CD99 / FLI-1 Double-positive Anaplastic Large Cell Lymphoma (ALCL): A Potential Diagnostic Pitfall

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Introduction

- ALK + ALCL is defined as a T-cell lymphoma consisting of lymphoid cells that are usually large and have abundant cytoplasm and pleomorphic, often horseshoe-shaped nuclei, with a chromosomal translocation involving the ALK gene and expression of ALK protein and CD30.
- comprises 10-20% of childhood lymphomas.
- About 70% of patients present with advanced disease with peripheral and/or abdominal lymphadenopathy, often associated with extranodal infiltrates and involvement of the bone marrow.
- Most patients have B symptoms, especially high fever.
- It has a wide range of variable morphologies including the so-called common pattern, the histiocytic pattern, the small cell pattern and the composite pattern.
Histopathologic variants of ALCL

Prognostic Impact of Morphologic and Phenotypic Features of Childhood ALK-Positive Anaplastic Large-Cell Lymphoma: Results of the ALCL99 Study
Laurence Lamant, Keith McCarthy et al
Case presentation

- 11 year old patient.
- High grade fever.
- Retroperitoneal mass.
- Laboratory tests including CBC, KFT, LFT and tumor markers were unremarkable except for LDH which was moderately increased to 361 u/l.
- MRI showed a well-defined left-sided retroperitoneal soft tissue mass measuring 6.5 X 4 cm displacing the left kidney.
- A needle-core biopsy was taken.
Microscopy H&E
Differential Diagnosis

- Ewing sarcoma
- Rhabdomyosarcoma
- Neuroblastoma
- Wilm's tumor
- Germ cell tumor
- Dedifferentiated liposarcoma
- Lymphoma
IHC

- CD99
- Desmin,
- Myogenin
- Synaptophysin
- S100
- MDM2
- WT-1
- SALL-4
- LCA
- CD43
CD99
Ewing Sarcoma ??
CD43
Lymphoblastic lymphoma??
CD3
CD99 positive lymphomas

Immunoreactivity of CD99 in Non-Hodgkin's Lymphoma: Unexpected Frequent Expression in ALK-positive Anaplastic Large Cell Lymphoma

Chang Ohk Sung, Young H. Ko, Sanghui Park, Kihyun Kim*, Wonseog Kim*
Departments of Pathology and Hematooncology*, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea
**CD99 positive lymphomas**

- T-lymphoblastic lymphoma 16/16 (100%)
- B-lymphoblastic lymphoma 3/5 (60%)
- NK/T-cell lymphoma 0/28 (0%)
- Angioimmunoblastic T-cell lymphoma 0/8 (0%)
- Peripheral T-cell lymphoma 0/23 (0%)
- Systemic anaplastic large cell lymphoma 8/15 (54%)
- ALK positive 7/10 (70%)
- ALK negative 1/5 (20%)
- Diffuse large B-cell lymphoma 2/37 (5%)
- Burkitt’s lymphoma 2/18 (11%)
- Follicular lymphoma 0/12 (0%)
- Small lymphocytic lymphoma 0/11 (0%)
- Mantle cell lymphoma 0/9 (0%)
Frequent CD99 and FLI-1 Expressions in Diffuse Large B-Cell Lymphoma and Their Association with Proliferative and Apoptotic Rates

Ufuk Berber,1 İsmail Yılmaz,1 Tolga Tuncel,2 Zafer Küçükdaci1 Aprullah Haholu1
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2 GATA Haydarpasa Training Hospital, Department of Medical Oncology, Istanbul

Immunohistochemical Evaluation of FLI-1 in Acute Lymphoblastic Lymphoma (ALL)
A Potential Diagnostic Pitfall

Oscar Lin, MD, PhD, Daniel A. Filippa, MD, and Julie Teruya-Feldstein, MD
FLI-1 positive lymphomas

- Angioimmunoblastic T-cell lymphoma 9/9 (100%)
- Anaplastic large cell lymphoma 4/5 (80%)
- Precursor T lymphoblastic lymphoma/leukemia (T-ALL) 16/16 (100%)
- Enteropathy T-cell lymphoma 1/1 (100%)
- Peripheral T-cell lymphoma, NOS 21/42 (50%)
- T/natural killer cell lymphoma 0/2 (0%)
- Diffuse Large B-cell lymphoma
CD99/FLI-1 positive tumors

- Ewing sarcoma
- Lymphoblastic lymphoma
- Diffuse large B-cell lymphoma
CD30
ALK
Diagnosis

Anaplastic Large T-cell Lymphoma
## Literature review

<table>
<thead>
<tr>
<th>Study Author Year</th>
<th>Total numbers of ALCL cases reviewed</th>
<th>CD99 + ALCL</th>
<th>FLI-1 + ALCL</th>
<th>CD99 - FLI-1 Double + ALCL</th>
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<td>15</td>
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<td>Oscar Lin et al 2009</td>
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<td>M S Shiran et al 2008</td>
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<td>Daniel Buxton et al 2009</td>
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<td>Steven Gustafson et al 2009</td>
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<td>Zhong Lin et al 2012</td>
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<td>Susan Swee-Shan Hue et al 2018</td>
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Take Home Message

The differential diagnosis of CD99/FLI-1 double positive small round blue cell tumors should include ALCL in addition to Ewing sarcoma and ALL.
References


Thank you!