History

- 48 year old male, right upper extremity mass
- Mass in same region excised in infancy (?lymphangiomata)
Initial Resection
Initial Resection

Equivocal CD35+
Focal CK+ (10%)
Diagnosis

- Outside consultation (1998)
  - Two experts: follicular dendritic cell tumor
  - Third expert: probable dendritic cell tumor, type unspecified

- Final Diagnosis:
  Follicular dendritic cell sarcoma
Follow-up

- Post-op radiation

- Two local recurrences, at 3 years and 8 years

- 1.5 cm recurrent mass was excised
Recurrent tumor
Recurrent tumor
CD31
Immunohistochemistry

- Tumor cells were positive for:
  - CD31 (diffusely), CD34 (25-50% of cells), Factor VIII (25% of cells), CK (10-15%)

- Tumor cells were negative for:
  - CD1a, CD21, CD23, CD35, CD68, S-100, HMB45, EMA, desmin, muscle-specific actin (MSA), or smooth muscle actin (SMA)
Final Diagnosis

- Epithelioid hemangioendothelioma (EH), with prominent lymphoid infiltrate (recurrent)
Follicular Dendritic Cell Tumor

- Rare tumor of antigen presenting cells
- Intermediate grade malignancy
- Predominantly in lymph nodes
- 40% recurrence, 25% metastasis, 17% mortality
  - Lung, lymph nodes, liver
Follicular Dendritic Cell Tumor
Epithelioid Hemangioendothelioma

- First described by Enzinger and Weiss in 1982
  - Tumor of endothelial cells, of low-grade malignancy
- Metastasis in up to 30%, but usually to local lymph nodes
- Mortality low
<table>
<thead>
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WHO Classification of Histiocytic/Dendritic cell sarcoma

- WHO Classification (2001) requires absence of clonal B/T cell receptor gene rearrangement
- Rare cases of H/DC sarcoma with Ig receptor gene rearrangement date back to 1985
Clonally related follicular lymphomas and histiocytic/dendritic cell sarcomas: evidence for transdifferentiation of the follicular lymphoma clone

Andrew L. Feldman, Daniel A. Arber, Stefania Pittaluga, Antonio Martinez, Jerome S. Burke, Mark Raffeld, Mireia Camos, Roger Warnke and Elaine S. Jaffe
High Frequency of Clonal Immunoglobulin Receptor Gene Rearrangements in Sporadic Histiocytic/Dendritic Cell Sarcomas

Wei Chen, MD,*† Sean K. Lau, MD,* Dean Fong, MD,‡ Jun Wang, MD,§ Endi Wang, MD, PhD,ǁ Daniel A. Arber, MD,‡ Lawrence M. Weiss, MD,* and Qin Huang, MD, PhD*

Abstract: The diagnosis of histiocytic/dendritic cell (H/DC) sarcomas is currently based on morphology and the presence of immunophenotypic features of H/DC differentiation. The issue whether clonal immunoglobulin receptor gene rearrangements are present in H/DC sarcomas has been debated over decades.

Key Words: histiocytic and dendritic cell sarcoma, immunoglobulin receptor gene rearrangements, differentiation

Results

}\textit{IG Receptor Gene Rearrangements in H/DC Sarcomas}\)

\begin{table}
\centering
\begin{tabular}{|l|l|c|c|c|c|c|c|c|}
\hline
\textbf{Case No.} & \textbf{Final Diagnosis} & \textbf{Clonal IgH Rearrangement} & \textbf{Clonal IgK Rearrangement} & \textbf{\(r(14;18)\) Translocation} & \textbf{In Situ Hybridization-Epstein-Barr virus} & \textbf{CD20} & \textbf{PAX5} & \textbf{BOB.1} & \textbf{Oct2} \\
\hline
23 & HS & + & + & + & NA & – & – & – & + \\
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\end{tabular}
\end{table}

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FDCS indicates follicular dendritic cell sarcoma; HS, histiocytic sarcoma; IDCS, interdigitating dendritic cell sarcoma; NA, unknown due to lacking of available materials.
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Conclusions

- Epithelioid hemangioendothelioma may occasionally be lymphocyte-rich, mimicking follicular dendritic cell tumors
- When encountering a spindled cell lesion with a prominent lymphocyte infiltrate, one must include vascular tumors and FDCT in the differential diagnosis
- Though the WHO has currently classified FDCT based on absence of Ig gene rearrangement, current research suggests it may be present in some histiocytic/dendritic cell tumors
References


Acknowledgment

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- Dr. Jeffrey Cao
- Dr. Craig Zuppan