



Diagnostic Problems Created by Dysplastic Follicular Dendritic Cells in Castleman's Disease on a Trucut Biopsy

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Dear Sir,

Castleman's disease (CD) is a type of reactive lymphoid hyperplasia presenting as nodal or extranodal mass in children and young adults. Common extranodal sites involved are mediastinum, lung, neck, axilla, pelvis and the retroperitoneum. Hyaline vascular variant (HV-CD) and plasma cell variant are the histological types. The histomorphological features of HV-CD are diagnostic; however, the presence of dysplastic follicular dendritic cells (FDC) and proliferation of FDCs encountered rarely, can cause difficulty and diagnostic dilemma especially in trucut biopsies. Here, we report a case of mesenteric castleman's disease which created difficulty in diagnosis on the trucut biopsy due to the presence of dysplastic follicular dendritic cells in the atrophic germinal centres.

A 13 year old boy presented with diffuse abdominal pain and few episodes of non-bilious vomiting and significant weight loss. Clinical examination revealed a firm to hard, non-tender, mobile mass in the left iliac fossa region. Ultrasonogram and computed topography revealed a lobulated soft tissue mass of 11 × 11 × 8 cm below the aortic bifurcation. A trucut biopsy from the lesion showed aggregates of mature lymphoid cells in a fibrocollagenous stroma. Few admixed histiocytes and eosinophils were seen. In addition, there were few large cells with

hyperchromatic nuclei, with nuclear lobulations and abundant cytoplasm in these lymphoid aggregates. The possibility of a Hodgkin lymphoma (HL) was considered due to the polymorphous nature of cells seen, sclerosis and few large cells. A second possibility of an inflammatory myofibroblastic tumor (IMFT) was also considered at this point as it was a mesenteric mass. Intra-operatively, the lesion was arising from the mesentery. The excision specimen was solid, capsulated and bosselated measuring 10 × 10 × 8 cm with homogenous grey white appearance. Microscopy revealed multiple lymphoid follicles with expanded interfollicular areas and areas of sclerosis. The lymphoid follicles showed hyalinised atretic germinal centres and onion skin mantle zones. Interfollicular areas showed prominence of high endothelial vessels, few of which were traversing through the germinal centres forming lollipop lesions (Fig. 1A). Few large cells with multilobation, hyperchromatic nuclei, prominent nucleoli and abundant cytoplasm were noted within the germinal centres, similar to the cells found in the trucut biopsy (Fig. 1B, C, D). No Reed- Stenberg cells were noted. The histomorphological features were classical of HV-CD except for the presence of these large atypical multilobated cells in germinal centres. On immunohistochemistry (IHC), CD 3 and CD 20 highlighted the reactive T and B zones respectively. The atypical cells were positive for CD21 (Fig. 1B-inset), confirming the cells to be follicular dendritic cells (FDC), while they were negative for CD23. The atypical cells were negative for CD30, CD15, PAX5, ALK-1, CD68 ruling out the possibilities of Hodgkin lymphoma (HL) and anaplastic large cell lymphoma (ALCL). EBV - LMP (Ebstein Barr Virus—Latent Membrane Protein) was negative. A final diagnosis of CD-HV with dysplastic FDCs was made based on morphology and IHC findings.

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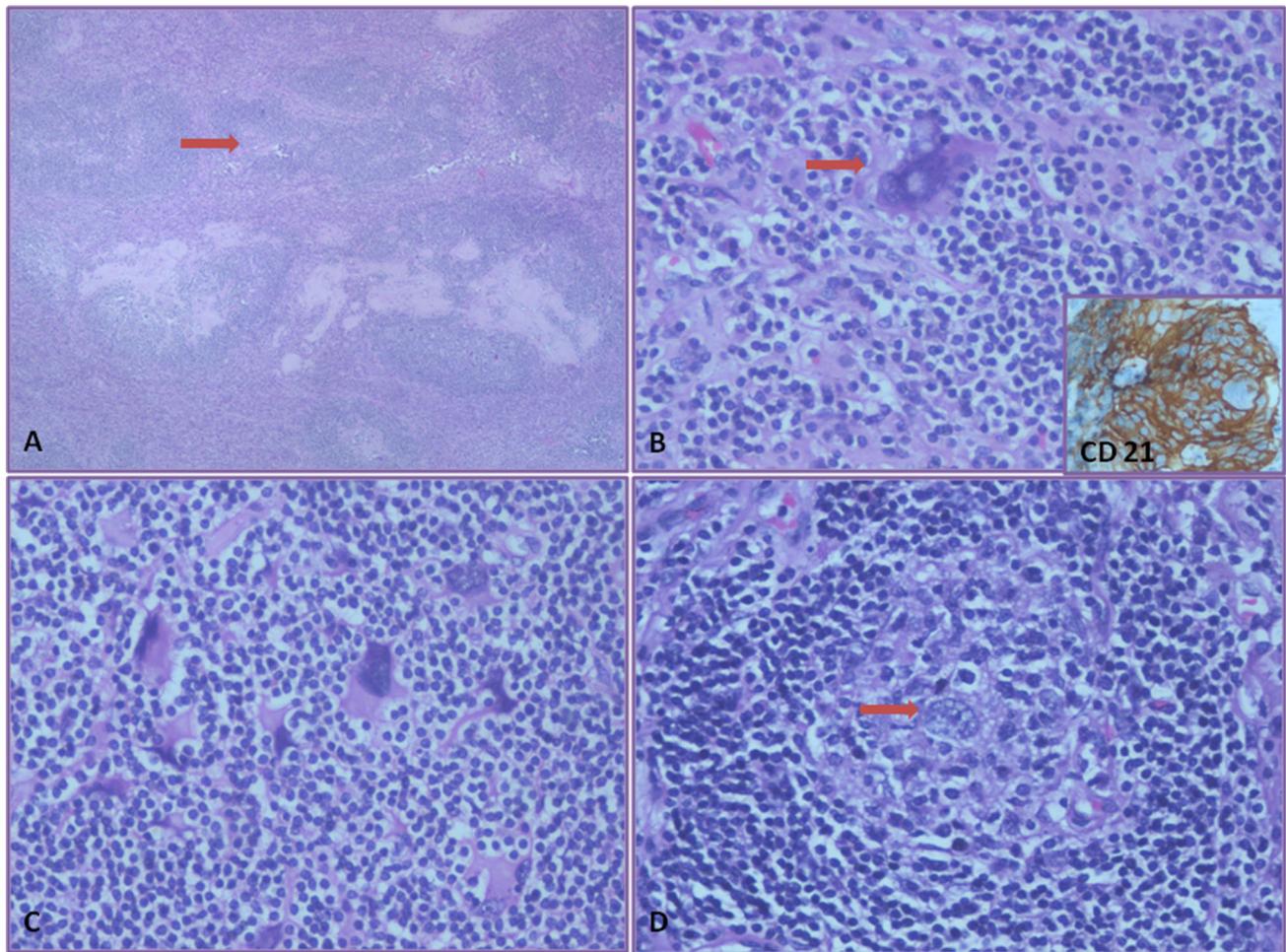


Fig. 1 **A** Low power view of mesenteric mass showing follicles with atretic germinal centres, arrow pointing a lollipop follicle (H&E $\times 100$); **B**, **C** and **D** showing dysplastic follicular dendritic

cells (FDC) in atretic follicles with hyperchromatic, pleomorphic and multinucleated nucleus (H&E $\times 400$); Inset in **B** showing positivity of FDC with CD21 (DAB $\times 400$)

Presence of dysplastic FDCs have been reported in HV-CD [1, 2]. Changes which may happen in the FDC network in HV-CD include expansion and/or disruption, and often proliferations of FDCs in tight clusters. FDCs in HV-CD can be nodules or proliferations that may occur within the atretic germinal centers, mantle zones, or in the interfollicular areas or as scattered cells. When these cells demonstrate cytologic atypia, it is termed as FDC dysplasia. Mass forming proliferation of FDCs with spindled morphology is called as FDC sarcoma [2, 3].

Presence of dysplastic FDCs in CD add to the diagnostic dilemma with the possible differentials of lymphomas having large atypical cells like HL and ALCL, especially on trucut biopsies and fine needle aspiration cytology (FNAC) both in nodal and extranodal sites. The complete morphology of the lesion is not depicted in the small biopsies and presence of these few atypical cells add to further confusion, that too in a site where CD is rare and lymphomas are common. However, immunomarkers can

be helpful in resolving the situation and differentiate lymphomas from dysplastic FDCs in CD. FDC cell sarcoma is a rare malignant neoplasm and 10–20% of these are found to occur in the background of HV-CD. Probably the dysplastic FDC proliferation may be a precursor lesion for FDC sarcoma [3]. Presence of dysplastic FDCs in CD is rare but known; awareness of the presence of these cells in HV-CD should alert the pathologists to consider a possibility of HV-CD especially in trucut biopsies (Fig. 1).

Compliance with Ethical Standards

As this is only a report of a rare histopathological finding in a disease entity, grants were not applicable for this report.

Conflict of interest The authors declare that there are no potential conflicts of interest.

Ethical Approval The study involves human participants. However, all the investigations/procedures done in the patient were done with a diagnostic/therapeutic intent and hence consent is taken from the individuals before the procedure as per institution protocols.

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