Follicular dendritic cell sarcoma of a cervical lymph node: Case report and review of the literature

Kanagasabai Sahadevan, MRCSEd; Anirvan Banerjee, FRCS (Otol); Richard Wight, FRCS (Otol)

Abstract
Follicular dendritic cell sarcoma is a rare tumor that occurs mainly in lymph nodes. We report a case of follicular dendritic cell sarcoma in a cervical lymph node that was initially diagnosed as a B-cell lymphoma by Tru-Cut biopsy. The correct diagnosis was established by excision biopsy and immunohistochemistry. This tumor is of interest to head and neck surgeons because its recurrence rate is significant and its metastatic potential has been underestimated because of its rarity and the difficulty in making the diagnosis. We discuss the salient clinical and pathologic features of this tumor, as well as its management protocol, and we review the literature.

Introduction
Dendritic cells are potent antigen-presenting cells that have the ability to initiate primary immune responses. Their main role is to capture and present antigen and immune complexes. These cells are present in lymph nodes, in nonlymphoid organs (e.g., the liver), in the gastrointestinal tract, and on epithelial surfaces. Follicular dendritic cells are classified as cells of the accessory lymphoid system; they are also known as dendritic reticulum cells. Proliferation of follicular dendritic cells occurs in a number of reactive and neoplastic conditions, including reactive follicular hyperplasia, follicular lymphoma, mantle cell lymphoma, nodular lymphocyte-predominant Hodgkin's lymphoma, and angioimmunoblastic T-cell lymphoma. Follicular dendritic cell sarcoma has been previously reported in the lymph nodes of the cervical and axillary regions and extranodally in the mediastinum, tonsil, soft palate, parapharyngeal region, thyroid, gastrointestinal tract, and liver.

Case report
A 74-year-old man presented with a 3-month history of a swelling over the left side of his neck. He was otherwise asymptomatic. Examination revealed the presence of a firm, mobile, 2 × 2-cm swelling in the anterior triangle on the left side. Findings on the remainder of the physical examination were normal. Analysis of a Tru-Cut biopsy revealed the presence of an infiltrate of medium- to large-sized lymphoid cells with irregular nuclei and nucleoli. Immunohistochemical staining was positive for CD79, and a moderate number of reactive T cells stained with CD3. These findings were considered to be suspicious for lymphoma, and a subsequent excision biopsy of the node was undertaken. The patient also underwent random biopsies of the draining region, a left tonsillectomy, and panendoscopy. Findings on computed tomography (CT) of the neck, which was undertaken after the histopathologic examination of the excised lymph node, were normal.

On gross inspection, the 4.0 × 3.5 × 2.5-cm specimen had a rubbery consistency and well-defined margins. Microscopically, nodal tumors of the head and neck tend to have a “pushing” rather than a permeating margin and are traversed by a sclerotic band or delicate fibrovascular septa. The tumor cells can be spindly, ovoid, or polygonal, and they can exhibit a storiform, fascicular, whorled, diffuse, fol-
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licle-like, or trabecular pattern. Most tumors have more than one growth pattern. The cellularwhorls are usually circular and centered on a blood vessel. In most cases, the tumor completely replaces the nodal architecture.10

The tumor cells exhibit a moderate amount of lightly eosinophilic cytoplasm and indistinct cell borders, giving rise to a syncytial appearance. The nuclei are oval to round, and their contours are usually smooth. Nucleoplasm can be clear or granular. Mitotic counts vary from case to case, ranging from 1 to 20 per high-power field (HPF). Coagulative necrosis is also seen in some cases. Another notable feature is the presence of a rich network of small blood vessels. A constant and highly characteristic feature is that the entire tumor is sprinkled throughout with small lymphocytes. Perivascular cuffing with lymphocytes is also noted in some cases.11 In 1998, Fonseca et al published immunohistochemical reactivities in follicular dendritic cell sarcoma (table).12

Follicular dendritic cell sarcoma most commonly presents in a young adult with lymphadenopathy (64% of all cases).11 Various parameters affect outcomes. Because most reported series have been retrospective in nature, there is a lack of uniformity of treatment reported in the literature. Greater morbidity and mortality appear to be associated with tumors larger than 6 cm, intraabdominal tumors, a lack of postoperative adjuvant therapy, and the presence of coagulative necrosis, a high mitotic count (>5/HPF), and moderate nuclear pleomorphism.10,11 Perez-Ordonez and Rosai reported that the association between a poor outcome and an intraabdominal location and marked nuclear pleomorphism is statistically significant.10 According to reports, the overall recurrence rate is 43%, the metastasis rate is 24%, the mortality rate is 17%, and the median time to recurrence is 12 months.3,5,11

From available studies, it is evident that follicular dendritic cell sarcoma is aggressive and should be regarded as a potentially recurrent and metastatic disease entity. Surgery appears to be the clear choice for operable tumors, and adjuvant chemotherapy or radiotherapy should be considered for tumors with marked nuclear pleomorphism and for incompletely resected tumors. Although follicular dendritic cell sarcomas are uncommon, they should be considered in the differential diagnosis of tumors in the head and neck region.

References

Table. Observed immunohistochemical reactivities12 in follicular dendritic cell sarcoma

<table>
<thead>
<tr>
<th>Antigen</th>
<th>No. tests</th>
<th>No. (%) positive</th>
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<tr>
<td>CD20</td>
<td>25</td>
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<tr>
<td>CD21</td>
<td>44</td>
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<td>CD35</td>
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<td>Ki-FDC1p</td>
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* Epithelial membrane antigen.