Case Report

Case report on follicular dendritic cell sarcoma: a rare malignant neoplasm of lymph node

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ABSTRACT

Follicular dendritic cells (FDC) are non-lymphoid, non-phagocytic, accessory immune cells, crucial for antigen presentation and regulation of the reactions in germinal centers. Sarcoma arising from FDC is very rare and it has low-to-intermediate malignant potential. Histopathological examination play a critical role for the diagnosis of the disease but in difficult cases immunohistochemistry has a great role and facilitates misinterpretation. Diagnosis of dendritic cell sarcoma (DCS) is a challenge for the histopathologists, because of its rare incidence and lack of familiarity of the histopathologists to see the slides. Here is a case of FDCS and the patient presented with left submandibular swelling.

Key words: *Follicular dendritic cell, dendritic cell neoplasm, follicular dendritic cell sarcoma.*

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INTRODUCTION

Dendritic cells are mononuclear phagocytic cells found in various sites and in lymph node.[1] Four types of dendritic cells are present in the lymph nodes: follicular, interdigitating, Langerhans and histiocytic/fibroblastic cells.[2] In other organs, this cell has role to present and process antigen to B lymphocytes, as well as phagocytosis. In lymph node, FDCs do not process antigen for presentation to T cells.[3] Tumours may arise from dendritic cells affecting lymphoid tissues. The occurrence might be <1% of tumours occuring inside the lymph nodes.[3,4] Follicular dendritic cell (FDC) sarcoma is a neoplastic proliferation of spindled to ovoid cells displaying morphological and immunophenotypic functions of FDCs. Chance of misdiagnosis is more, because of its unusual microscopic features, consists of both the features of sarcoma and lymphoma. There is an extensive patient age variety of this tumour and the median age is 50 years. For the most not unusual organ where in follicular dendritic cells sarcoma (FDCS) occurs is in cervical lymph nodes.[5] At the same time, as few cases are located in extra nodal organs which includes liver and spleen.[5] Surgical excision is the primary treatment, now and again mixed with adjuvant radiotherapy or chemotherapy.[6] Histologically, the neoplastic-spindled cells are dispersed inside a prominent lymphoplasmacytic infiltrates. Here, we discuss a case report of follicular dendritic cell sarcoma and its presentation and evaluation.

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CASE REPORT

A 38-year-old male religious teacher presented with gradual swelling at submandibular region for two years. From the beginning of the swelling, he noticed mild pain and occasional fever. For these complaints, he got treatment from local doctor as a case of acute tonsillitis with some medications including antibiotics and analgesics. After taking these medications the pain and fever was subsided but the swelling did not completely disappear. Over the last five months, the size of the swelling became rapidly increased and the diameter of the swelling reached up to 3 cm (Figure 1).

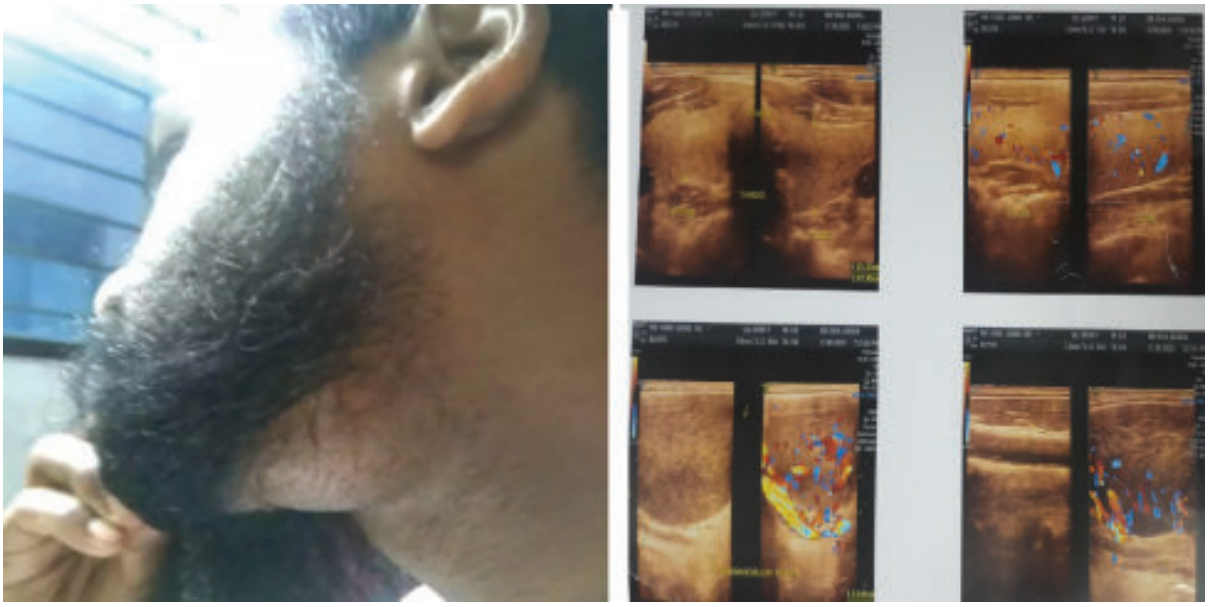

Figure 1. Swelling at submandibular region measuring collectively (3 cm x 2.5 cm x 2 cm) and large solid mass in the left submandibular region in ultrasonography

He also observed about 15 lb weight loss in last seven months and there was generalized weakness. Then he reported to an ENT specialist and the specialist advised for fine needle aspiration cytology (FNAC) from left submandibular swelling along with other related investigations which included haematological, biochemical and radiological investigations. The laboratory investigations revealed normal level of blood sugar, blood urea, serum creatinine, cholesterol, triglycerides and liver function test. His Hb level was 12.4gm/dl. Ultrasonography of left submandibular swelling revealed a mass measuring 5 cm × 3 cm x 4 cm in size with properly-described contour possibly submandibular lymph node.

Chest X-ray was done and revealed no abnormality. FNAC reported as features are suspicious of malignant salivary gland neoplasm but lymphoepithelial carcinoma could not be excluded. A second FNAC from the left submandibular swelling was suggestive of lymphoepithelial neoplasm and advice for excision biopsy was made (Figure 2).

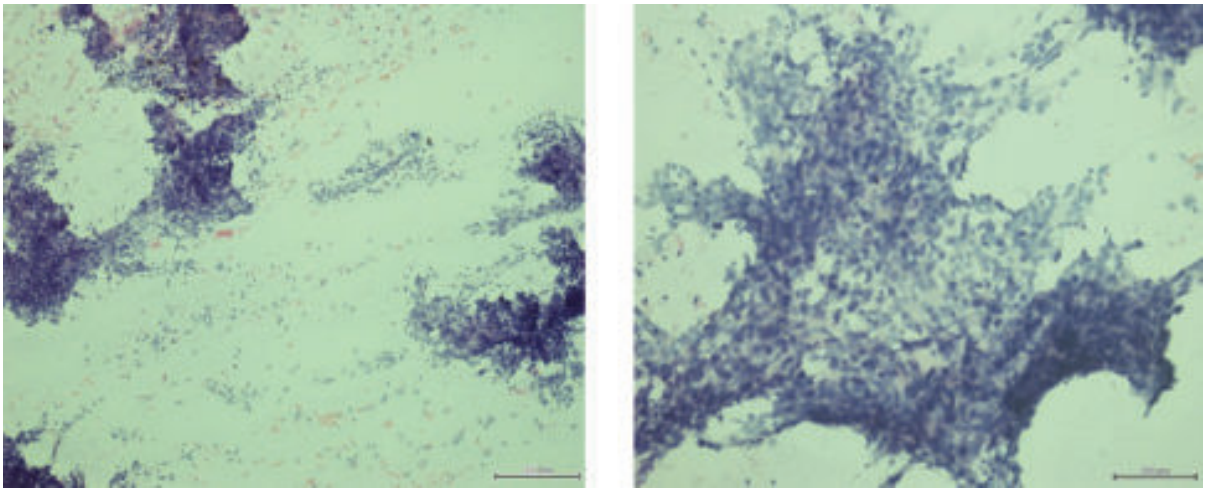

Figure 2. Smears showing presence of spindle to ovoid shaped cells as sheets and singly

The histopathological diagnosis was lymphoepithelial carcinoma and recommended for immunohistochemistry. Then the patient reported to Armed Forces Institute of Pathology (AFIP) with slides and blocks for review and immunohistochemistry for further evaluation. Examination of the histopathology slides revealed the specimen was from a lymph node and the nodal structure was nearly effaced by diffuse proliferations of spindle to oval shaped cells organized in storiform as well as whorled pattern. The neoplastic cells are having vague cellular border, vesicular nuclei with slight amount of cytoplasm. Few binucleate and multinucleate tumour cells are also seen (Figure 3). Then immunohistochemistry markers are applied for further evaluation. Immunohistochemical (IHC) staining confirmed that the neoplastic cells stained positively for CD21, CD23, HLA-DR Fascin and vimentin but the stain turned into negative for S-100, CD68, Pan cytokeratin, CD1a, CD3, CD20, CD4 and EMA (Figure 4). Based on the microscopic findings and the IHC staining, the patient diagnosed as a case of follicular dendritic cell sarcoma.

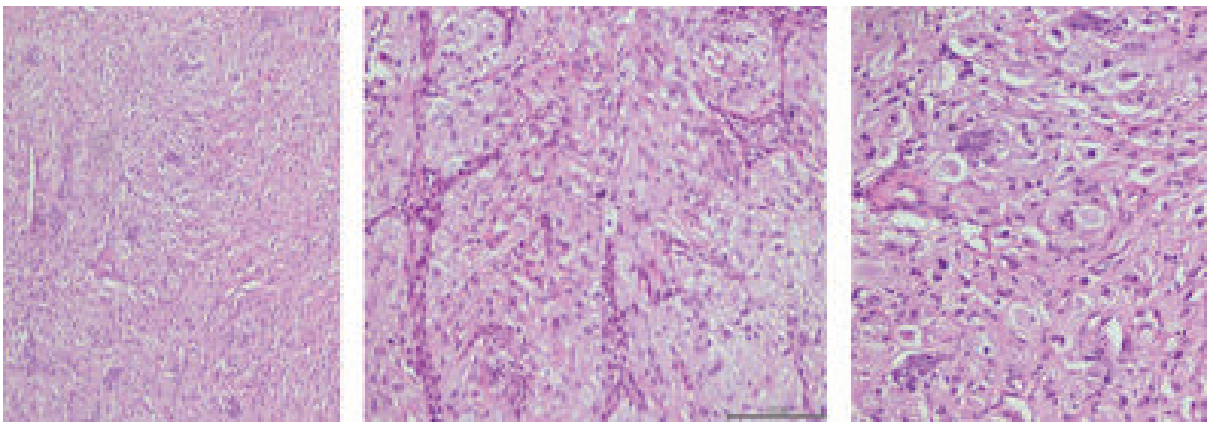

Figure 3. Microscopic examination revealed proliferation of spindle to oval shaped cells organized in storiform as well as whorled pattern

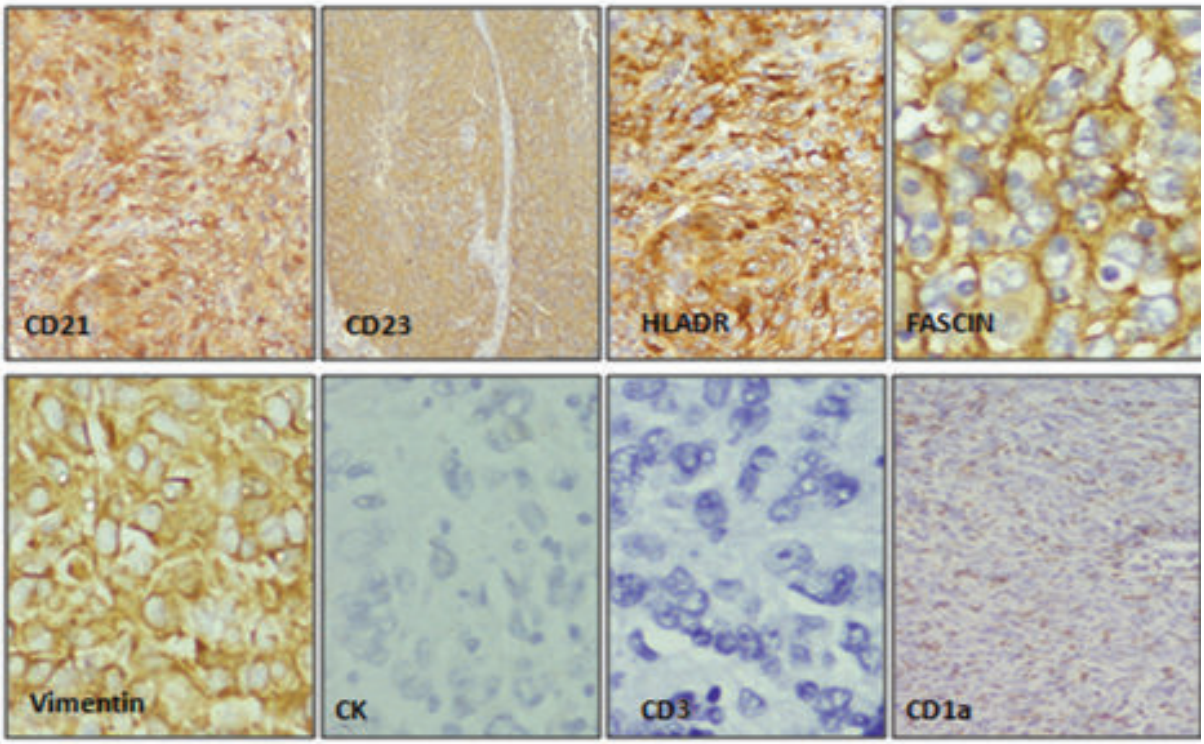

Figure 4. Immunomoreactivity of the different antibody of the case

DISCUSSION

FDCS is a low to mild malignant tumour but it has a high risk to relapse and metastasis.[7] The mechanisms of carcinogenesis of FDCS which includes its initiation and development are normally unknown. It is speculated that FDCS may be derived from hyperplastic dendritic cells which is the useful source of VEGF and about 10-20% of sufferers typically be affected by hyaline-vascular Castleman Disease (HVCD).[8] As a malignant tumour of the lymphatic system, there may be no proof that it has any carcinogenic relation with Epsteim-Barr virus and Epstein-Barr-encoded-RNA, that is taken into consideration as one of the manipulator for the lymphoid neoplasm.[9] It was first describe by Monda's in 1986. FDCS is normally detected in lymph nodes, in particularly the cervical lymph node.[10] FDCS may also developed in different lymph nodes which incorporates the mediastinum, retroperitoneum, mesentery and tonsil. FDCS can also additionally involve extronodal tissues, including the liver, stomach, spleen, pancreas, gut, muscle and skin.[10] Since the medical manifestation of this tumour has no specificity, it is difficult to detect and the correct diagnosis primarily based totally on a histopathological examination. FDCS taking place within the lymph is normally a slow process and presents with painless enlarged lymph nodes. FDCS that occurs intra-abdominally can also additionally metastasize to different organs, including the liver and lung.[11] The cytological features of the tumour cells are characterized through spindle to ovoid or fusiform cells forming typically sheets and nest and singly or it can give fascicles, whorled, diffuse sheet or nodular shapes with lymphocyte infiltration.[12] The tumour cell nuclei are round or oval in shape with vesicular nucleus with clear nuclear membrane. The cytoplasm of the tumour cells appears as thin eosinophilic. A necrosis is usually not seen and the area of coagulative necrosis is commonly determined with poor prognoses.[13]

FDCS in electron microscopy shows long spindled cells connected together by way of desmosome junctions. Immunohistochemical characteristics of FDCS are high quality staining for CD21, CD23 and CD35, which are the main diagnostic markers to distinguish from other diseases which include histeocytic sarcoma, soft tissue sarcoma, lymphoma, and specifically interdigitating dendritic cell sarcoma (IDCS). There are different non-specific high quality immunohistochemistry biomarkers consisting of CD68, S-100, CD1a, D240 and Ki-67. Positive Ki-67 expression is also a crucial index of prognosis.[10]

The potential misdiagnosis of FDCS consists of histiocytic sarcoma (HS), Langerhans cell sarcoma (LCS) and IDCS. The immunohistochemical biomarkers being used most often for those four diseases (FDCS, HS, LCS and IDCS) are CD68, CD1a, CD21, CD23, fascin, clusterin and S-100.[14] The immunohistochemical expression profile is the subsequent: HS: CD68 (100%), CD1a (0%), CD21/CD35 (0%), LYS (94%), S100 (33%); Langerhans cell sarcoma (LCS): CD68 (96%), CD1a (100%), CD21/CD35 (0%), LYS (42%), S100 (100%); inter digitating dendritic cell sarcoma (IDCS): CD68 (50%), CD1a (0%), CD21/CD35 (0%), LYS (25%), S100 (100%); follicular dendritic cell sarcoma (FDCS): CD68 (54%), CD1a (0%), CD21/CD35 (100%), LYS (8%), S-100 (16%).[14] Immunohistochemical tests of CD21, CD23 and CD35 are the most correct indicators for distinguishing FDCS from non-FDCS disease. Currently, there is no valid therapeutic strategy for FDCS. Surgical resection with adequate marginal clearance is the main stay of the disease, and the roles for adjuvant radiotherapy and chemotherapy remain controversial. Reports indicate that the CHOP program has no satisfactory effects, however doxorubicin (DXR), etoposide, methylprednisolone, cisplatin and cytarabine (ESHAP) are recommended.[15]

Consent: Informed written consent was taken from the patient for reporting the case report and any accompanying images.

Conflict of interest: Nothing to declare.

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