Follicular Dendritic Cell Sarcoma and its Management: A Case Report and Review

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ABSTRACT

Follicular dendritic cell sarcoma is a rare neoplasm arising in lymph nodes and also in extranodal sites from accessory cells of the immune system which are essential for antigen presentation and germinal centre reaction regulation. Most common sites are cervical or axillary lymphnodes but extranodal sites like oral cavity, tonsil, gastrointestinal tract, soft tissue or breast may occur. It has high potential for recurrence and metastasis. We reported a case of dendritic cell sarcoma of right cervical lymph node in a 60-year old female. The case was treated with lymph node dissection followed by adjuvant radiation. At present, the patient is on regular follow up since last four months.

KEYWORDS: Follicular dendritic cell sarcoma, lymph node, management

INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) is the most common subtype (70%) among all dendritic cell neoplasms. It is a rare neoplasm arising from antigen-presenting cells of the B-cell follicles of nodal and extranodal sites. It was first described in 1986.[1] Approximately only 200 cases have been reported worldwide till 2010. Most of the cases have a median age of 40 years with a slight female predominance.[2] FDCS was more frequently localised at diagnosis than those with interdigitating cell sarcoma (84.7% versus 60%). FDCS typically has a indolent course, but with a metastatic potential.[3] We report a case of FDCS of right cervical lymph node successfully diagnosed and treated with surgery followed by radiotherapy.

CASE REPORT

A 60 year old female was initially presented with swelling over right side neck. Physical examination revealed right level II cervical lymph node enlargement of size 4x4cm, mobile, and hard in consistency (Figure-1). Contrast enhanced CT scan of neck examination revealed right level II cervical lymphadenopathy. Biopsy from right cervical lymph node revealed undifferentiated sarcoma more in favour of dendritic cell sarcoma (Figure-2).

On immunohistochemical examination vimentin, CD35, S-100 markers were positive (Figure-3) and CD1a, CD23, CD34, HMB45 were negative (Figure-4) conforming the diagnosis of FDCS. Further systemic examination ruled out any distant metastasis. Radical lymph node dissection was performed. She was received external beam radiotherapy of 45Gy in 25 fractions to right side upper and lower neck by IMRT technique. Now, she was kept under regular follow up since last four months without any disease.

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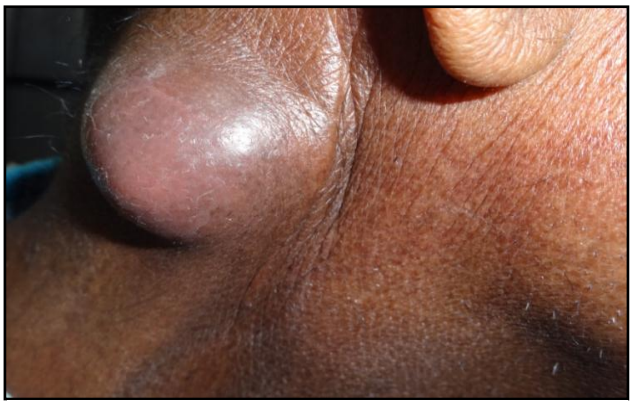

Figure-1: A lymph node of size 4x4cm found in right side of neck

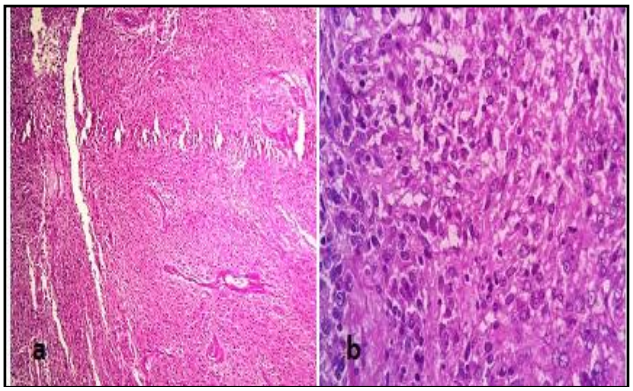

Figure-2: a)Microsection shows spindle cells arranged in fascicles and storiform pattern (H & E,100X), b) Microsection shows oval to spindle cells with eosinophilic cytoplasm, oval nucleus with vesicular chromatin and small distinct nucleoli. There is characteristic sprinkling of small lymphocytes (H & E,400X).

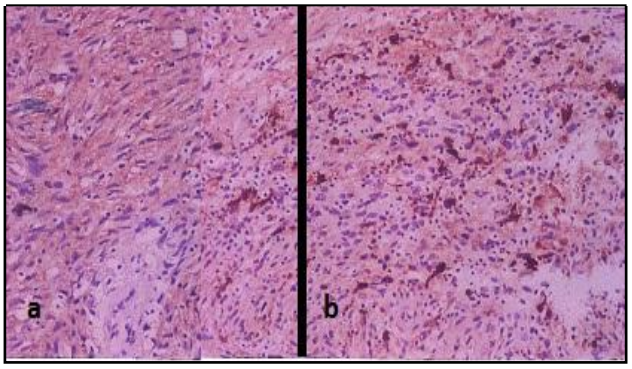

Figure-3: a) Immunohistochemistry showing positivity of CD35, b) Immunohistochemistry showing positivity of S-100

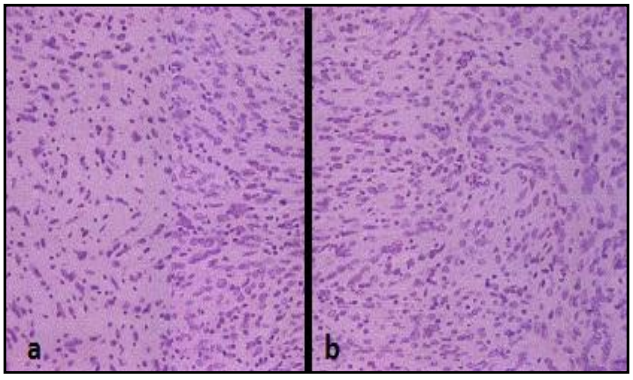

Figure-4: a) Immunohistochemistry showing negativity of CD1a, b) Immunohistochemistry showing negativity of CD23

DISCUSSION

In the 2008 WHO classification of Hematopoetic and Lymphoid Tissue tumours, malignancies originating from histiocytic and dendritic cells such as histiocytic sarcoma, Langerhans cell histiocytosis, Langerhan cell sarcoma, interdigiting dendritic cell sarcoma, disseminated juvenile xanthogranuloma, and FDCS are included in a separate category.[4] Aberrant proliferation of antigen-presenting cells in lymphoid follicles leads to tumour formation. FDCS is a rare neoplasm and can involve both nodal and extranodal sites.[5] Majority of the cases of FDCS occurs at nodal sites. The cervical and axillary group of lymph nodes are predominantly affected.[5] Extranodal FDCS has been reported in sites like the tonsil, spleen, oral cavity, GI tract, lung, skin, and breast.[5] Recent larger reports with longer follow up shows FDCS is an aggressive tumour and should be considered as intermediate malignancy. Storiform, fascicular, and whorled patterns are most commonly seen histologically with sprinkling of small lymphocytes throughout the tumour. The spindle quality is a characteristic feature of neoplastic cells in FDCS. The tumour cells of FDCS reflect the phenotype of non-neoplastic follicular dendritic cells on immunohistochemical staining. The tumour cells are positive for one or more markers among CD21, CD23, and CD35. They are consistently negative for markers like CD1a, CD34, lysozyme, CD3, CD30, CD79a, HMB45 and cytokeratins, and mostly lack S100 expression. Clusterin, fascin, and podoplanin are also positive in FDCS and are useful in differentiating FDCS from other neoplasms arising from histiocytes and dendritic cells.[6] Over expression of epidermal growth factor receptor in tumour cells of FDCS was found in literatures.[7]

The treatment of FDCS is critical, as the optimal treatment has not been defined due to rarity of the disease. Published report on treatment and outcomes are based on retrospective data, and there is no such retrospective data available for FDCS. The first line treatment as combined modality approach with surgery, radiation, and chemotherapy has not been tested prospectively. Surgery is the mainstay of treatment in localized disease. But recurrence rate

can be up to 50%.[8] Therefore adjuvant treatments is necessary. The resected localized disease can be prevented from recurrence by adjuvant radiotherapy or chemotherapy.[9] Relapse rates are better with radiotherapy in comparison to chemotherapy. A recent analysis showed that a trimodality treatment has the best outcome [8].Combined chemotherapeutic regimens designed for NHL like CHOP and its derivatives have been shown to elicit good, objective responses.[7] Targeted therapy, imatinib in combination with gemcitabine and cisplatin have promising outcomes.[10]

CONCLUSION

FDSC is a very rare entity and mostly seen in cervical and axillary group of lymph nodes. Radical lymph node dissection followed by adjuvant radiotherapy and chemotherapy is the treatment of choice. Due to rarity of the disease, the case needs documentation for further knowledge regarding clinical presentation, pathological study, and treatment response. A longer follow up with further studies are necessary to determine whether combination treatment is useful in the treatment of FDCS.

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