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Case Report

Follicular dendritic cell sarcoma of liver: a rare case report

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ABSTRACT

Follicular dendritic cell sarcoma (FDCS) most commonly involves lymph nodes but also affects extranodal sites like liver, spleen, pancreas less commonly. It is a low to intermediate grade malignancy. Unusual presentation, morphology or immunophenotyping makes the diagnosis of FDCS very challenging. Immunohistochemistry plays a major role in confirming the diagnosis of FDCS. A 40 year male presented with dull abdominal pain. Ultrasonography revealed two hypoechoiec lesions in liver. Cytomorphology, later histological and immunohistochemistry (IHC) diagnosis of FDCS was established. Patient was put on chemotherapy.

Keywords: Follicular dendritic cell sarcoma, Liver, Extranodal site

INTRODUCTION

FDCS has morphologic and immunophenotypic features of follicular dendritic cells (FDC), which are immune cells found in germinal centres of lymphoid follicles in secondary and tertiary lymphoid organs, thus rendering the differential diagnosis unusual.[1]

FDCS originate from mesenchymal cell lines, play an integral immunological role in the presentation of antigens to B cells. Primary sarcomas of the liver represent 0.1% of all primary hepatic tumors, FDCS of liver is rare, accounting of only 30% of all FDCS tumors in extranodal sites.[2,3] FDCS can occur at any age, but presents in adults and has no sex predilection. Most patients present with isolated adenopathy, but extranodal disease can also occur. It grows slowly as painless mass, devoid of systemic symptoms. Microscopically, FDCS are composed of spindle to ovoid cells that have range of architectural features such as fascicle formation, storiform arrays, whorls, diffuse sheets and vague nodules.

Complete surgery with or without adjuvant chemotherapy or radiotherapy is the choice of therapy for the treatment of hepatic FDCS. Recurrence of hepatic FDCS and development of distant metastases are major causes of therapeutic failure.

The present case of hepatic FDCS with involvement of lymph nodes was rare. Surgical removal of tumor from various sites was not possible, so chemotherapy was started.

CASE REPORT

A 40 year old male presented with pain in right hypochondrium since 1-1.5 months. Ultrasonography of abdomen was done which showed two hypoechoiec lesions in liver.

USG guided FNAC from liver mass was taken. Cytology smears were air dried and stained with Giemsa stain. The smears showed large histiocytes showing emperipolesis of lymphocytes, plasma cells, polymorphs, sheets of histiocytes, spindle shaped cells, polymorphs, plasma cells and lymphocytes, along with few normal hepatocytes were seen in background. Cytomorphological diagnosis of Rosai Dorfman disease with differential diagnosis of FDCS was made (Figure 1).

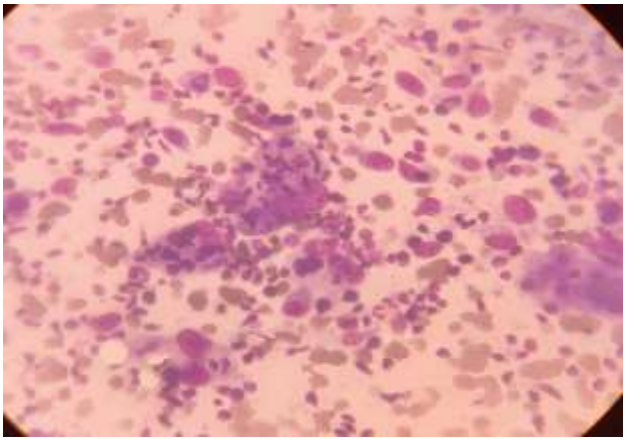

Figure 1: Cytosmears stained with Giemsa stain, 40 ×, shows histiocytes, emperipolesis of lymphocytes, plasma cells, polymorphs, spindle shaped cells, normal hepatocytes.

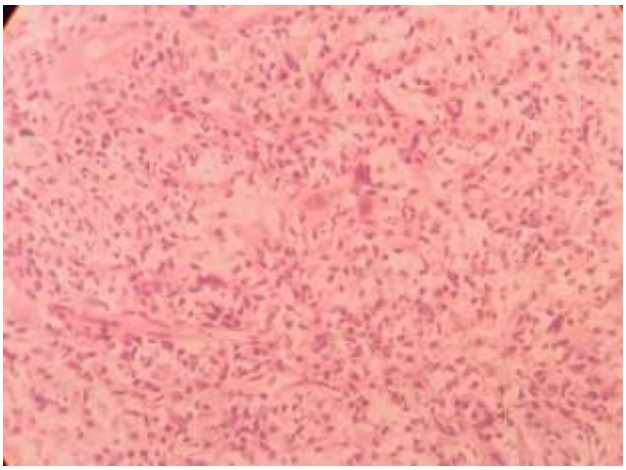

Figure 2: H and E stain, 40 ×; the section shows short fascicles, lymphocytes, histiocytes, plasma cells.

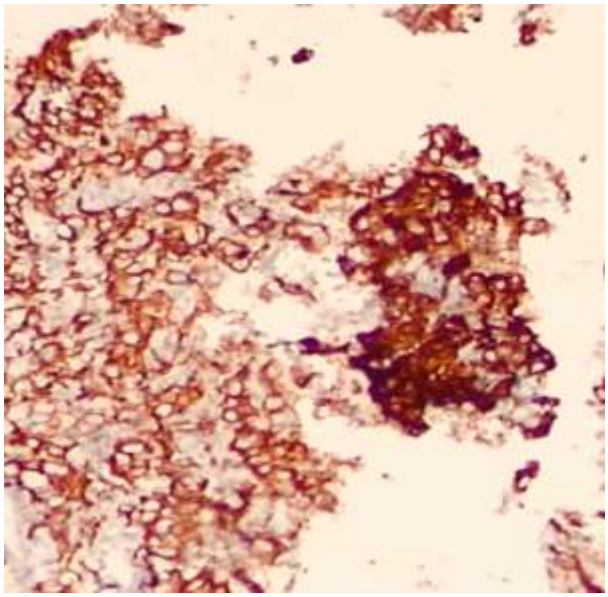

Figure 3: CD 21.

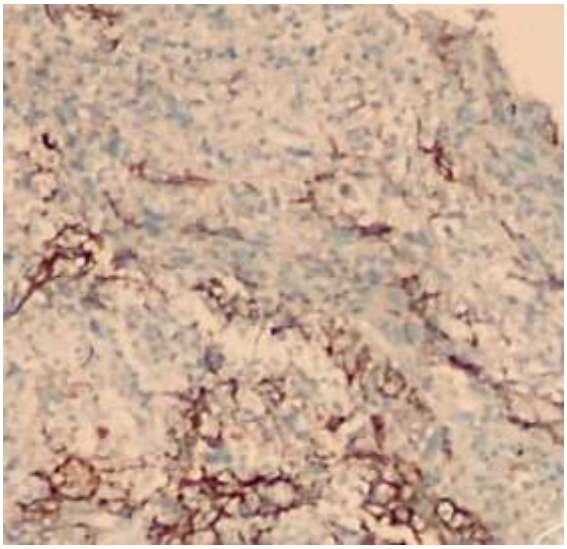

Figure 4: CD 23.

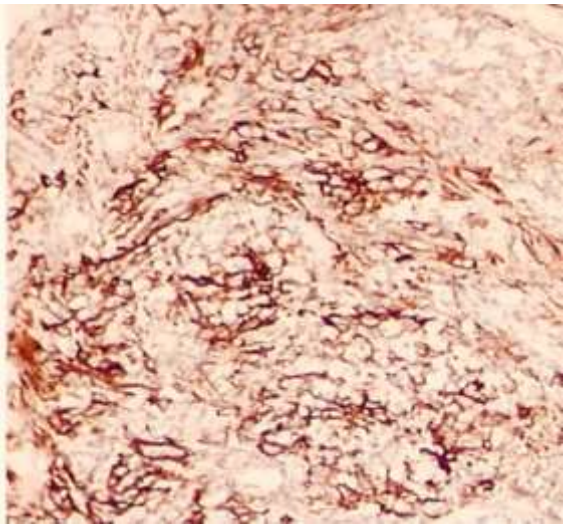

Figure 5: D2-40.

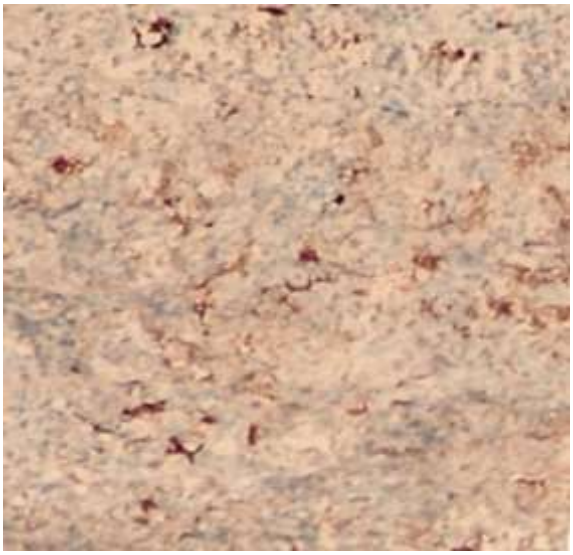

Figure 6: S-100.

CT scan and PET scan was done to see the extent of disease. CT scan done initially showed multiple enlarged, hypoenhancing mass lesion in liver of varying sizes scattered in both lobes, largest measuring 9×8 cm. PET scan done later after about 4 months showed left supraclavicular, left infraclavicular, pericardiac, perigastric (3×1 cm), peripancreatic (2×2 cm), periaortic, periportal, portocaval (3×1 cm), aortocaval lymph nodes with multiple discrete heterogeneously enhancing soft tissue density, hypodense lesions in both lobes of liver parenchyma, largest measuring 12×8×9 cm, with central area of necrosis. Mild diffuse increased FDG uptake in spleen was noted.

Tru cut biopsy using biopsy gun was taken from liver mass for H and E staining and IHC.

H and E stained slides showed spindle shaped cells arranged in short fascicles, at places forming storiform pattern, marked chronic inflammatory cell infiltrate comprising of lymphocytes, histiocytes and plasma cells. Mitotic activity was seen. No necrosis was evident in slides. Histopathological diagnosis of FDCS was made (Figure 2). Differential diagnosis of Rosai Dorfman disease was kept, as there was evidence of emperipolesis and chronic inflammatory cells. Hodgkin's lymphoma was also kept as differential diagnosis. Block was subjected for IHC for confirmation of diagnosis.

IHC showed monomorphic spindle cell proliferation positive for CD 21, focally positive for CD 23, smooth muscle actin, D2-40, CD 68, S-100 (Figure 3-6). Ki 67 proliferation index was 20-30%.

Tumor cells were non-immunoreactive for CK, CD 3, CD 20, CD 30, CD 15, LCA, ALK-1, CK-7, CK 20, CD 117, CD 34, CD 31, Desmin, DOG 1, SDHB, CD 4, CD 8, CD 7, CD 138, EMA, P 63, CD 79a, MUM 1, PAX 5, Glypican, CD 45, Tdt.

Based on cytomorphology, histomorphology and IHC, diagnosis of FDCS was confirmed.

As the lesion was not single, resection of the tumor was not possible. Chemotherapy was started using CHOP (cyclophosphamide, doxorubicin, vincristine, prednisolone) regime.

DISCUSSION

FDCS was a rare malignant tumor derived from the abnormal proliferation and differentiation of follicular dendritic cells in the lymphoid follicles.[1] It was first reported by Monda in 1986.[4] It only accounted for 0.4% of all soft tissue sarcoma but had significant recurrent and metastatic properties.[5] FDCS commonly occured in cervical and axillary lymph nodes, 30% occured in extranodal sites like oral cavity, spleen, liver, small bowel, pancreas, peritoneum, head and neck soft tissue.[3,5,6] The rate of hepatic FDCS was 13%.[8] It had no age or sex predilection in the incidence. It presented as large mass with mean size of 7-10 cm, appeared in relatively young population with median age at presentation of 41 years.[3,8] Most intrabdominal tumors were significantly larger and more aggressive than their extra-abdominal counterparts with frequent sites of metastases in lung, liver and lymph nodes.[8,9] Hyaline vascular type Castleman disease was probably precursor in 20% cases. 12% were associated with EBV infection.[5,10]

Histopathological examination was the gold standard for the diagnosis of FDCS. Hepatic or splenic FDCS was composed of spindle to ovoid shaped cells, lymphocytes, plasmacytes, arranged in clusters, storiform or spiral pattern. IHC markers CD 21, CD 23, Clusterin were generally expressed in FDCS. Rate of Ki 67 positive expression was important index of prognosis.[11]

Pathognomonic evidence for diagnosis of FDCS was microtubuloreticular structures (MTRS) and increased levels of intracellular clusterin.[12]

There were no guidelines for treatment of FDCS as primary hepatic sarcomas were rare. Primary choice of treatment was surgery followed by chemotherapy with CHOP or adjuvant radiotherapy can be used to improve survival in case of resectable FDCS.[4] Local recurrence rate was 23% with metastatic spread rate being 21%.[8]

Data available showed 5 year post operative recurrence free survival rate of 27-32% and 5 year overall survival rate of 79%.[6,8] A higher mitotic count >5/10 hpf and larger size of tumor has strong association with recurrence. Hepatic FDCS behaved like a low grade soft tissue sarcoma.[8]

CONCLUSION

Hepatic FDCS is rare soft tissue sarcoma, diagnosis is confirmed on histological and IHC markers showing positivity for CD 21, CD 23, CD 35, D2-40, Clusterin. CT scan and PET scan helps in detecting the recurrence and metastases of FDCS. FDCS is an important differential diagnosis in liver masses as surgery is the mainstay of treatment if detected early with adjuvant chemotherapy and radiotherapy.

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