Follicular Dendritic Cell Tumor of Neck
- A case report

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ABSTRACT

Follicular dendritic cell (FDC) sarcomas, also known as dendritic reticulum cell tumors, are uncommon neoplasms arising from antigen-presenting cells in B-lymph follicles of nodal and extra-nodal sites. It is considered as an intermediate grade malignancy since it has significant recurrent and metastatic potential. A 45-year-old female presented with a right side neck swelling. Neck dissection was performed. Microscopically, the tumor showed spindle-shaped stromal cells with large oval and polygonal nuclei. Immunohistologically, the cells were positive for CD21 and CD35, consistent with Follicular Dendritic Cell sarcomas. Adjuvant chemotherapy of Cyclophosphamide/Doxorubicin/Vincristine/Prednisone (CHOP) was given. Literature review provides the current information for the diagnosis and treatment of this unusual tumor.

Keywords: Dendritic cell, Sarcoma, Antigen presenting cells, immunohistochemistry, CHOP, Castleman’s Disease

Introduction

Follicular dendritic cell (FDC) sarcomas, also known as dendritic reticulum cell tumors, are uncommon neoplasms arising from antigen-presenting cells in B-lymph follicles of nodal and extra-nodal sites. It is considered as an intermediate grade malignancy since it has significant recurrent and metastatic potential. Neoplasms showing follicular dendritic cell differentiation are uncommon. In recent years, Follicular Dendritic Cell Tumor is receiving increasing attention because of the availability of more sensitive markers to confirm the Follicular Dendritic Cell lineage. Most reported cases have involved lymph nodes of the neck, mediastinum, and axilla. Approximately 30% of the cases are located in extranodal sites, such as palate, pharynx, liver, tonsil, thyroid and intra-abdominal soft tissue. Microscopically, follicular dendritic cell tumor is composed of oval to spindle cells (Figure 5) with eosinophilic cytoplasm arranged in sheets, fascicles, and whorls, sometimes admixed with foci showing a storiform pattern of growth. (Figure 4) The tumor cells are
characteristically admixed with small lymphocytes. The tumor nuclei are oval to spindle, with thin nuclear membranes, small basophilic nucleoli, and clear or dispersed chromatin. Scattered multinucleated tumor cells are frequently seen. Necrosis, marked cellular atypia, high mitotic rate, and abnormal mitoses may occur and are harbingers of an aggressive behavior. The tumor cells typically express CD21, CD35, Ki-M4p, Ki-FDRC1p, and vimentin, with occasional positivity for S-100 protein, muscle-specific actin, and epithelial membrane antigen.

Ultrastructural examination shows complex interdigitating cytoplasmic processes joined by desmosomes. The behavior of these tumors is more akin to that of a low-grade soft tissue sarcoma than a malignant lymphoma and is characterized by local recurrences in 36% of cases and metastases in 28%. A small proportion of cases have arisen against a background of Castleman’s disease of the hyaline-vascular type which suggests that it may represent a precursor lesion. Follicular dendritic cell proliferation and dysplastic changes occurring in Castleman’s disease can form the background from which a Follicular dendritic cell sarcoma develops and others in association with Epstein-Barr virus infection. Follicular dendritic cell sarcoma has a slight recurrent and metastatic potential and it should be viewed as an intermediate grade malignancy. Complete surgical resection is the treatment of choice whenever feasible. Adjuvant radiotherapy or chemotherapy (CHOP REGIMEN) appears indicated in cases having adverse pathological features and in recurrent or incompletely resected lesions. But the value of these adjuvant treatments to effectively improve survival rates has not been convincingly demonstrated.

Much still needs to be learned about the most effective adjuvant therapy and the molecular biology of these tumors.

**Conclusion**

**Follicular Dendritic Cell Tumor**

Tumor is a rare tumor which is difficult to diagnose based on its non-specific presentation. Diagnosis of the Follicular dendritic cell sarcoma is based on the findings of morphology and immunohistochemistry. Follicular dendritic cell sarcoma exhibits distinctive histologic features that permit its presumptive recognition, but a firm diagnosis requires confirmation with special studies. Because it has a significant recurrent and metastatic potential (the latter risk having been previously underestimated), it should be viewed as an intermediate grade malignancy.

**REFERENCES**


**Figure 1**: Presentation at Preoperative phase

**Figure 2**: Post op Day 2

**Figure 3**: Histopathology appearance Scattered Multinucleated Tumor Cells with eosinophilic cytoplasm arranged in sheets.

**Figure 4**: Histopathology 2 denoting spindle cells with oval nuclei with scattered small lymphocytes.

**Figure 5**: Post chemotherapy (no recurrence)