“Stepping in the Right Direction: Effective Management of Diffuse Large B-Cell Lymphoma - Leg Type"

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History of Present Illness

- Case of 56 y/o Female patient who complaints of right foot weakness, multiple skin lesions and rapidly growing mass on right ankle with 8 months of evolution prior to his initial presentation.

- Patient underwent an extensive biochemical, infectious, and radiologic workup which was unremarkable.

- Excisional biopsy of the skin lesion:

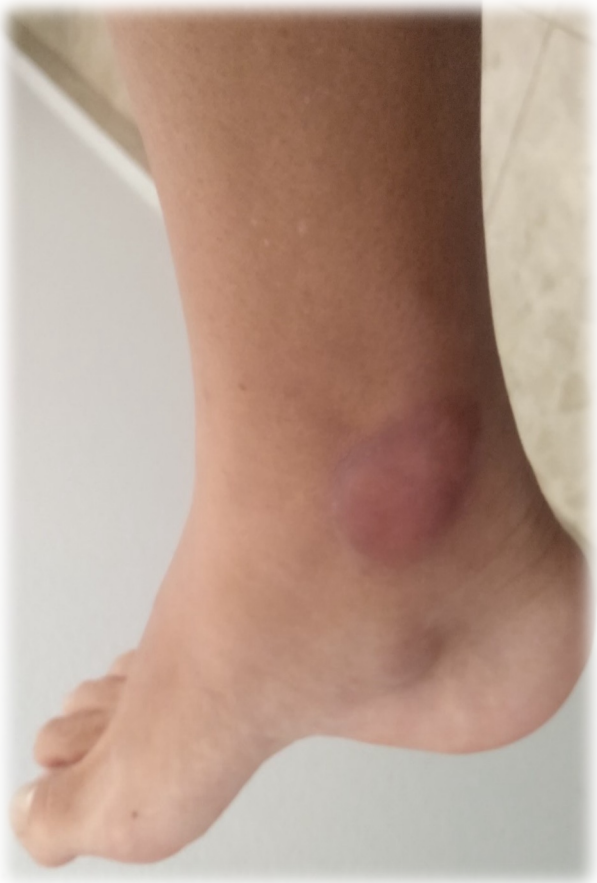
> *Primary Cutaneous Diffuse Large B-Cell Lymphoma –Leg Type*

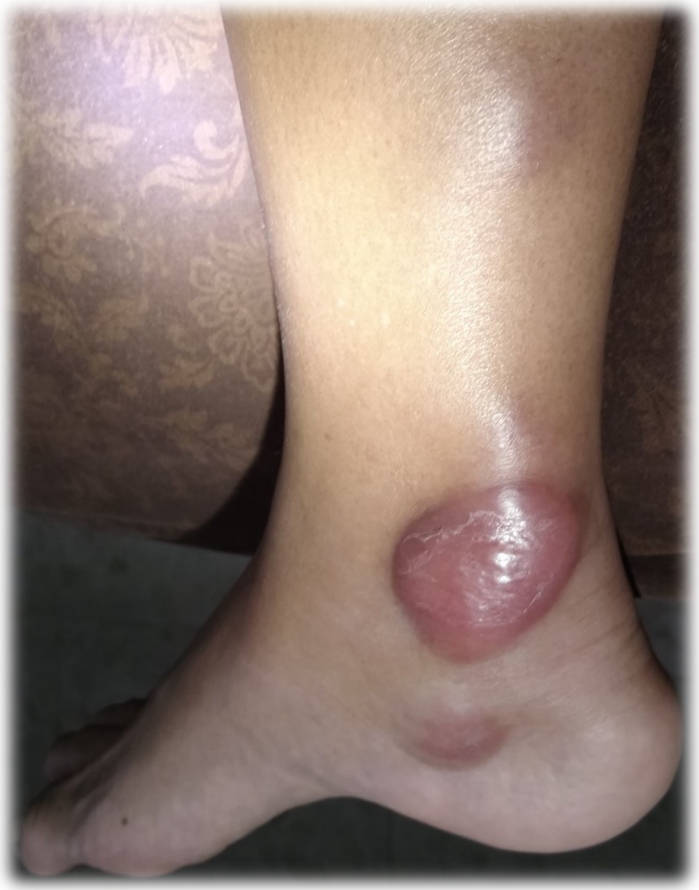
Physical Examination

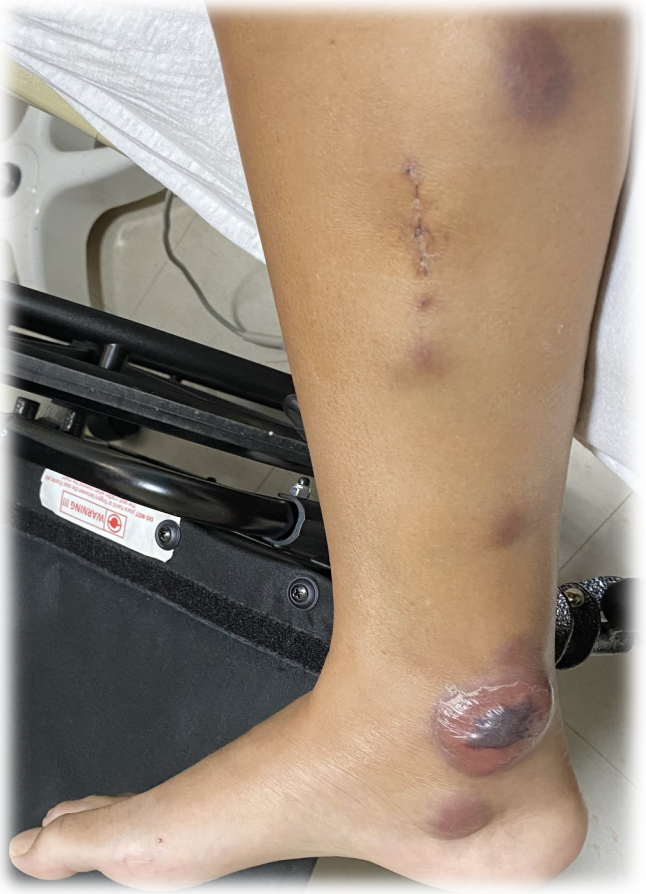
Temp: 98 F **BP**: 107/89 **PR**: 78 **RR:** 14 **O2 Sat:** 98% **Weight:** 83lbs **Height**: 60 in **BSA:** 1.28
ECOG: 0

- **General**: AAO x3. Normal appearance, No acute distress. Cooperative
- **HEENT:** PERRLA, Anicteric. External ears and throat normal, hearing grossly normal, oropharynx unremarkable.
- **Respiratory**: Breathing comfortably, CTA x 2
- **Cardiovascular**: RRR, No murmur or gallops
- **Abdomen**: No masses, Non tenderness, no hepatomegaly, no splenomegaly
- **Lymph Nodes**: No cervical, axillary, supraclavicular, inguinal or popliteal adenopathy
- **Skin**: **Multiple violaceous lesions present along R leg ranging in size between 2 cm to 6 cm. The largest lesion is in the medial part of the right ankle.**
- **Extremities**: Right leg with multiple palpable lesions. Mild edema R>L. Without cyanosis.

Images

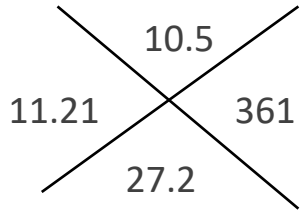






Laboratories:

CBC:

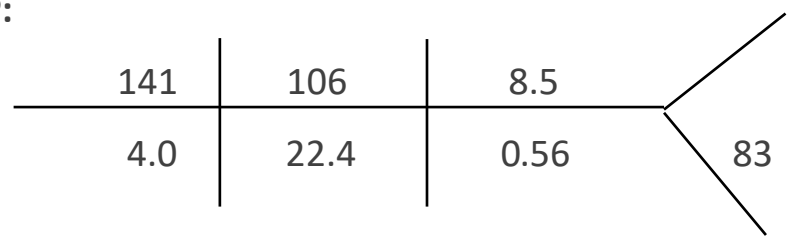


MCV: 90.4

MCHC: 31.3

RDW: 15.2

CMP:



AST: 20

ALT: 11

ALK: 101

LDH: 466

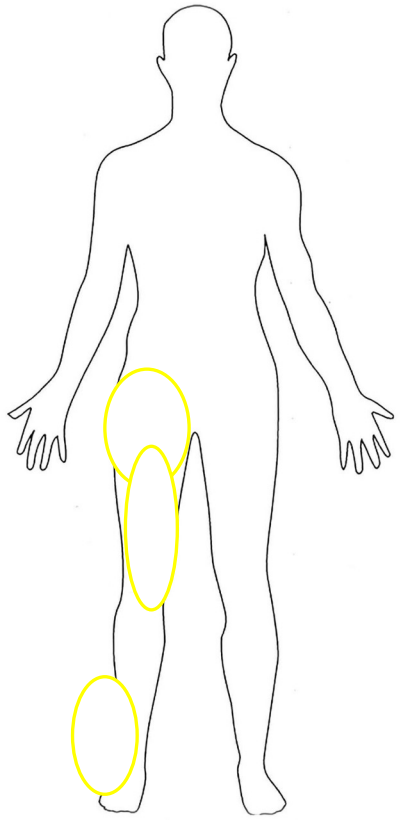
Ca2+: 8.0

Alb: 2.7

PET CT Scan:

- Large, irregular shaped strongly hypermetabolic mass involving the articular space of the right ankle, extending to the soft tissues of the lateral and medial aspects, consistent with malignancy SUV_{max} of 23.54.

- Metastatic disease to multiple lymph nodes along the medial aspect of the right leg, right thigh inguinofemoral and right internal and external iliac chains, with SUV_{max} ranging from 10.76-16.69.

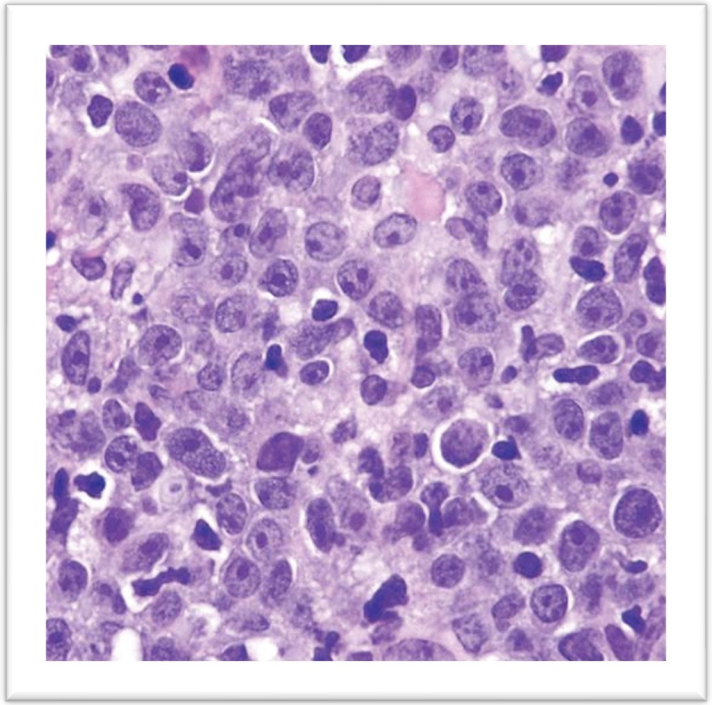


Surgical Pathology Report

Ø**Final Diagnosis:**

-Right Leg Mass:

-Diffuse Large B-Cell Lymphoma



Final Pathology Report / IHC:

CD Marker:	Result:
CD3	Negative
CD5	Positive, diffuse
CD20	Positive, diffuse
CD79	Positive, diffuse
Ki67	Positive in near 100% of neoplastic cells
CD68	Positive in background histiocytes
EBV	Negative
C-Myc	Positive in near 100% of neoplastic cells
BCL-2	Positive

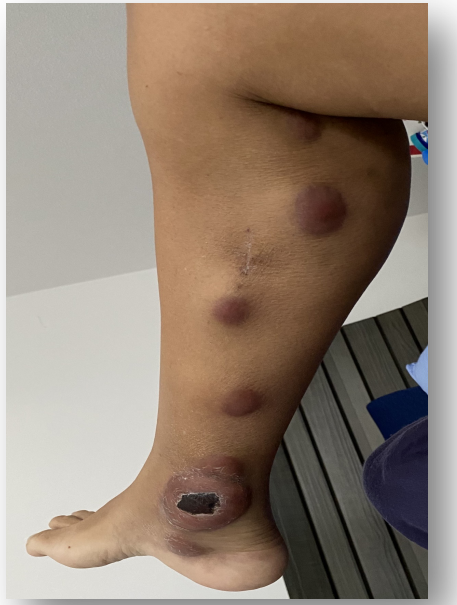
CD Marker:	Result:
MUM-1	Positive in 70% of neoplastic, variable cells
CD10	Positive
PAX5	Positive, diffuse
CD44	Negative
CD23	Negative
CD30	Negative
ALK1	Negative
CD38	Positive

FISH: Neg BCL-2 / Neg BCL-6 / + C-Myc = No DH

Treatment Course

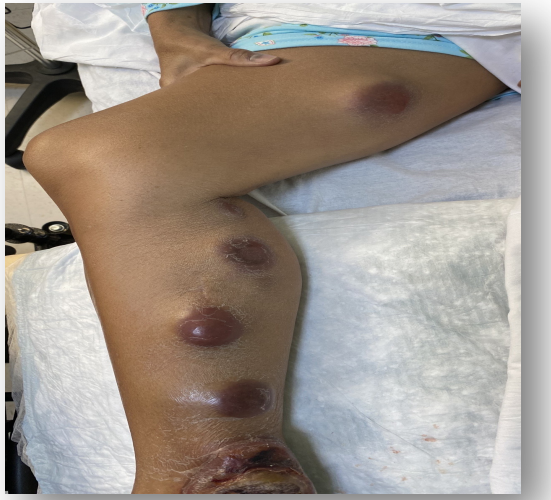
R-CHOP x 2 + RTX

Partial Response

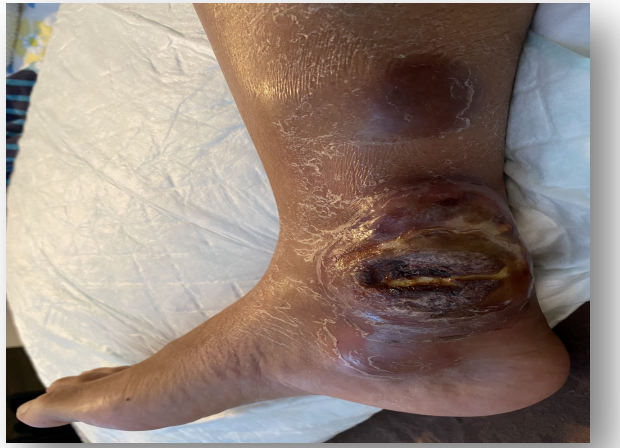


ICE

Progressive Disease



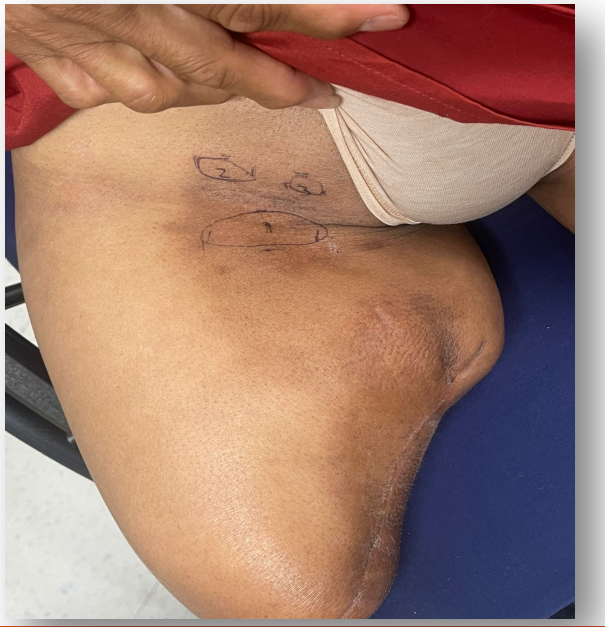
(Surgery) Right AKA



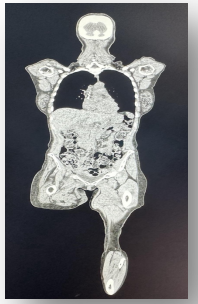
Treatment Course:

Tafasitamab-cxix + Lenalidomide + RTX

Progressive Disease



PET CT Scan



Manifested by two residual FDG Avid right inguinal nodes. The largest one measures 1.7 x 1.3 cm maximal SUV's was estimated to be 5.9 Deauville score sequences 5).

Epcoritamab-bysp Protocol

Primary Cutaneous Diffuse Large B-Cell Lymphoma-Leg Type

- In contrast to nodal lymphomas, PCL cases are primarily of T-cell origin.
- Represents only 4% of all cutaneous lymphomas.
- Has a significantly worse prognosis, with a 5-year overall survival of only 50–60%
- Leg type has the higher rates of relapse and recurrence.
- The median age at initial presentation is approximately 70 with a female predominance.
- PCDLBCL-LT preferentially involves the lower extremities with characteristic rapidly enlarging red to purple nodules.
- Regional lymph nodes and bone marrow represent the typical sites of extracutaneous spread.

Primary Cutaneous Diffuse Large B-Cell Lymphoma-Leg Type/Diagnosis:

- An accurate diagnosis is essential in order to provide prompt treatment, especially considering the immunophenotypic overlap with other cutaneous lymphoma subtypes.

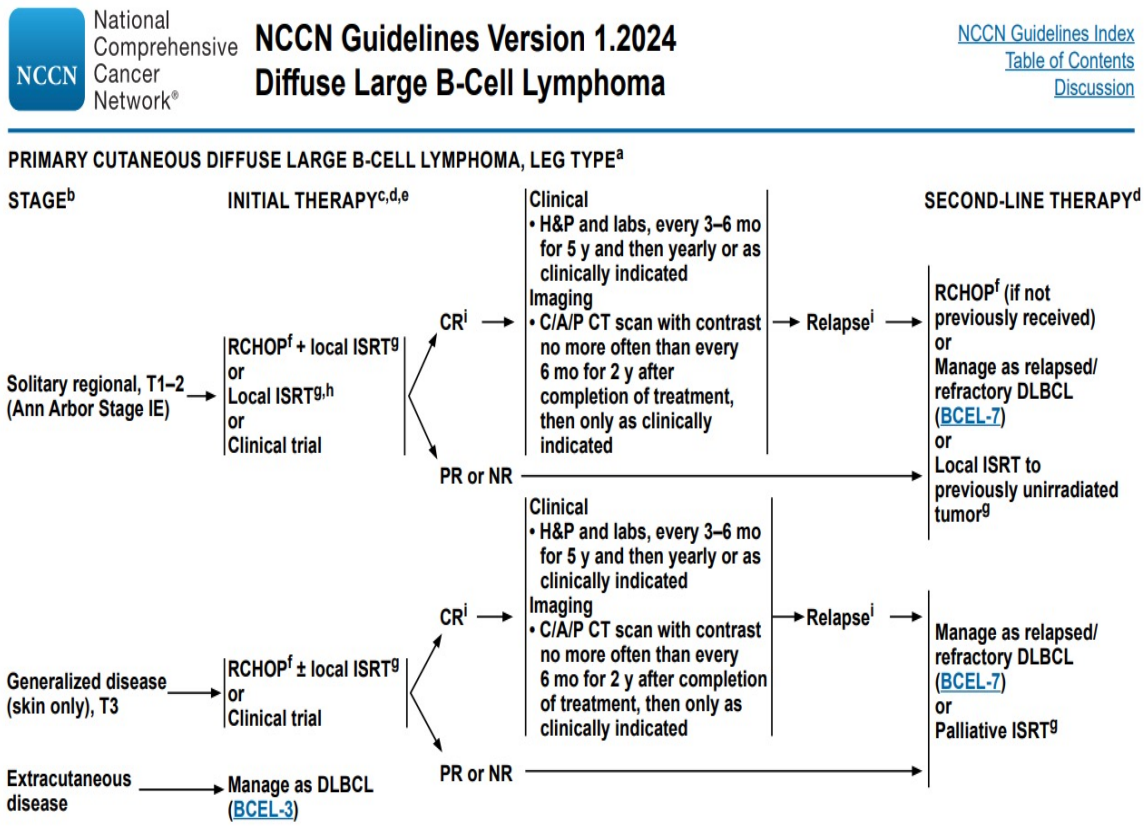
- Tissue specimens predominantly show *MUM-1 and BCL-2 expression*.

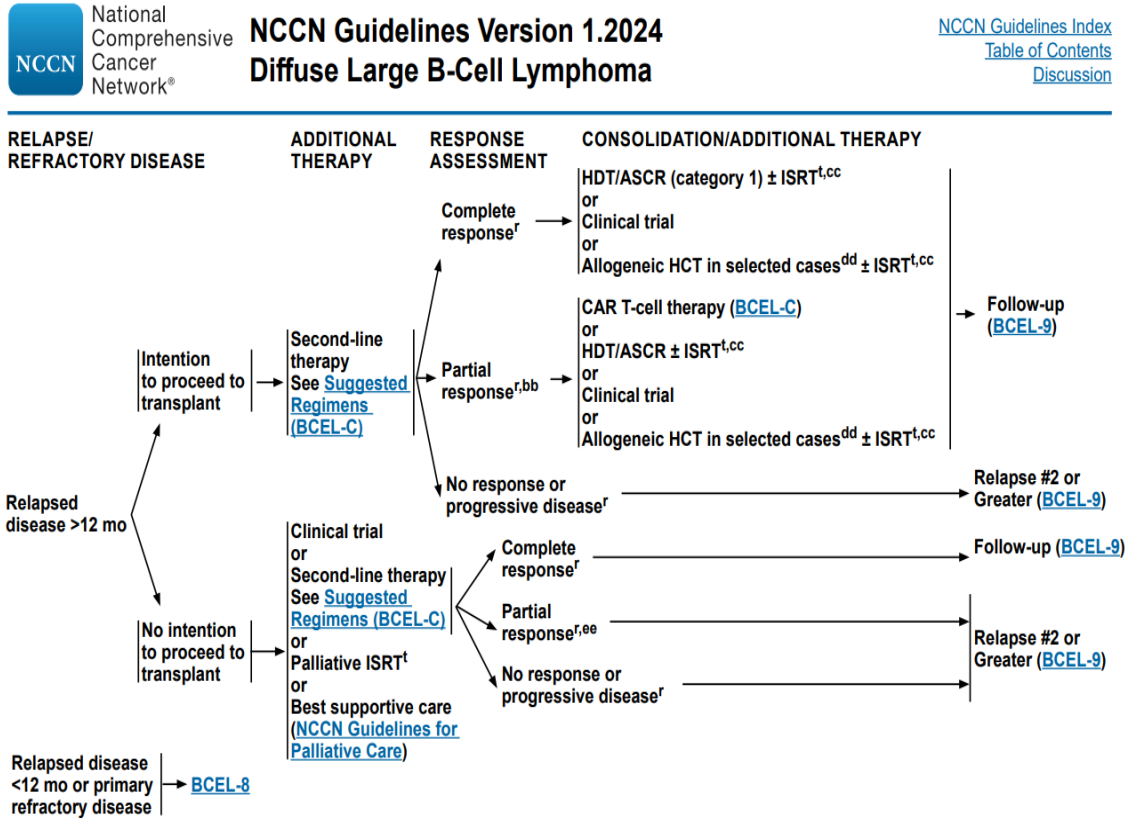
- PCDLBCL-LT frequently expresses *BCL-6, CD20, PAX-5, and CD79a.*

- The germinal center marker *CD10 is usually negative.*

Primary Cutaneous Diffuse Large B-Cell Lymphoma-Leg Type / Treatment:

- First-line treatment for PCLs is anthracycline-based chemotherapy combined with rituximab.
- Therefore, various rituximab plus polychemotherapy combinations are recommended, regardless of the clinical stage.
- Despite the lower incidence of PCDLBCL-LT, clinical trial data has been published that suggests the immunomodulatory agent, lenalidomide, is efficacious in management of relapsing and refractory PCDLBCL-LT.
- **Surgery** is typically NOT the primary treatment for diffuse large B-cell lymphoma (DLBCL), regardless of its subtype or location.





NCCN Guidelines 2024

Conclusion:

- Violaceous nodules on the lower limbs should raise the suspicion of PCDLBCL-LT.
- Lesions can involve areas of the body other than the lower limbs.
- Existing on the spectrum of extranodal cutaneous lymphomas, PCDLBCL-LT is an aggressive cutaneous lymphoma variant and its diagnosis, if delayed, can significantly affect prognosis.
- The primary treatment for DLBCL usually involves chemotherapy, often combined with immunotherapy or targeted therapy.
- Some cases, radiation therapy may also be used to target specific areas of disease involvement.
- Surgery is typically not curative for PCTDLBCL-LT on its own and is usually part of a multidisciplinary approach to treatment.

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Thank you