Tumors arising from follicular dendritic cells are relatively uncommon. Although most of them originate from the lymph nodes, extranodal sites can also be involved. In this article, we report a 62-year-old male case with a very rare tonsillar follicular dendritic cell sarcoma. The diagnosis was based on histopathologic examination and immunohistochemical staining. He was treated successfully with surgical excision, neck dissection and postoperative radiotherapy. No recurrence was observed during a 18 month follow-up.

Key Words: Follicular dendritic cell sarcoma; neck dissection; radiation therapy; tonsil.

Follicular dendritic cells are non-lymphoid and non-phagocytic accessory cells of the immune system and are generally found in the germinal centers of primary and secondary lymphoid follicles.\cite{1,2} They serve as antigen-presenting cells, play a major role in the induction and maintenance of the humoral immune response and stimulate B-cell proliferation and differentiation.\cite{3,4} Follicular dendritic cells have complement receptors and human leukocyte antigen-DR on their surface and can be identified immunohistochemically.\cite{1}

Follicular dendritic cell sarcoma (FDCS) is a rare malignant tumor originating from follicular dendritic cells. Although most of these tumors occur in lymph nodes, various extranodal sites can also be affected. The tonsil is an uncommon site of occurrence for FDCS. Wide local excision with or without neck dissection is the primary treatment of these tumors. Adjuvant treatment such as radiotherapy and chemotherapy are also recommended. We report a rare case of FDCS originating from the right tonsil.

CASE REPORT

A 62-year-old man presented with a three-month history of difficulty on swallowing, a gradually enlarging painless mass around his right tonsil and globus sensation when swallowing. He did not report any symptoms associated with recent upper respiratory infection. There were no co-occurring diseases or history of tobacco intake. A thorough head and neck examination was performed and a 25x15 mm indurated mass located between the anterior and posterior tonsillar pillars was detected (Figure 1). The mass was lobulated and completely occupying the right tonsillar fossa. Neck examination did not reveal any enlarged lymph nodes. Magnetic resonance imaging (MRI) and computed tomography (CT) scans demonstrated a 22x12 mm
lobulated, well-defined soft tissue mass involving the right tonsillar fossa and slightly extending to the parapharyngeal space (Figure 1 and 2). Although the gross appearance of the mass was consistent with a neoplastic disease, a punch biopsy was performed before definitive surgical treatment due to the atypical appearance of the lesion. Histopathologic examination was consistent with FDCS.

The tumor and the right tonsil were excised completely with wide surgical margins. We also performed a right supraomohyoid neck dissection. The diagnosis of FDCS was confirmed by postoperative histopathologic examination. The surgical margins were tumor-free. The tumor was made up of spindle-shaped cells with hyperchromatic nuclei and high mitotic activity under the stratified squamous epithelium (Figure 3). Immunohistochemical staining showed that the tumor cells were positive for CD21 (Figure 4), CD23, CD35, CD68, vimentin and S-100. Histopathologic examination of the neck dissection specimen did not reveal any lymph node metastasis. The patient received radiotherapy following surgical treatment. Postoperative positron emission tomography did not show any evidence of residual or recurrent disease at both primary site and the neck. Follow-up of 18 months showed no recurrence.

**DISCUSSION**

Follicular dendritic cell sarcoma is a rarely seen neoplasm arising from follicular dendritic cells of the immune system. This tumor is generally seen in young and middle-aged adults. Most of these tumors occur in lymph nodes; however, several extranodal sites including the liver, tonsil and intraabdominal soft tissue can also be affected.

As the gross appearance of the tumor is not specific, the diagnosis of FDCS of the tonsil is typically based on histopathologic examination. Histopathological findings of follicular dendritic cell tumors include spindle-shaped arrangement of cells, high mitotic activity and focal storiform or whorled growth pattern of cells. Follicular dendritic cell sarcoma is generally positive for CD21, CD35, KiM4p, KiFDC1p, vimentin, S-100 protein, CD68 and specific muscle actin. CD21 and CD35 are the most useful antibodies because of their sensitivity and specificity. CD21 is expressed strongly in approximately 96% of cases, but occasionally the staining can be patchy or weak. In our case
spindle-shaped cells with hyperchromatic nuclei and high mitotic activity under the stratified squamous epithelium were found in histopathologic examination. Immunohistochemical staining showed that the tumor cells were positive for CD21, CD23, CD35, CD68, vimentin and S-100.

The differential diagnosis of FDCS includes ectopic meningioma, interstitial reticulum cell sarcoma, lymphoepithelial carcinoma, undifferentiated carcinoma, malignant melanoma, thymoma, malignant fibrous histiocytoma and large cell lymphoma. None of these tumors express CD21, CD35, KiM4p or KiFDC1p in immunohistochemical studies and their structural profiles are substantially different from FDCS. The microscopic features such as storiform pattern, syncytial and spindle cells, bland nuclei with small but distinct nucleoli are additional diagnostic clues.

Follicular dendritic cell sarcoma is considered as a low- and intermediate-grade malignant neoplasm. According to Chan et al., several factors may be associated with poor prognosis including tumor size (≥6 cm), intraabdominal location, presence of coagulative necrosis, high mitotic count, significant cellular atypia and lack of adjuvant therapy.

The primary treatment for FDCS is wide surgical excision. According to some authors regional lymph node dissection should be performed only in case of radiologically detected metastasis. Perez-Ordonez et al. suggested that adjuvant radiotherapy may provide some benefit for residual or locally recurrent tumors. Some authors suggest that adjuvant radiotherapy or chemotherapy is indicated in cases with adverse pathologic features, and in advanced or incompletely resected tumors. In the current case, following definitive surgical treatment, adjuvant radiotherapy was performed due to high mitotic activity of the tumor cells.

In conclusion, FDCS should be included in the differential diagnosis of any tonsillar mass. If the diagnosis of FDCS is suspected, immunohistochemical staining should be done. Once the diagnosis is confirmed, wide surgical excision and if required, adjuvant therapies should be performed.

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