Follicular dendritic cell sarcoma of the tonsil: A case report and literature review

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Abstract
We describe a case of follicular dendritic cell sarcoma (FDCS) of the tonsil in a 59-year-old woman. She was successfully treated with excision of the mass and postoperative radiation therapy. According to our review of the literature, only 25 cases of extranodal FDCS in the head and neck have been previously reported, including only 10 cases that involved a tonsil. We briefly review these earlier reports, and we discuss the diagnosis and management of FDCS.

Introduction
Follicular dendritic cells are nonlymphoid, nonphagocytic accessory cells of the lymphoid system. Their function is to capture and present antigens and immune complexes to B cells. Follicular dendritic cell sarcoma (FDCS) is a rare entity, as evidenced by the small number of cases reported in the literature. Only 25 cases of extranodal FDCS in the head and neck have been published, and only 10 of these have involved the tonsil. In this article, we describe the eleventh such case, and we discuss the diagnostic and treatment considerations.

Case report
A 59-year-old woman presented to the otolaryngology clinic with a 2-year history of a right tonsil mass that had been found by a physician at another institution. The patient had a history of obstructive sleep apnea with snoring. She denied dysphagia, odynophagia, weight loss, and otalgia. Examination confirmed the presence of a 4-cm exophytic mass involving the right tonsil. No other masses or lymphadenopathies were noted. Computed tomography (CT) of the neck with contrast demonstrated a right oropharyngeal mass with no extension into the parapharyngeal space (figure 1).

The patient was brought to the operating room for endoscopy and biopsy. Histopathologic analysis of the specimen identified FDCS. Hematoxylin and eosin (H&E) stains identified spindle- and oval-shaped cells forming whorls with frequent multinucleated giant cells (figure 2). Immunohistochemical stains were positive for CD21, CD23, and CD35 and negative for CD1p, S-100, CD20, CD3, CD31, CD34, and cytokeratin.

The mass was excised (figure 3) via wide local excision of the right tonsil with primary closure. The patient had no evidence of regional or distant metastasis. She underwent postoperative radiation therapy, receiving 60 and 50 Gy to the oropharynx and neck, respectively. Eighteen months after the cessation of treatment, she exhibited no evidence of recurrence.

Discussion
Our analysis of the 10 previously reported cases of FDCS of the tonsil revealed a 1:1 distribution of men and women and an age range of 18 to 77 years (mean: 45). Except for 1 patient who received preoperative radiation, all patients were treated primarily with tonsillectomy. Radical neck dissection was included in 4 of these cases. Only 1 instance of cervical metastasis was found during the initial workup. Postoperative radiation was performed in 3 cases, with chemotherapy added in 2 of them. Follow-up, which ranged from 3 months to 8 years, yielded 2 cases of recurrence. One case involved a local recurrence with cervical metastasis at 4.5 years, and the other involved metastasis to the lung.

The neoplastic process associated with follicular dendritic cells was first described by Monda et al in 1986. The diagnosis of FDCS is based on morphology, immunohistochemistry, and electron microscopy. FDCS cells are spindle- and oval-shaped with plump cytoplasm arranged in whorl and storiform patterns. The cytoplasm is eosinophilic and fibrillar. The nuclei are also spindle- and
oval-shaped within nuclear membranes. There are variable degrees of mitotic activity.\textsuperscript{12}

FDCS stains positive for CD21, CD23, and CD35.\textsuperscript{13} Monoclonal FDCS-specific markers include R4/23, KiM4, KiM4p, and Ki-FDC1p.\textsuperscript{10} The reported specificity of the FDCS tumor markers ranges from 63 to 94%.\textsuperscript{14} The diagnosis of FDCS can be elucidated if the cell is immunoreactive to one or more of the above-mentioned FDCS markers.

The differential diagnosis for FDCS includes ectopic meningioma, ectopic thymoma, metastatic carcinoma, malignant melanoma, malignant fibrous histiocytoma, and large-cell lymphoma.\textsuperscript{12} These entities differ from FDCS structurally and immunohistochemically.

The behavior of FDCS is similar to that of low- and intermediate-grade malignant neoplasms and much like that of soft-tissue sarcoma. The similar properties include an increased incidence of local recurrence and occasional regional and distant metastases.\textsuperscript{12}

The optimal treatment of FDCS has not been determined because of the rarity of this tumor. Surgical excision has been the most common treatment. Postoperative radiation and even chemotherapy have been tried, but their survival benefit is unknown.\textsuperscript{12}

References