Case Report

Interdigitating dendritic cell tumor

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ABSTRACT

Interdigitating dendritic cell tumor is an extremely rare neoplasm that mainly occurs in lymph nodes. In this case report, we describe the histopathological and clinical features of a typical interdigitating dendritic cell case in a 30-year-old Saudi female who presented with a painless unilateral left neck mass. Microscopically, the lesional tissue in the affected lymph node showed whorls and fascicles of spindle cells intermingling with small lymphocytes. The neoplastic cells were strongly and diffusely positive for S-100 protein, vimentin and alpha 1-antichemotrypsin and focally for macrophages marker (CD68). The combined morphological features and immunohistochemical results were diagnostic of interdigitating dendritic cell tumor.

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Interdigitating dentritic cells are associated with the T-cell areas of peripheral lymphoid organs, namely, in the paracortex of lymph nodes and tonsils and in the periarteriolar lymphoid sheaths of the spleen. Functionally they belong to the category of immune accessory cells involved in antigen presentation to T-lymphocytes. Ultrastructurally they have complex cell prolongations that interdigitate with each other but desmosomes are absent. Immunohistochemically they are reactive for S-100 protein. Neoplastic proliferation of these cells is extremely rare with only approximately 26 reported cases. We report a new case of interdigitating dendritic cell tumor and review the previous reports to point out the main pathological feature that may help in the diagnosis of this rare tumor.

Case Report. A 30-year-old Saudi female presented with a recent history of painless unilateral left neck mass. She had no previous history of malignancy. Radiological investigation showed no other thoracic or intra-abdominal masses and all biochemical and hematological data were reported as within normal limits. The mass was subsequently excised and sent for histopathological assessment.

Gross pathological features. The excised lymph node was almost entirely replaced by a grey white tumor measuring 6 x 5 x 4 cm. The cut surface was firm, nodular with focal areas of hemorrhage.

Histological findings. The lymph node architecture was almost completely effaced by a spindle cell neoplasm with sharp boundaries between the neoplasm and residual normal lymphoid tissue. A proliferation of oval to spindle cells was seen, forming a storiform pattern with occasional whorls, reminiscent of the features commonly observed in meningiomas **(Figure 1)**. The nuclei were oval and had a relatively bland, vesicular chromatin pattern with inconspicuous nucleoli. The mitotic rate was 3 per 10 high power fields. The cytoplasm of the neoplastic cells was abundant, slightly eosinophilic and had indistinct borders. Small lymphocytes were found within the lesion, either as single cells sprinkled among the tumor cells or as discrete

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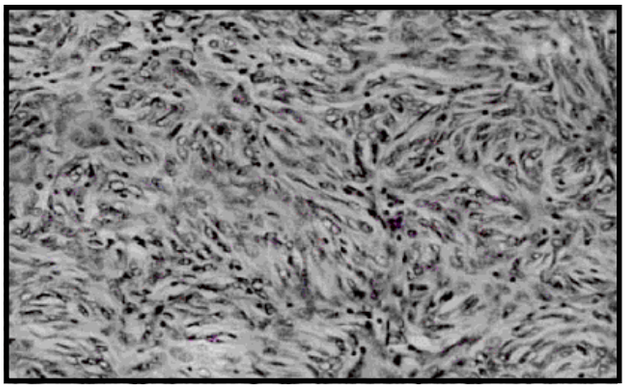

Figure 1 - Interdigitating dendritic cell tumor composed of spindle cells arranged in a clustered and vague storiform pattern. Note the presence of scattered mature lymphocytes within the neoplasm. Hematoxylin & Eosin stain x 200.

collections of cells, often present in perivascular spaces. A panel of immunohistochemical stains was performed on formalin-fixed and paraffin-embedded tissue. The neoplastic spindle cells were strongly and diffusely positive for S-100 protein, vimentin and alpha1-antichemotrypsin. They were also focally positive for macrophages marker (CD68). The tumor cells were however, uniformly negative for leukocyte common antigen, CD20, CD3, CD1a, CD21, CD35, CD34, a melanocyte marker (AMB45), smooth muscle antigen (SMA), muscle specific actin (MSA), desmin, epithelial membrane antigen and AE1/AE3. The small background lymphocytes were strongly positive for leukocyte common antigen and most of these cells were CD3-positive T cells, with only few CD20-positive B cells.

Discussion. Dendritic cells are a heterogenous group of immune accessory cells present in lymphoid and non lymphoid organs. They have a major role in the processing and presentation of antigens to both T and B cells. Based on their immunophenotype, 4 major subtypes have been identified including follicular dendritic cells (FDC), interdigitating dendritic cells (IDC), indeterminate and Langerhan's cells.[1] Neoplasms of dendritic cell origin are among the rarest of tumors affecting the hematopoietic and lymphoid tissue. Interdigitating dendritic cells are found throughout the paracortex of normal lymph nodes, tonsils and in the periarteriolar lymphoid sheaths of the spleen.[2] Ultrastructurally, these cells have complex prolongations that interdigitate with each other, but desmosomes are absent.[3-5] By immunohistochemistry, IDC tumor cells consistently express S-100 protein and vimentin with CD 1a were being negative in most cases. They are negative for markers of FDC (CD21, CD35), myeloperoxidase, CD34, specific B-cell and T-cell associated antigens, CD30, epithelial membrane antigen and cytokeratins. The ki-67 index usually ranges between 10 and 20% (median 11%).[3] The admixed small lymphocytes are almost always of T-cell lineage with few B-cells. Neoplasms of the IDC are uncommon among dendritic cell tumors with fewer than 30 cases reported in the literature. It is generally a disease of adults (mean age 48.5 years). Males are marginally more frequently affected (M:F=15:11)[6] and most of the reported cases have arisen in lymph nodes but extranodal sites such as skin,[7,8] small intestine,[9,10] nasopharynx[11] and testis[12] have been reported. In contrast to the distinctive microscopic appearance of FDC tumors, IDC tumors are morphologically heterogenous. Many of the previously reported cases have demonstrated histiocytic features with pleomorphic nuclei, abundant eosinophilic cytoplasm and a sinusoidal growth pattern.[5,13-15] Nevertheless, one very consistent and diagnostically valuable feature of IDC tumor is the presence of a variable number of reactive background lymphocytes closely intermingled within the tumor cells. Such histological appearance is, however, also shared by neoplasms derived from other dendritic cells, notably follicular dendritic cell tumors. In comparison with follicular dendritic cell tumors, IDC tumors appear to be more clinically aggressive and detailed clinical analysis has been severely limited by the extreme rarity of IDC tumors, and the best treatment protocol is still unknown. The differential diagnosis of IDC neoplasms with spindle cell morphology includes all spindled lesions that occur in the lymph nodes. Metastatic carcinoma, metastatic malignant melanoma with primary Kaposi sarcoma and metastatic sarcomas of other types may be excluded by paying attention to their malignant cytological characteristics and performance of appropriate immuno-histochemical studies. Inflammatory pseudo tumor shares some histological features with IDC tumor like the presence of a significant population of a small reactive lymphocytes. However, the high cellularity and the pleomorphic appearance of the spindle cells in IDC tumors are in direct contrast to inflammatory pseudo tumor.[16] Rarely malignant lymphoma has a spindled appearance and immunohistochemical studies are important for identification of these cases. Furthermore, thymoma may closely resemble IDC tumor, especially since both neoplasms may have collections of lymphocytes in perivascular spaces but the keratin reactivity of thymoma should easily differentiate it from IDC.

In conclusion, in patients presenting with enlarged lymph nodes showing a histological pattern of spindle cell neoplasm; the possibility of the rare dendritic cell tumour should be considered and investigated. The practicing histopathologist and oncologist should be aware of the immunohistochemical characteristics of these neoplasms and of the differential diagnoses that

ought to be considered and excluded in such cases. There is a need to study larger groups of these rare neoplasms in order to establish a reliable and effective therapeutic regime.

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