

Beleid MERKEL CEL CARCINOMEN

1. Diagnose

APO onderzoek met immunohistochemie om kleincellig longcarcinoom uit te sluiten.

Volledig huidnazicht, palpatie klierstreken, labo (uitgebreid, met NSE tumormerker), PET-CT, sentinel lymph node biopsy

2. TNM stadiëring

Overgenomen uit UpToDate®

Primary tumor (T)			
T category	T criteria		
TX	Primary tumor cannot be assessed (eg, curetted)		
T0	No evidence of primary tumor		
Tis	<i>In situ</i> primary tumor		
T1	Maximum clinical tumor diameter ≤2 cm		
T2	Maximum clinical tumor diameter >2 but ≤5 cm		
T3	Maximum clinical tumor diameter >5 cm		
T4	Primary tumor invades fascia, muscle, cartilage, or bone		
Regional lymph nodes (N)			
Clinical (N)			
N category	N criteria		
NX	Regional lymph nodes cannot be clinically assessed (eg, previously removed for another reason, or because of body habitus)		
N0	No regional lymph node metastasis detected on clinical and/or radiologic examination		
N1	Metastasis in regional lymph node(s)		
N2	In-transit metastasis (discontinuous from primary tumor; located between primary tumor and draining regional nodal basin, or distal to the primary tumor) without lymph node metastasis		
N3	In-transit metastasis (discontinuous from primary tumor; located between primary tumor and draining regional nodal basin, or distal to the primary tumor) with lymph node metastasis		
Distant metastasis (M)			
Clinical (M)			
M category	M criteria		
M0	No distant metastasis detected on clinical and/or radiologic examination		
M1	Distant metastasis detected on clinical and/or radiologic examination		
M1a	Metastasis to distant skin, distant subcutaneous tissue, or distant lymph node(s)		
M1b	Metastasis to lung		
M1c	Metastasis to all other visceral sites		
Prognostic stage groups			
Clinical stage group (cTNM)			
When T is...	And N is...	And M is...	Then the stage group is...
Tis	N0	M0	0
T1	N0	M0	I
T2-3	N0	M0	IIA
T4	N0	M0	IIB
T0-4	N1-3	M0	III
T0-4	Any N	M1	IV

Primary tumor (T)	
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Tis	<i>In situ</i> primary tumor
T1	Maximum clinical tumor diameter ≤2 cm
T2	Maximum clinical tumor diameter >2 but ≤5 cm
T3	Maximum clinical tumor diameter >5 cm
T4	Primary tumor invades fascia, muscle, cartilage, or bone

Regional lymph nodes (N)	
Pathological (pN)	
pN category	pN criteria
pNX	Regional lymph nodes cannot be assessed (eg, previously removed for another reason or not removed for pathological evaluation)
pN0	No regional lymph node metastasis detected on pathological evaluation
pN1	Metastasis in regional lymph node(s)
pN1a(sn)	Clinically occult regional lymph node metastasis identified only by sentinel lymph node biopsy
pN1a	Clinically occult regional lymph node metastasis following lymph node dissection
pN1b	Clinically and/or radiologically detected regional lymph node metastasis, microscopically confirmed
pN2	In-transit metastasis (discontinuous from primary tumor; located between primary tumor and draining regional nodal basin, or distal to the primary tumor) without lymph node metastasis
pN3	In-transit metastasis (discontinuous from primary tumor; located between primary tumor and draining regional nodal basin, or distal to the primary tumor) with lymph node metastasis

Distant metastasis (M)	
Pathological (M)	
M category	M criteria
M0	No distant metastasis detected on clinical and/or radiologic examination
pM1	Distant metastasis microscopically confirmed
pM1a	Metastasis to distant skin, distant subcutaneous tissue, or distant lymph node(s), microscopically confirmed
pM1b	Metastasis to lung, microscopically confirmed
pM1c	Metastasis to all other distant sites, microscopically confirmed

Prognostic stage groups			
Pathological stage group (pTNM)			
When T is...	And N is...	And M is...	Then the stage group is...
Tis	N0	M0	0
T1	N0	M0	I
T2-3	N0	M0	IIA
T4	N0	M0	IIB
T1-4	N1a(sn) or N1a	M0	IIIA
T0	N1b	M0	IIIA
T1-4	N1b-3	M0	IIIB
T0-4	Any N	M1	IV

3. Behandeling

3.1 Locoregionaal

3.1.1 Primaire tumor

Heelkunde

Brede excisie met ruim (2 cm) tumorvrije randen indien mogelijk

Sentinel lymph node biopsy

Primaire radiotherapie (brede marges tot 5 cm)

Alternatief voor heelkunde indien inoperabel letsel

Adjuvante radiotherapie bij patiënten met volgende risicofactoren:

- primaire tumor ≥1 cm

- primaire tumor in hoofdhals regio
- positieve of nipte snedevlakken (marges < 1 cm)
- lymfovasculaire invasie
- Immuungecompromitteerde patiënt
- Satellietletsels
- Positieve lymfeklieren

Dosis schema radiotherapie

- 45-50 Gy in 25 fracties
- 55-60 Gy zo microscopisch positieve sectievlakken
- 60 to 66 Gy zo macroscopisch positieve SV, inoperable primaire letsels of inoperable regionale LN

3.1.2 Lymfeklieren

Klinisch negatieve klieren: steeds sentinel lymphnode biopsy te doen

Klinisch positieve klieren:

- histologisch te bevestigen
- lymfeklierevidement
- adjuvante radiotherapie

3.2 Gemetastaseerde ziekte

1^{ste} lijn: immuuntherapie onder vorm van Avelumab tweewekelijks

2^{de} lijn: chemotherapie (platinum-etoposide) of studiedeelname

4. Follow-up

Eerste 3 jaar om de 4 maanden, daarna om de 6-12 maanden: klinisch onderzoek van huid en lymfeklieren.

Zelfinspectie maandelijks.

Beeldvorming afhankelijk van risico-inschatting.