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CASE REPORT

Follicular dendritic cell sarcoma of inguinal lymph node – A case report

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Abstract

Follicular dendritic cell sarcomas (FDCS) are rare neoplasms that involve lymph nodes or extranodal sites. They show varied histological features and thus can be mistaken for carcinoma or sarcoma. Correct identification is important for further management.

A 43-year-old Indian female presented with a three-month history of progressive swelling at the right inguinal region. It was excised completely and was reported as lymph node with metastatic poorly differentiated carcinoma based on Haematoxylin and eosin (H&E) stain findings. Computerized tomography (CT) scans of thorax, abdomen and pelvis were normal and did not reveal a primary site. Following this, the case was referred to one of the authors. The slides were reviewed and a variety of immunocytochemical markers were done. The tumour cells were negative for epithelial, melanocytic, neural, leucocyte and soft tissue tumour markers. They were immunopositive for CD21, CD35 and negative for CD68. Based on the immunocytochemical findings, a final diagnosis of FDCS was made. This case highlights the histological and immunophenotypical profile of a rare tumour which requires a high index of suspicion for diagnosis.

Keywords: Lymph node, histiocytic disorders, follicular dendritic cell sarcoma, CD21, CD35.

INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) is a rare tumour of follicular dendritic cells (FDC). FDC are normally present in primary and secondary lymphoid follicles and serve as antigen-presenting cells that play a major role in the initiation and maintenance of humoral immune response.[1] FDCS may occur in lymph nodes of the neck, axilla and mediastinum or at extranodal sites.[2] Histologically, the growth pattern of cells may be more than one pattern such as diffuse, storiform or fascicular. The cells may be ovoid, spindly or epithelioid. The epithelioid cells may mimic metastatic carcinoma. With the usual immunohistochemical work-up, the diagnosis may be missed because FDC markers such as CD21 and CD35 are often not included in the routine panel of antibodies used for investigations of undifferentiated neoplasms. We report a case of FDCS of the inguinal lymph node which mimicked metastatic undifferentiated carcinoma on routine H&E section.

CASE REPORT

A 43-year-old Indian lady presented with a three-month history of progressive swelling in the right inguinal region. The lesion was excised completely. It consisted of a fleshy brownish lymph node measuring 4.5 cm in largest dimension. The initial histological diagnosis was metastatic undifferentiated carcinoma based on H&E stain. Following this, CT scans of thorax, abdomen and pelvis were done to identify a primary site and were found to be normal. Peripheral blood examination was normal and serological tumour markers were negative. The paraffin blocks and sections were sent for a second opinion to one of the authors.

Pathology

The lymph node showed loss of normal architecture and infiltration by epithelioid cells arranged in diffuse pattern (Fig. 1a). The cells had large vesicular nuclei with small nucleoli and pale eosinophilic cytoplasm (Fig. 1b). A few

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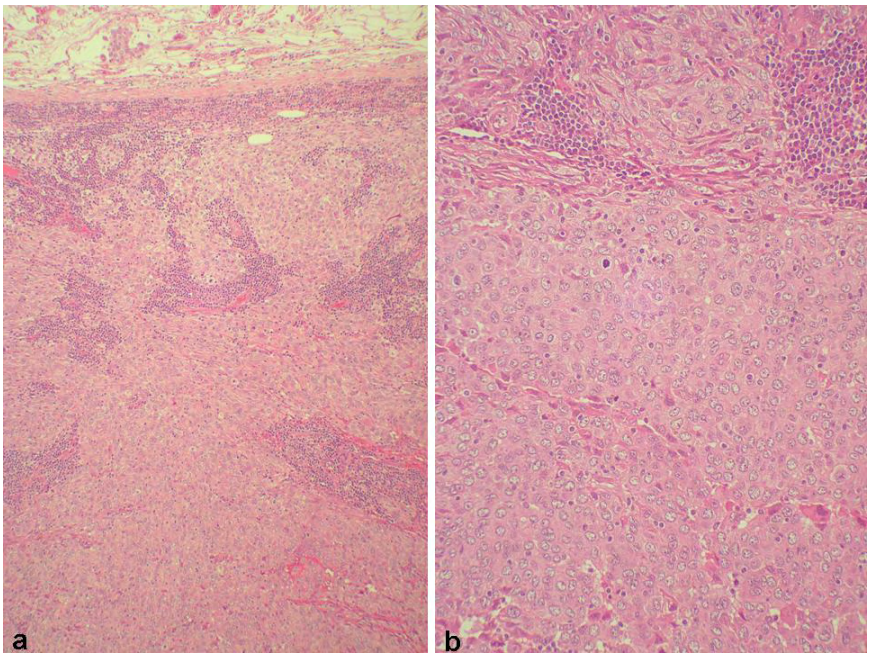

FIG. 1a: Inguinal lymph node showing loss of architecture and replacement with diffuse sheets of pale staining cells (H & E; X 40). 1b: Cells resemble epithelial cells and have indistinct cell borders (H&E; X200).

areas showed spindle-shaped cells (Fig. 2a). An occasional multinucleated cell was also present. Mitotic figures were readily seen, more than 15 per 10 high power fields. Atypical mitotic figures were noted (Fig. 2b). There were no features of Castleman's disease in the residual node, such as presence of prominent lymphoid follicles with vascularisation and hyalinization.

The tumour cells were immunopositive for vimentin, CD21 and CD35. They were negative

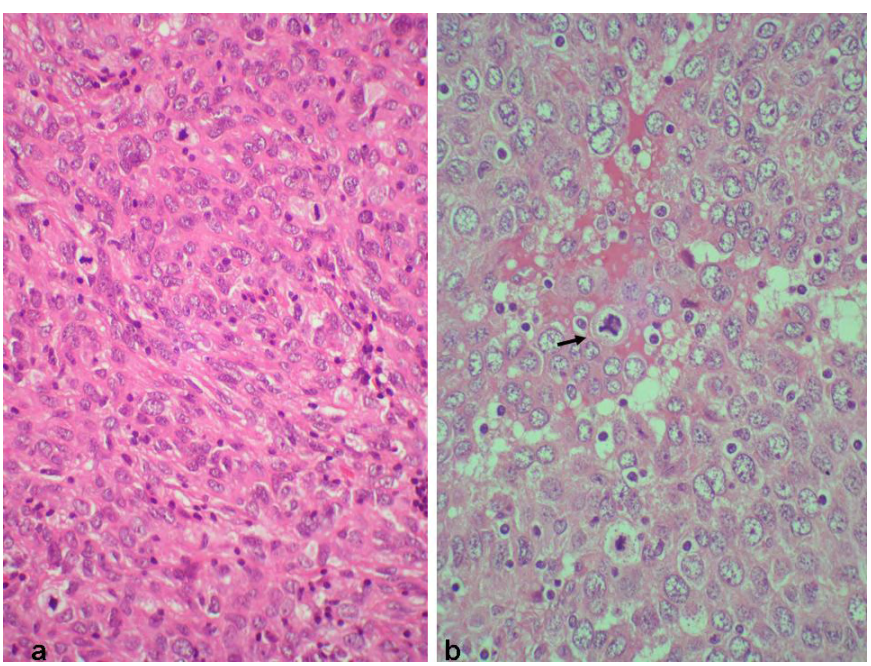

FIG. 2 a: Focal spindle cell pattern with whorled arrangement and numerous mitoses (H&E X 200). b: abnormal mitosis (arrow). (H&E X 400)

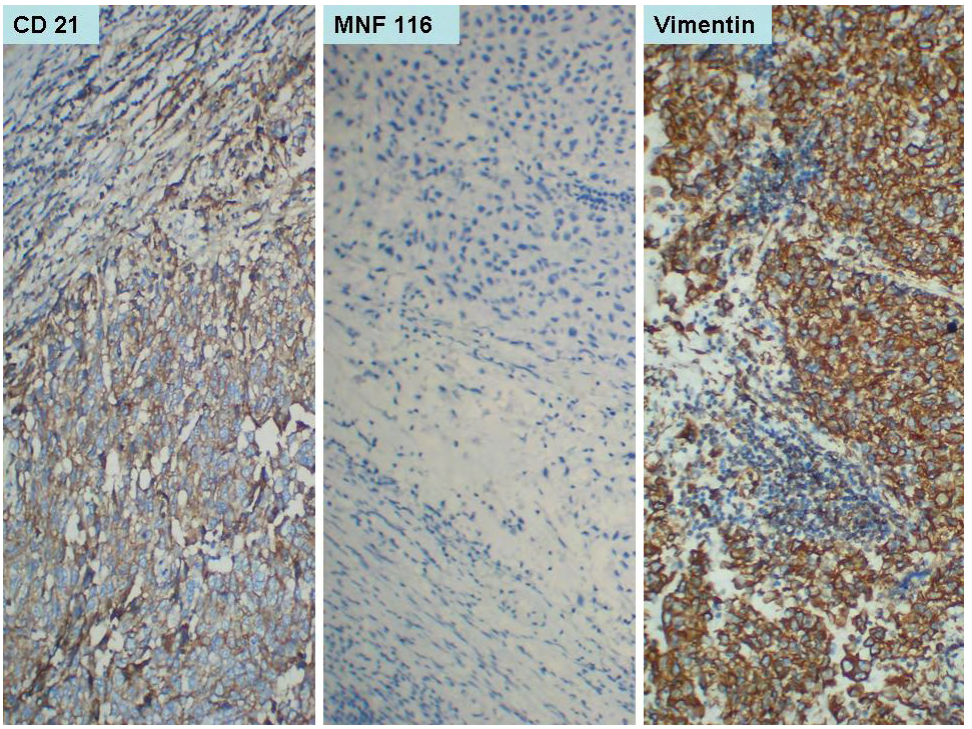

FIG. 3. Tumour cells are positive for CD21 and vimentin, while epithelial marker, MNF116 is negative (Immunoperoxidase stain X 200).

for CD45RO, epithelial membrane antigen, cytokeratin MnF116, cytokeratin AE1/3, S-100 protein, smooth muscle actin, HMB45, CD20, CD3, CD1a, CD68, CD34 and EBER (Fig.3). The combined microscopical and immuno-histological features suggested FDC sarcoma.

Further management

Patient is well 4 months following excision and is referred to an oncologist at the time of publication.

DISCUSSION

Follicular dendritic cell sarcoma is a rare tumour first described in 1986 by Monda *et al*.[3] It predominantly arises in the lymph nodes of young to middle-aged adults. The tumour may present in the extranodal sites such as tonsil, oral cavity, soft tissue,[4] liver,[5] and breast.[6] Most cases of FDCS are published as case reports of single or a few cases. The largest series consisted of 17 cases,[2] 13 cases [3] and a recent report of 14 cases.[7]

FDCS is a tumour of accessory dendritic cell. The International Lymphoma Study Group[8] has classified the tumours of accessory dendritic cells and histiocytes using an immunohistochemical approach based on 61 cases as shown in Table 1.

Follicular dendritic cell sarcomas are usually well-circumscribed and solid. Microscopically the most common arrangement is the storiform pattern. Other patterns include fascicular, whorled, diffuse, myxoid and follicle-like or trabecular patterns. The tumour cells are usually spindly, but can be ovoid or polygonal. Rarely are epithelioid cells seen.[2] Binucleated or multinucleated cells may be seen. A tumour composed of predominantly ovoid or polygonal cells may be mistaken for undifferentiated carcinoma or melanoma. Identification of more typical spindle-cell foci would raise the suspicion of FDCS. The diagnosis of FDCS requires a high index of suspicion and requires confirmation by immunohistochemical stains. CD21 and CD35 directed against the C3d and C3b receptors are the most commonly used FDCS markers and these stains can be done on formalin-fixed paraffin embedded materials.[2] Clusterin and fascin are the recently employed markers.[9]

The aetiology and predisposing factors for FDCS are not ascertained. Association with Epstein-Barr infection has been suggested particularly in extranodal FDCS involving the liver. Five of six cases in the review by

TABLE 1: Immunophenotypic classification and histogenesis of histiocytic and dendritic cell tumours.

Cell of origin	Tumour	CD68	Lysosome	CD1a	S100	CD21	CD35
Macrophage / histiocyte	Histiocytic sarcoma	+	+	-	-/+	-	-
	Langerhan's cell tumour/sarcoma	-/+	-/+	+	+	-	-
Dendritic cel	Interdigitating cell tumour /sarcoma	+/-	- -	-	+	-	-
	Follicular dendritic cell tumour /sarcoma	+/-	-	-	-/+	+	+

+ = 75-100 % of neoplastic cells (NC) are positive; +/- = 50-75% of NC are positive;
-/+ = 25-50% of NC are positive. Rare = 10-25% of NC are positive; - = < 10 % of NC are positive.

Shia et al exhibited positive EBER by in situ hybridization.[10] Some nodal as well as extranodal FDCS appear to be associated with hyaline vascular type Castleman's disease suggesting that it may be a precursor for this condition.[11, 12]

The prognosis of FDCS is not well established. It has been viewed as an indolent tumour with a tendency for local recurrence. Patients are treated with surgical excision with or without adjuvant chemotherapy. Cases with intraabdominal involvement and microscopical features such as significant cytological atypia, extensive coagulative necrosis, high proliferative index (mitotic count of >5/10 HPF), tumour size greater than 6 cm. and lack of adjuvant therapy have poor prognosis whereas lesions arising in lymph nodes behave as low grade sarcoma with a relatively good prognosis.[2] Soriano *et al*[7] observed that patients who underwent a combination of surgery, chemotherapy and radiation therapy as initial treatment had longer disease-free intervals.

In conclusion, we present a case of FDCS arising from the inguinal lymph node which mimicked poorly differentiated carcinoma. Final diagnosis was based on the histological and immunocytochemical findings. Use of CD21 and CD35 stains are helpful in confirming the diagnosis when epithelial markers are negative. Awareness of this entity is important and should be considered in the differential diagnoses of undifferentiated malignant tumour.

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