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Follicular Dendritic Cell Sarcoma of Lymph Node - a Rare Entity

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Case Report

Follicular Dendritic Cell Sarcoma of Lymph Node - a Rare Entity

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Abstract

Follicular dendritic cells (FDC) are non-lymphoid, non-phagocytic accessory cells in the immune system that are essential for antigen presentation and germinal center reaction regulation[1]. These cells are CD21+, CD35+, CD1a- and S100 protein ± and they show desmosomes ultrastructurally.

The most commonly involved sites by FDC tumors are lymph nodes but may arise at a variety of extranodal sites including oral cavity, tonsil, gastrointestinal tract and liver. Most studies represent single case reports or case series.

Our patient presented with tumor in the lymph nodes. Histology revealed tumor cells with abundant eosinophilic cytoplasm, hyperchromatic and pleomorphic nuclei, and prominent nucleoli. The tumor cells were found to be positive for CD21 which is a specific marker for follicular dendritic cells.

Introduction

Follicular dendritic cells (FDC) are non-lymphoid, non-phagocytic accessory cells in the immune system that are essential for antigen presentation and germinal center reaction regulation.[1] They form tight meshworks within lymphoid follicles. These cells are CD21+, CD35+, CD1a- and S100 protein ± and they show desmosomes ultrastructurally.

Neoplasms showing FDC differentiation were first recognized by Monda et al in 1986.[2] The most commonly involved sites are lymph nodes but may arise at a variety of extranodal sites including oral cavity, tonsil, gastrointestinal tract and liver. Most studies represent single case reports or case series. The apparent rarity may be explained in part by under-recognition of the entity.[3]

Histologically FDC tumor is composed of spindle to ovoid cells that are arranged in fasicles, storiform patterns and whorls. The individual neoplastic cells generally have plump, slightly eosinophilic cytoplasm with indistinct cell borders. The nuclei are elongated, with vesicular or granular finely dispersed chromatin, small but distinct nucleoli, and a delicate nuclear membrane. Lymphocytes are characteristically sprinkled throughout the tumor. Nonetheless, the recognized morphologic spectrum of FDC sarcoma has broadened over the years, such as endocrine tumor like vasculature, polygonal cells, hyaline cytoplasm, and myxoid stroma; and a definitve diagnosis always requires confirmation by immunohistochemical and/ or ultrastructural studies.[4]

We report a case of follicular dendritic cell sarcoma of the lymph node.

Case Report

We describe a 46 year old male having cervical lymphadenopathy since two months. There was no history of generalized lymphadenopathy. General physical and systemic examination was unremarkable. No visceromegaly was present. Haematological examination revealed Haemoglobin 13.8gm/dl, Haematocrit 40%, TLC $6x10^9$/L and Platelets $320x10^9$/L. Excisional biopsy of the lymph node was done which showed effacement of the nodal architecture by a neoplastic lesion exhibiting sheets of large tumor cells with moderate amount of cytoplasm. Spindly appearance of the tumor cells was also noted. The nuclei showed moderate to severe pleomorphism and hyperchromasia (Figure 1). Occasional prominent nucleoli were seen. Abundant mitotic figures were present. No glycogen positivity was seen on special stains. Initial panel of immunohistochemistry was negative for epithelial markers (cytokeratin MNF, CK Cam 5.2, EMA) and Lymphoid panel (LCA, Pan B markers; CD 20 and CD 79a, Ki-1, CD 30, ALK protein) and S-100, CD 68, CD31, CD34, HMB45, Vimentin, ASMA and Desmin. The only positive mother was CD 21 (Figure 2). The case was diagnosed as Follicular dendritic cell sarcoma/tumor according to WHO/REAL classification system.

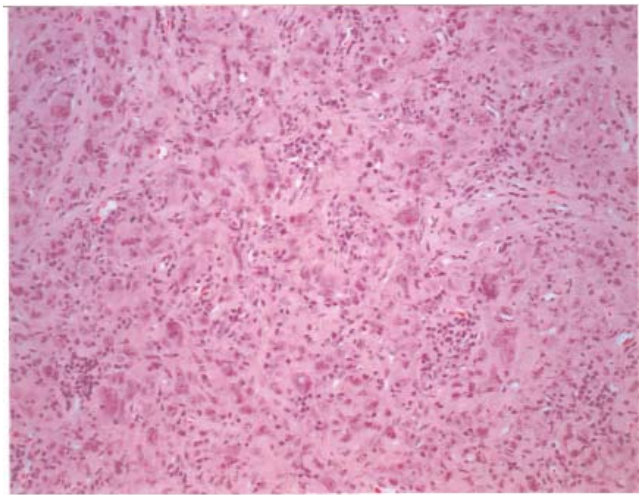

Figure 1:H & E Sections showing effaced nodal architecture by large tumor cells with moderate cytoplasm and pleomorphic nuclei. (Mag 20x)

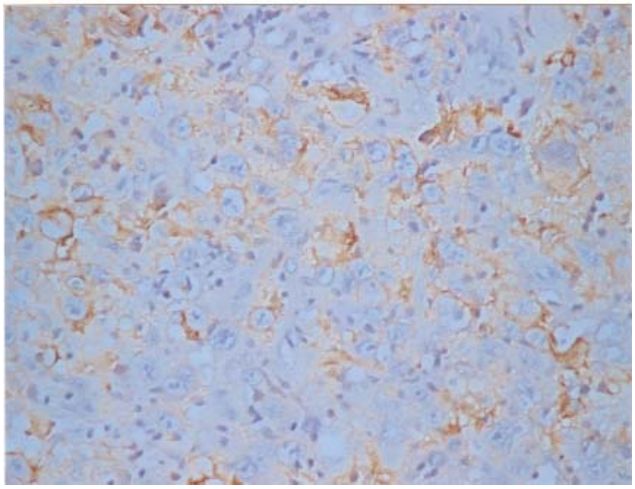

Figure 2. Membrane staining of the tumor cells by CD 21 antibody. (Mag 40x)

Discussion

Monda et al[2] first described Follicular dendritic cell tumor (FDC), since then several other individual cases and small series have been reported.[3-7] It is usually a disease of young and middle aged adults with no sex predilection.

FDC is a low grade malignant neoplasm and distant metastases have been described[3]. The local recurrence rate is 40% and metastatic rate is 25%.[8] When FDC sarcoma metastasizes, the lung is a predilection site in addition to lymph node and liver.[3]

Our patient presented with tumor in the lymph nodes. Histology revealed tumor cells with abundant eosinophilic cytoplasm, hyperchromatic, pleomorphic and prominent nucleoli. The tumor cells were found to be positive for CD21 which is a specific marker for follicular dendritic cells.

The differential diagnosis depends, in part, on the tumor location and may include thymoma, large cell lymphoma, peripheral nerve sheath tumor, histiocytic lymphoma, Langerhan's cell histiocytosis, spindle cell carcinoma and a variety of other mesenchymal lesions. The diagnosis should be confirmed with the use of immunohistochemistry, especially in extranodal organs.

Little is known about the etiology of FDC sarcoma. There is an association of FDC sarcoma with Castleman disease of hyaline-vascular type.[4,9] The tumor has been found to be associated with EBV in cases occurring predominantly in liver and spleen[7,10] usually in cases with an inflammatory pseudotumour like appearance.

In conclusion, FDC sarcoma is a rare entity and awareness of this tumor requires an alert mind, especially if a preliminary panel of immunostains fails to reveal cytokeratin expression.

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