Case Report

Follicular dendritic cell sarcoma in supraclavicular lymphnode and gastro intestinal stromal tumor- stomach- synchronous primary neoplasms- A case report

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Abstract:
Follicular dendritic cell sarcoma (FDCS) is an uncommon tumor arising from follicular dendritic cells that normally form meshwork in lymphoid follicles , this case of ours is a 65yr old female , presented with one month history of cervical swelling along with abdominal discomfort . She has a past history of radical mastectomy for carcinoma breast treated with chemoradiation 12 years back. Endoscopy revealed polypoid lesion in the body of stomach. Both the tumors - Lymph node & Gastric lesion, were excised and submitted to routine histopathological examination. Lymphnode swelling was confirmed as FDCS on immunohistochemistry. Polypoidal lesion of stomach was confirmed as Benign Gastro intestinal stromal tumor(GIST).Cases of FDCS associated with other malignancies have been described in literature. However to our knowledge, this is the first report in literature, were FDCS is seen simultaneously with GIST – stomach. However further studies are needed for possible relationship between two such neoplasms occurring concurrently.

Key words: Follicular dendritic cell sarcoma, gastro intestinal tumor, interdigitating cells, intranuclear inclusions, immunohistochemistry

Introduction:
Dendritic cells participate in the immune system as antigen presenting cells. Four different types of dendritic cells exist in lymph nodes, namely, histiocytic, fibroblastic, interdigitating and follicular cells. Follicular dendritic cells, also called dendritic reticulum cells, are seen forming the network in the germinal centers of the lymph nodes. They present antigens to the B lymphocytes so that the memory B cells and plasma cells can be generated [1]. The tumors of these cells are called follicular dendritic cell sarcomas (FDCS), which are rare and are low grade , when compared to intermediate grade neoplasms which may recur after the surgery [2].

Lymph nodes are involved frequently, with the cervical region being most common. Supraclavicular, axillary, mesenteric, and retroperitoneal lymph nodes can also be involved whereas Inguinal lymphnodal group involvement is very rare [3]. FDC sarcoma can also involve extranodal sites including the oral cavity, tonsil, palate, nasopharynx, breast, pleura, gastrointestinal tract, liver, pancreas, spleen, soft tissue, pelvis, and peritoneum [4]. The clinical presentation of FDC sarcoma is of a painless, indolent enlarging mass, however are otherwise asymptomatic [5], account for 0.4% of soft tissue sarcomas, with significant recurrence and metastatic potential and are of intermediate grade malignancy [6].

Case report:
A 65 year old female patient presented with right sided supra clavicular lymphnodal swelling for 2 months. She also complained of abdominal discomfit of one month duration. General examination was nil remarkable with no organomegaly and no other group of lymphnodes were palpable. Routine hematological parameters were within normal limits .Bone marrow examination showed micronormoblastic erythropoiesis .Computed Tomography of neck showed lymph nodal mass measuring 1*0.6*1 cms. Upper Gastro Intestinal Endoscopy revealed polypoidal lesion with central ulceration in body of stomach measuring 4cm in diameter. Excision of supraclavicular lymph nodal mass and gastric polypectomy was performed and submitted for histopathological examination.
Macroscopy:
a. Received supraclavicular lymph nodal mass measuring 4*2*2 cm, was encapsulated and cut surface showed solid grey/white, homogeneous appearance.
b. Received Excision biopsy of sessile polyp from body of stomach measuring 4*3*2 cms. Cut surface showed grey white fleshy appearance. Both specimens were routinely processed.

Microscopy:
a. Lymphnode: Haematoxylin & Eosin sections showed capsule, subcapsular sinus with total effacement of architecture, replaced by ovoid to polygonal cells (epithelioid cell like) intermixed with mature lymphocytes and plasma cells(fig 1 A,B). Neoplastic cells were large, -2-3times the small lymphocyte, seen in sheets, with moderate to scant cytoplasm had round to ovoid vesicular nuclei with irregular nuclear membranes and prominent nucleoli and few showing intranuclear inclusions (fig 1C). Mitotic activity 1-2/10 HPF. No areas of cellular atypia, necrosis, or abnormal mitosis were noticed.
b. Submucosal polyp: Haematoxylin & Eosin stained sections showed ulcerated gastric mucosa.Submucosa showed tumor tissue arranged in whorls and short intersecting fascicles, with frequent and prominent nuclear palisading. Individual cells were bland, spindled, with pale to eosinophilic fibrillar cytoplasm, numerous perinuclear vacuoles that indent nucleus, and extensive stromal hyalinization (fig 5A).Muscularis propria,serosa nil remarkable.

Figure 1: (A) Totally effaced lymphnode (H&E, scanner), (B) tumor tissue arranged in short fascicles,whorls.individual cells are spindled to epithelioid.(H&E 40x), (C) Tumor cell with intra nuclear inclusion.(H&E 100x)

Immunohistochemistry: Performed on paraffin embedded tissue on both specimens, briefed in the following table.

<table>
<thead>
<tr>
<th>IHC marker</th>
<th>CLONE</th>
<th>Gastric polyp</th>
<th>Supraclavicular mass</th>
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<tbody>
<tr>
<td>CD117</td>
<td>Polyclonal rabbit</td>
<td>Positive</td>
<td>Negative</td>
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<tr>
<td>CD23</td>
<td>SP23</td>
<td>Negative</td>
<td>Positive</td>
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<tr>
<td>S100</td>
<td>Polyclonal rabbit</td>
<td>Negative</td>
<td>negative</td>
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<tr>
<td>PCK</td>
<td>AE1 – AE3</td>
<td>Negative</td>
<td>negative</td>
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<tr>
<td>VIMENTIN</td>
<td>V9</td>
<td>Positive</td>
<td>Positive</td>
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Figure 2(A) Immunohistochemistry of lymphnodal mass showing cytoplasmic positivity of Vimentin 40x (B) : showing CD23 membranous positivity 40x

Figure 3 Immunohistochemistry of lymphnodal mass (A) Pancytokeratin 40x (B) S100 negative in the tumor cells 40x

Figure 4 GIST – (A) H&E 40x , (B) Immunohistochemistry CD 117, 40x
Based on histology and Immunohistochemistry we could diagnose Supraclavicular lymphnodal mass as Follicular dendritic cell sarcoma and Gastric polyp as Gastric intestinal stromal tumor. Both are Low grade - occurring as simultaneous primaries.

Discussion

Follicular dendritic cell (FDC) sarcoma was first described in 1986 by Monda et al.[7] on the basis of a series of four cases, all of which occurred in lymph Nodes. In 1994, Chan et al., [8] reported the first 2 cases of extranodal FDC sarcoma, both of which occurred in the oral cavity. The tonsil, nasopharynx, pancreas, peripancreatic, and peritoneal tissues are the most common extranodal sites.

FDCS is the most common histological subtype of dendritic cell tumors with a fairly benign course. It behaves like an intermediate grade sarcoma with a substantial risk of local recurrence (28.1%) and distant metastasis (27.2%). Similar to other soft tissue sarcomas, large tumor size (≥6 cm), presence of coagulative necrosis, high mitotic count (≥5 per 10 high-power fields) and significant cytologic atypia were shown to be associated with poor prognosis [9].

Its etiopathogenesis remains unknown, but Castleman disease, hyaline vascular variant, appears to be a precursor lesion in a subset of patients [10]. EBV is involved in pathogenesis of a small subset of FDCS cases, FDCs express CD21, a receptor for EBV, which enables the entry of oncogenic virus into cell [11].

The diagnosis was challenging as the neoplasm looks very similar to other tumors of the gastrointestinal tracts like gastrointestinal stromal tumor (GIST), solitary fibrous tumor, inflammatory myofibroblastic tumor, smooth muscle and fibrous tumors, fibrohistiocytic tumor, metastatic sarcoma, undifferentiated carcinoma and also melanoma at times. On considering the possibility of a FDCS, the diagnosis requires the application of immunohistochemistry with antibodies to CD21, CD23, and CD35 which aid in diagnosis. They are also usually positive for desmoplaakin, vimentin, fascin, human leukocyte antigen-DR and EMA, and are variably positive for S-100 and CD68. CD20 and CD45 are occasionally expressed.

The typical histologic characteristics described in FDCS include proliferations of spindled or ovoid cells and occasional giant cells with whorled, storiform or fascicular patterns and lymphocyte-rich stromas. Mitotic rates are generally 0–10 per 10 high-powered fields. High grade histologic features such as high mitotic count (>5 per 10 HPF), coagulative necrosis and significant cellular atypia have been described and may be associated with unfavorable outcome [12].

The clinical course of affected patients is variable, but with long clinical follow-up a substantial number of patients developed local recurrence. A smaller subset of patients, including most of those in whom disease recurred developed distant metastases. The most common sites of metastases are the lung, liver, pancreas, and lymph nodes [13].

Treatment administered have varied in accord with clinical context of the affected patients. In patients with localized disease, surgical resection has been the mainstay treatment. The roles of chemotherapy and radiotherapy have not been clearly defined. Several chemotherapeutic regimens have been tried including CHOP, ABVD (doxorubicin, bleomycin, vinblastine, dacarbazone). The longest disease-free survival intervals occurred in patients treated with a combination of surgery, radiation therapy, and chemotherapy.

Conclusion:

To conclude, FDCS is an extremely rare tumor of lymph nodes and extranodal tissues. Once FDCS is suspected histologically, immunohistochemical stains for follicular dendritic cell differentiation should be performed to avoid the risk of misdiagnosis.

This case was an unreported presentation of occurrence of simultaneous FDCS with GIST, which was identified and confirmed by histological and IHC studies.

Source of funding: Nil

Conflicts of interest: Nil

Acknowledgement:

The authors are grateful to authors/editors/publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

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