CASE 35
CLINICAL HISTORY

• Female, 24
• Painful ulcerated lesion
• Left buttock
• Developed over a few weeks
• ?Abscess
• Excision
• Two months later developed a similar lesion on right buttock
The main diagnosis to consider is:

A. Pyoderma gangrenosum
B. Cutaneous vasculitis
C. Intravascular CD30 positive T-cell lymphoma (intralymphatic CD30 positive lymphoproliferative disorder)
D. Lymphomatoid papulosis type A
E. Peripheral T cell lymphoma, NOS
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DIAGNOSIS

?INTRAVASCULAR CD30 POSITIVE ANAPLASTIC LARGE T CELL LYMPHOMA

OR

BENIGN ATYPICAL INTRAVASCULAR CD30(+) T-CELL PROLIFERATION

(INTRALYMPHATIC CD30+ LYMPHOPROLIFERATIVE DISORDER
AM J SURG PATHOL, MAY 2014. EPUB AHEAD OF PRINT)
INTRAVASCULAR LYMPHOMAS

• Most have a B cell lineage and are associated with very poor prognosis
• T cell, NK (EBV positive) intravascular lymphomas are very rare
• CD30 positive intralymphatic lymphoproliferative disorder has only recently been recognized as a distinctive entity
INTRAVASCULAR LARGE B CELL LYMPHOMA

CLINICAL FINDINGS

• F = M
• Adults (elderly)
• Most common in the Asia
• Frequent involvement of the CNS and skin
• Hepatosplenic involvement (26%), bone marrow involvement (32%), lymph node involvement (only 11%)
• Usually sistemic involvement by the time cutaneous lesions develop
• Violaceous ill-defined lesions on trunk and limbs (inverted livedo reticularis pattern)
• B symptoms frequent
• Poor prognosis
• In cases with cutaneous involvement only (around 26%), prognosis appears to be better
INTRA VASCULAR LARGE B CELL LYMPHOMA

HISTOLOGICAL FINDINGS

- Dilated dermal and subcutaneous vascular channels including venules, capillaries and arterioles
- Blood vessels appear dilated and contain numerous large pleomorphic lymphoid cells
- Most cases have a pan-B cell phenotype (CD20 and CD79a positive)
Intravascular ALK-negative anaplastic large cell lymphoma with localized cutaneous involvement and an indolent clinical course: toward recognition of a distinct clinicopathologic entity.


Department of Pathology, Stanford University School of Medicine, Stanford, CA 94305, USA.

- **A case of localized cutaneous intravascular anaplastic lymphoma kinase-negative ALCL (cIALCL) with a very indolent clinical course.** The patient experienced a single cutaneous relapse and remains alive without disease 4 years after diagnosis. Our index case of cIALCL and 1 other tested case were immunohistochemically confirmed to be intralymphatic (contained within D2-40+vessels) as compared with the blood vessel localization of cIT/NKL. Recognition of cIALCLs as a distinct clinicopathologic entity, and in particular their distinction from aggressive, usually EBV cIT/NKLs, may be possible on the basis of a combination of clinicopathologic criteria, allowing for localized therapy in a subset of patients.

Am J Surg Pathol. 2013 Apr
PROPOSAL

• Large T-cell lymphoma with typical ALCL morphology and immunophenotype
• as well as an intralymphatic localization
• represents a distinct clinicopathologic entity that
• should be distinguished from intravascular usually EBV+ aggressive T-cell or NK-cell lymphomas involving blood vasculature
A rare case of intravascular large T-cell lymphoma with an unusual T helper phenotype.

Deetz CO, Gilbertson KG 2nd, Anadkat MJ, Dehner LP, Lu D.

Lauren V. Ackerman Laboratory of Surgical Pathology, Washington University Medical Center, St Louis, MO 63110, USA.

Abstract

- We present a case of a rare intravascular large T-cell lymphoma in a 59-year-old man with an unusual CD3+, CD4+, CD5-, CD30+, CD56-, TIA-1-negative and EBER-negative phenotype. This T helper or CD30 phenotype is particularly uncommon. To our knowledge, it has only been described once before and never in the absence of the cytotoxic marker TIA-1. This case exemplifies the particular diagnostic challenges raised by intravascular large cell lymphomas generally and should encourage the use of endothelial immunohistochemical staining in questionable cases. While evaluating skin punch biopsies, it is critical to keep this rare entity on the differential diagnosis along with the relatively more common intravascular large B-cell lymphoma and epithelial malignancies. Additionally, our understanding of intravascular large natural killer/T-cell lymphoma as a heterogeneous phenotypic entity continues to evolve. This case demonstrates that the degree of this phenotypic heterogeneity may be even greater than previously thought.
Cutaneous intravascular anaplastic large cell lymphoma.

Wang L, Li C, Gao T.

Department of Dermatology, Xijing Hospital, Fourth Military Medical University, Xian, China.

• The patient was a 47-year-old woman who had developed multiple erythematous patches and plaques on her back.

• The lesions responded well to CHOP (cyclophosphamide, hydroxydoxorubicin, oncovin, prednisone) chemotherapy, but relapsed shortly after therapy. The patient was surviving with the disease for eight years but was ultimately lost to follow up.

• Histopathologically, the neoplasm evolved from IL to extravascular lymphoma. This was showed in biopsies obtained at different stages of the disease. The lymphoma cells stained positively for CD30, CD45, CD3, CD4, CD5 and Ki67, and lacked expression of anaplastic lymphoma kinase (ALK), CD8, CD45RA, CD45RO, CD20, CD79, CD56, perforin and granzyme B.

• Our results suggest that IALCL represents a distinct subtype of IL and is histopathologically and biologically different from IL with B, NK or T cell phenotype.
Benign atypical intravascular CD30(+) T-cell proliferation: a reactive condition mimicking intravascular lymphoma.

Riveiro-Falkenbach E, Fernández-Figueras MT, Rodríguez-Peralto JL.

Department of Pathology, Hospital Universitario 12 de Octubre, Instituto de Investigación i+12, Universidad Complutense, Madrid, Spain.

- Report of 2 patients with skin lesions showing an atypical intravascular CD30 T-cell proliferation. Both the patients did not present systemic disease and therefore exhibit a favorable outcome.
Intralymphatic Cutaneous Anaplastic Large Cell Lymphoma/Lymphomatoid Papulosis: Expanding the Spectrum of CD30-positive Lymphoproliferative Disorders.


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Abstract

Intravascular large B-cell lymphomas and EBV NK/T-cell lymphomas commonly follow an aggressive clinical course. We recently reported an entirely intravascular anaplastic large cell lymphoma (ALCL) in the skin with a surprisingly indolent clinical course; interestingly, this lymphoma involved the lymphatic rather than the blood vasculature. We hypothesized that intravascular skin-limited ALCL is distinct from aggressive systemic intravascular lymphomas in its intralymphatic localization and clinical course. We now describe 18 cases of cutaneous intravascular large cell lymphoproliferations from 4 institutions. All 12 intravascular large T-cell lesions were intralymphatic; the majority (9) were CD30 T-cell lymphoproliferative disorders (TLPDs), 5 further classified as intravascular ALK ALCL. One ALK ALCL and 2 benign microscopic intravascular T-cell proliferations were also intralymphatic. A single case of otherwise typical cutaneous follicle center lymphoma contained intralymphatic centroblasts. The clinical and pathologic characteristics of the CD30 TLPDs were similar to those of their extravascular counterparts, including extralymphatic dermal involvement in a subset, DUSP22-IRF4 translocations in half of tested ALK ALCLs, and associated mycosis fungoides in 1; most were skin-limited at baseline and remained so at relapse. All 5 cases of intravascular large B-cell lymphoma involved the blood vasculature and behaved in a clinically aggressive manner; the ALK ALCL, although intralymphatic, was systemic and clinically aggressive. We propose that cutaneous ALK ALCL and related CD30 ALK TLPDs involving the lymphatics are part of an expanding spectrum of CD30 TLPDs. The identification of intralymphatic as distinct from blood vascular localization may provide critical prognostic and therapeutic information.
CD3, CD30(+) intravascular lesions with an indolent clinical course

- Few cases all with good behavior
- However, cases that are ALK positive are associated with involvement elsewhere including lymph nodes and are associated with poor prognosis
CONCLUSION

• Newly described entity
• Indolent and part of the spectrum of CD30 positive lymphoproliferative disorders
• Exclude other lymphomas (systemic ALK1 negative anaplastic lymphoma, B/T cell/NK intravascular lymphoma)
References

1. Riveiro-Falkenbach E, Fernández-Figueras MT, Rodríguez-Peralto JL. Benign atypical intravascular CD30(+) T-cell proliferation: a reactive condition mimicking intravascular lymphoma. Am J Dermatopathol. 2013 Apr;35(2)


