of primary tumor	T0	T0	U	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in patient record	TX	TX	U	U

**Mycosis Fungoides****CS Tumor Size/Ext Eval****See Standard Table****Mycosis Fungoides****CS Lymph Nodes**

**Note 1:** Only peripheral lymph nodes are assessed; these include the cervical, supraclavicular, epitrochlear, axillary, and inguinal nodal groups. Clinically abnormal peripheral lymph node(s) is defined as any palpable peripheral node that on physical examination is firm, irregular, clustered, fixed, or 1.5 centimeter (cm) or larger in diameter. Central nodes, which are not generally amenable to pathologic assessment, are not currently considered in the nodal classification unless used to establish N3 histopathologically.

**Note 2:** Clinically abnormal nodes must be histopathologically confirmed as positive to assign codes 110-405.

**Note 3:** Dutch grade system includes:

Grade 1 - Dermatopathic lymphadenopathy (DL)

Grade 2 - Early involvement by mycosis fungoides (MF), (presence of cerebriform nuclei larger than 7.5 micrometers (um))

Grade 3 - Partial effacement of lymph node architecture; many atypical cerebriform mononuclear cells (CMCs)

Grade 4 - Complete effacement of lymph node architecture

**Note 4:** National Cancer Institute - Lymph Nodes (NCI-LN) grade system includes:

LN 0 - No atypical lymphocytes

LN 1 - Occasional and isolated atypical lymphocytes not arranged in clusters

LN 2 - Many atypical lymphocytes or in 3-6 cell clusters

LN 3 - Aggregates of atypical lymphocytes; nodal architecture preserved

LN 4 - Partial/complete effacement of nodal architecture by atypical lymphocytes or frankly neoplastic cells

**Note 5:** A T-cell clone (clone negative or clone positive) is defined by polymerase chain reaction (PCR) or Southern blot analysis of the T-cell receptor gene.

**Note 6:** For this site, code all lymph node (regional and distant) involvement in this field.

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
000	No regional lymph node involvement No clinically abnormal peripheral lymph nodes	N0	N0	NONE	NONE
100	Clinically enlarged palpable lymph node(s) (adenopathy), <b>WITH</b> pathologically negative nodes	N0	N1	RN	RN
110	Clinically normal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone negative	N1a	N2	RN	RN
115	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone negative Stated as N1a with no other information on regional lymph nodes	N1a	N3	RN	RN
120	Clinically normal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone positive	N1b	N2	RN	RN
125	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone positive	N1b	N3	RN	RN

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
125 cont'd	Stated as N1b with no other information on regional lymph nodes	N1b	N3	RN	RN
130	Clinically normal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone unknown	N1NOS	N2	RN	RN
135	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 1 or NCI LN 0-2: Clone unknown	N1NOS	N3	RN	RN
140	Stated as N1 [NOS] with no other information on regional lymph nodes	N1NOS	N1	RN	RN
200	<b>OBSELETE DATA RETAINED V0200</b> ; No clinically enlarged palpable lymph node(s) (adenopathy); pathologically positive lymph node(s)	ERROR	N2	RN	RN
210	Clinically normal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone negative	N2a	N2	RN	RN
215	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone negative Stated as N2a with no other information on regional lymph nodes	N2a	N3	RN	RN
220	Clinically normal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone positive	N2b	N2	RN	RN
225	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone positive Stated as N2b with no other information on regional lymph nodes	N2b	N3	RN	RN
230	Clinically normal peripheral lymph	N2NOS	N2	RN	RN

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
230 cont'd	node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone unknown	N2NOS	N2	RN	RN
235	Clinically abnormal peripheral lymph node(s) <b>AND</b> histopathologically Dutch grade 2 or NCI LN 3: Clone unknown	N2NOS	N3	RN	RN
240	Stated as N2 [NOS] with no other information on regional lymph nodes	N2NOS	N2	RN	RN
300	<b>OBSOLETE DATA RETAINED V0200</b> ; Both clinically enlarged palpable lymph node(s) (adenopathy) and pathologically positive lymph nodes	ERROR	N3	RN	RN
310	Clinically normal peripheral lymph nodes <b>AND</b> histopathologically Dutch grades 3-4 or NCI LN 4: Clone positive or negative or unknown	N3	N2	RN	RN
315	Clinically abnormal peripheral lymph nodes <b>AND</b> histopathologically Dutch grades 3-4 or NCI LN 4: Clone positive or negative or unknown Stated as N3 with no other information on regional lymph nodes	N3	N3	RN	RN
400	Clinically normal peripheral lymph node(s), histopathologically confirmed <b>BUT</b> Dutch grade and NCI LN grade unknown: Clone positive, negative or unknown	N1NOS	N2	RN	RN
405	Clinically abnormal peripheral lymph node(s), histopathologically confirmed <b>BUT</b> Dutch grade and NCI LN grade unknown: Clone positive, negative or unknown	N1NOS	N3	RN	RN

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
800	Lymph nodes, NOS	N1NOS	N1	RN	RN
850	Clinically abnormal peripheral lymph nodes <b>AND</b> no histopathologic confirmation	NX	N1	U	U
999	Unknown; regional lymph nodes not stated; Regional lymph node(s) cannot be assessed; Not documented in patient record	NX	NX	U	U

**Mycosis Fungoides  
CS Lymph Nodes Eval  
See Standard Table**

**Mycosis Fungoides  
Regional Nodes Positive  
See Standard Table**

**Note:** Record this field even if there has been preoperative treatment.

**Mycosis Fungoides  
Regional Nodes Examined  
See Standard Table**

**Mycosis Fungoides  
CS Mets at DX**

**Note 1:** For this site, code all lymph node (regional and distant) involvement in CS Lymph Nodes field. Only visceral involvement is coded in CS Mets at DX. Bone marrow and peripheral blood involvement is coded in CS Site-Specific Factor 1.

**Note 2:** Visceral involvement is considered metastatic and should be questioned in the absence of node or blood involvement.

**Note 3:** For visceral disease (Stage IVB), documentation of involvement by only one organ outside the skin, nodes, or blood is needed.

**Note 4:** Visceral involvement must be pathologically confirmed, except for the liver and spleen, which may be documented with imaging (code 60). If liver or spleen is involved clinically without imagines, use code 05.

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
00	No visceral organ involvement	M0	M0	NONE	NONE

Code	Description	TNM 7 Map	TNM 6 Map	SS77 Map	SS2000 Map
05	Clinical confirmation only of visceral (non-cutaneous, extra nodal) involvement (Except for liver and spleen imaging, see code 10)	M0	M1	NONE	NONE
10	Imaging confirmation of liver and/or spleen metastasis	M1	M1	D	D
40	<b>OBSOLETE DATA RETAINED V0200</b> Visceral (non-cutaneous, extra nodal) involvement: Carcinomatosis Distant metastasis, NOS MFCG Stage IV	ERROR	M1	D	D
45	Visceral (non-cutaneous, extranodal) involvement, pathologically confirmed: Involvement by at least one organ outside the skin, nodes, blood, or bone marrow	M1	M1	D	D
60	Carcinomatosis, unknown if clinical or pathological confirmation	M0	M1	D	D
70	Distant metastasis, NOS Stated as M1 with no other information on distant metastasis	M1	M1	D	D
99	Unknown; distant metastasis not stated Distant metastasis cannot be assessed Not documented in patient record	M0	MX	U	U

**Mycosis Fungoides****CS Mets Eval****See Standard Table**

**Note:** This item reflects the validity of the classification of the item CS Mets at DX only according to the diagnostic methods employed.

**Mycosis Fungoides****CS Site-Specific Factor 1****Peripheral Blood Involvement****Note:** See page A-115

**Note 1:** Sezary cells are circulating atypical, malignant T lymphocytes with hyperconvoluted cerebriform nuclei, with a mature memory T-cell phenotype (CD3+, CD4+) with loss of CD 7 and CD26.

**Note 2:** Assessment of blood tumor burden is performed using two types of tests:

A. Using the microscope to count neoplastic (Sezary) cells. The results can be given as counts per cubic millimeter (mm<sup>3</sup>) or as a percentage of the total lymphocytes that are abnormal (Sezary cells).

B. Flow cytometry to characterize the cell surface markers on the neoplastic cells, where the neoplastic cells show the loss of some specific markers (CD26, for example.) Interpretation of the test results is complicated and may vary. In addition, clonality may be assessed as explained in Note 4.

**Note 3:** The categories for peripheral blood involvement are:

B0: No significant blood involvement

B1, Low blood tumor burden

B2: High blood tumor burden.

**Note 4:** Clonality may be assessed on tissue and blood samples for this disease. The tests look for what is called "TCR gene rearrangement" a detectable genetic pattern of the T-cell receptors on the neoplastic T-cells. A finding of "monoclonal" means the cells have derived from a single ancestor cell. In this staging system, "monoclonal" is termed "clone +" or "clone positive," and this generally indicates a worse prognosis. "Polyclonal" means the cells in question have not derived from a single ancestor cell, and this finding is termed "clone -" or "clone negative." This may indicate a better prognosis. Test methods for clonality include Southern blot and polymerase chain reaction (PCR).

**Note 5:** Record the B rating as stated by the physician. If counts or percentages of neoplastic cells, flow cytometry test results, and/or clonality test results are performed but a B rating is not stated by the physician, use code 090.

**Note 6:** If there is no peripheral blood involvement or Sezary cell count is less than 1000, code 030.

Code	Description	B Map
000	<b>OBSOLETE DATA CONVERTED V0203;</b> See code 030 No peripheral blood involvement: Less than 1000 Sezary cells	B0
001	<b>OBSOLETE DATA RETAINED V0200</b> Atypical circulating cells in peripheral blood: Less than 5% Greater than or equal to 1000 Sezary cells	BX
002	<b>OBSOLETE DATA RETAINED V0200</b> Atypical circulating cells in peripheral blood: Greater than 5%	BX

Code	Description	B Map
003	<b>OBSOLETE DATA RETAINED V0200</b> % Not stated	BX
010	Absence of significant blood involvement: 5% or less of peripheral blood lymphocytes are atypical (Sezary) cells Clone negative Stated as B0a	B0a
020	Absence of significant blood involvement: 5% or less of peripheral blood lymphocytes are atypical (Sezary) cells Clone positive Stated as B0b	B0b
030	Absence of significant blood involvement: 5% or less of peripheral blood lymphocytes are atypical (Sezary) cells Clone unknown Stated as B0 [NOS]	B0NOS
040	Low blood tumor burden: More than 5% of peripheral blood lymphocytes are atypical (Sezary) cells but does not meet the criteria of B2 Clone negative Stated as B1a	B1a
050	Low blood tumor burden: More than 5% of peripheral blood lymphocytes are atypical (Sezary) cells but does not meet the criteria of B2 Clone positive Stated as B1b	B1b
060	Low blood tumor burden: More than 5% of peripheral blood lymphocytes are atypical (Sezary) cells but does not meet the criteria of B2 Clone unknown Stated as B1 [NOS]	B1NOS
070	High blood tumor burden: 1000 microliter (uL) Sezary cells or more Clone positive Stated as B2	B2

Code	Description	B Map
080	% of atypical peripheral blood lymphocytes not stated and B rating not stated	BX
090	Sezary cell counts, blood flow cytometry, and/or clonality results in chart, B rating not stated	BX
100	<b>OBSOLETE DATA CONVERTED V0203</b> See code 997 Sezary cell counts, blood flow cytometry, and/or clonality tests ordered, test results not in chart, B rating not known	BX
988	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 988 will result in an edit error.)	BX
997	Sezary cell counts, blood flow cytometry, and/or clonality tests ordered, test results not in chart , B rating not stated	BX
999	Unknown or no information Not documented in patient record	BX