A Rare Case of Follicular Dendritic Cell Sarcoma with Pseudochylous Effusion and Review of Literature From India

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Case Report

A 55-year-old man, presented with complaints of bilateral neck swelling, abdominal distension and loss of appetite for 2 months. Past medical history was unremarkable. On general physical examination patient was emaciated and had pallor, generalized lymphadenopathy and Beau's line in the nails of both hands (Fig. 1c). Systemic examination showed hepatosplenomegaly and tense ascites. Complete blood counts showed anaemia (Hb-8.9 gm/deciliter), thrombocytopenia (97,000/microliter) and WBC count of 12,900/microliter with normal differentials. Contrast CT imaging of chest and abdomen showed generalized lymphadenopathy, hepatosplenomegaly, gross ascites and moderate bilateral pleural effusion. Excisional biopsy of left axillary lymph node showed effacement of nodal architecture by proliferation of spindle to ovoid shaped tumour cells in the background of reactive lymphocytes, plasma cells and scattered eosinophils. The tumour cells were arranged in fascicles with whirling and storiform pattern (Fig. 2c). Immunostain for CD23, CD68 and S-100 were positive in tumour cells (Fig. 2d–f). On this basis, diagnosis of follicular dendritic cell sarcoma (FDCS) was made. Bone marrow examination

also showed infiltration by spindle to ovoid cells with bland nuclei, irregular nuclear contours and scanty cytoplasm (Fig. 2a, b). Ascitic fluid aspirate was milky in appearance, hence possibility of chylous ascites was kept (Fig. 1a). Ascitic fluid triglyceride level was 59 mg/dl which was within normal limits. After addition of ether to ascitic fluid, no change in the appearance of the fluid was found, which confirmed the diagnosis of pseudo-chylous ascites (Fig. 1b). Malignant cytology of the same were negative on three different occasions. He underwent therapeutic peritoneocentesis for symptomatic relief and was given 1st cycle of adriamycin based chemotherapy (doxorubicin, cyclophosphamide, vincristine, prednisolone). The patient had remarkable improvement following the first course of chemotherapy, with complete resolution of ascites. Currently, he is undergoing remaining chemotherapy.

Discussion

Follicular dendritic cell sarcoma also known as “dendritic reticulum cell sarcoma” is one of the rare malignancies. It has a unique feature of combination of both lymphoma and sarcoma [1]. Follicular dendritic cells are the neoplastic cells localised in germinal centres of lymph nodes which appear like sarcomatous spindle shaped cells. Lymph nodes are the most common site of involvement (approximately 2/3rd) followed by extranodal sites in the remaining cases. A wide variety of extra-nodal sites like colon, lung, thyroid, abdomen and parapharynx have been reported in literature [2–6]. This disease is rare not only due to its low incidence, but also due to its propensity to masquerade other disorders due to similarities in presentation and immunohistochemistry. Recent reports of associations of FDCS with Castleman's disease [5] and pemphigus [6]

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Fig. 1 **a** Milky appearance of ascitic fluid tapped. **b** “Ether test” for confirming the pseudochylous nature of ascitic fluid. After addition of ether in the test-tube (*left side*), there was no clearing of milky appearance of fluid. **c** Transverse depressions along the fingernails (*Beau’s line*)

Fig. 2 **a** Trephine biopsy of the bone marrow infiltration by scattered elongated cells with inconspicuous nucleoli in a background rich in granulocytes. (Haematoxylin and eosin stain, $\times 40$). **b** Multiple elongated cells, with irregular nuclear margins and foldings and small nucleoli infiltrating the marrow spaces. (Trephine Biopsy, Haematoxylin and Eosin stain, Magnification $\times 100$). **c** Lymph node biopsy showing total effacement of architecture by oval to spindle shaped cells arranged in sheets and fascicles. (H&E, $\times 100$). **d** Photomicrograph is highlighting CD23 expression in follicular dendritic cells. (CD23 immunostain $\times 200$). **e** Photomicrograph shows CD68 expressing macrophages in the background. (CD68 Immunostain $\times 100$). **f** Photomicrograph shows few S100 positive interdigitating reticulin cells. (S100 Immunostain $\times 100$)

have added to the variegate presentation of this disease which can cause delay in diagnosis. Another unique subtype of this special entity is an inflammatory pseudo-

tumour like variant. Unlike classical FDCS, this variant has the significant inflammatory background, female predominance, EBV positivity and restriction to abdomen [7].

Table 1 Reported cases of “Histiocytic Sarcoma” from India in world literature and their outcomes

S No.	Author	Year	Sex/ age	Site involved	Treatment given	Outcome
1	Nangal et al. [11]	2014	F/40	Left breast lump	Wide Excision of lump and CHOP × 6 cycles (Planned)	NA
2	Baghmar et al. [6]	2013	M/20	Right hemipelvic mass and paraneoplastic phephigus	RCHOP × 1 cycle	Expired due to Pseudomonas infection
3	Sunder et al. [12]	2013	M/64	Scalp swelling	Wide local excision and local radiotherapy	Died 10 months after surgery
4	Sunder et al. [12]	2013	F/45	Left breast lump	Modified left radical mastectomy with adjuvant radiotherapy	Died 14 months after surgery
5	Mondal et al. [13]	2012	M/27	Left tonsillar swelling	Wide local Excision and Adjuvant radiotherapy	Doing fine last 6 months
6	Malik et al. [14]	2012	M/56	Anal Canal	NA	NA
7	Sunder et al. [15]	2012	F/28	Stomach and jejunal mass	Distal gastrectomy, proximal jejunal resection	NA
8	Patnaik et al. [16]	2012	M/22	Extradural mass at thoracic level	T8, T10 Laminectomy	Alive 5 months after surgery
9	Cyriac et al. [17]	2010	M/69	Neck lump	Chemotherapy plus radiotherapy	Died
10	Vaideswar et al. [18]	2009	M/50	Tonsillar mass	Tonsillectomy	Doing fine after 4 years of surgery
11	Current report	2014	M/55	Generalised lymphadenopathy	Chemotherapy (CHOP)	Doing fine at present

FDCS usually behaves as an indolent tumour. Unlike our case, bone marrow involvement is rare to occur [8].

Due to the rarity of this disease, diagnosing FDCS is challenging for histopathologists. Classically described, FDCS markers on immunohistochemistry are CD 21, CD 23, CD35, clusterin, vimentin and KiM4p [9]. Exact treatment guidelines are still lacking, however complete excision of the mass alone or in combination with radiotherapy/chemotherapy have been done successfully as reported in world literature [10]. We reviewed literature in relation to similar cases reported from India. Total 10 such cases were reported from India and Table (1) illustrates the details of the published cases. Including our case, we report that FDCS is relatively more common in males (M:F = 7:4) with a median age of presentation at 45 years. Interestingly 6/11 cases had extra-nodal involvement and the affected sites were breast, anal canal, stomach and thoracic cavity. The fact that there is no standard guideline for treatment of FDCS is reflected in our literature search as well. Of the 11 cases, four were given chemotherapy, three patients were treated with adjuvant radiotherapy while seven patients underwent surgical resection of the mass. The outcome was reported in six of these case-reports of which four patients died within the first year of diagnosis. Our patient presented with disseminated involvement and is currently undergoing chemotherapy and has shown initial response in the form of complete resolution of ascites.

Conclusion

FDCS usually behaves as an indolent tumour and it can have varied symptomatology depending upon the site of involvement. Our patient's disease was aggressive as he presented with generalized lymphadenopathy and rapid filling ascites along with bone marrow involvement. The rarity of this entity is due to lack of awareness about presentation, morphology, immunohistochemistry. Greater awareness is required amongst pathologists and clinicians to suspect and diagnose this tumor.

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