Follicular Dendritic Cell Sarcoma: A Rare Entity with Variable Pathologic Features and Prognosis

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CLINICAL HISTORY

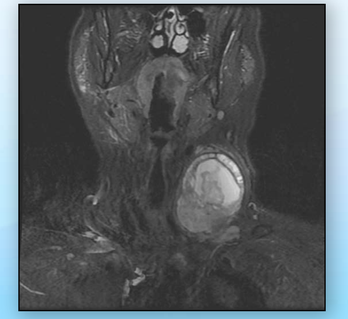


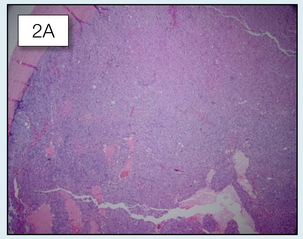
Figure 1. MRI showing left supraclavicular mass.

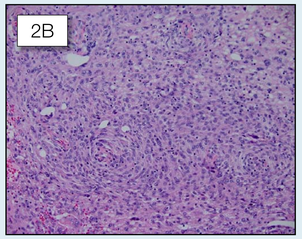
The patient is a 66-year-old female who presented with a left supraclavicular mass (5.7 cm on ultrasound). An MRI scan ***(Fig.1)*** was performed and confirmed a mass displacing the left sternocleidomastoid muscle anteriorly with mixed cystic and solid appearance. CT scan of the chest and abdomen did not show additional lymphadenopathy. The patient reported history of chronic fatigue, and a remote history of right nephrectomy. A CBC was unremarkable except for a chronic normocytic anemia with hemoglobin of 10.4 g/dL.

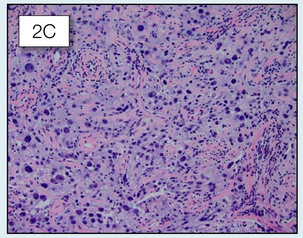
HISTOLOGIC FINDINGS

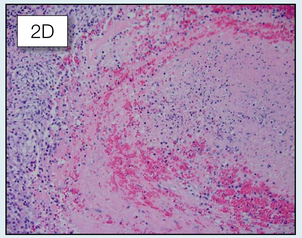
Histologic sections revealed a well-circumscribed, mostly encapsulated tumor composed of spindle to ovoid cells, which in areas formed whorls, particularly around small vessels ***(Fig. 2A, 2B).*** The tumor cells showed indistinct cell borders and nuclei with vesicular to granular chromatin, variably prominent nucleoli, occasional multinucleation, and scattered pseudo-nuclear inclusions. There was significant cytologic atypia/pleomorphism ***(Fig. 2C).*** Areas of necrosis were noted ***(Fig. 2D).*** Perivascular lymphocyte aggregation was also seen. The mitotic count ranged from 4 to 9 mitoses per 10 high power fields.

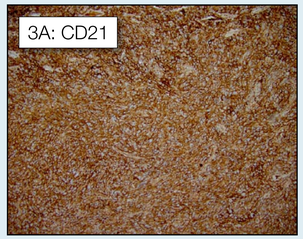
An extensive panel of immunostains was performed ***(Fig. 3)*** and revealed that the tumor cells were strongly and diffusely positive for vimentin, CD21, CD35, CD23, and showed patchy membranous staining with D240. The tumor cells were negative immunostains for CD1a, AE1/3, EMA, CD45, CD20, CD3, CD4, CD8, CD34, CD56, CD68, chromogranin, synaptophysin, p63, PAX8, TTF1, Mart1, S100, and HMB45.

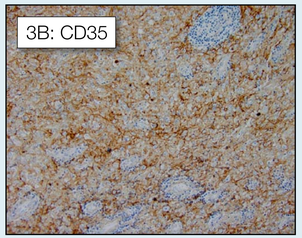


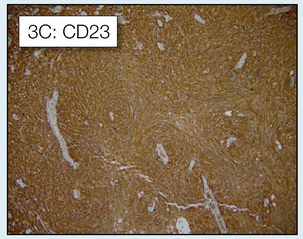


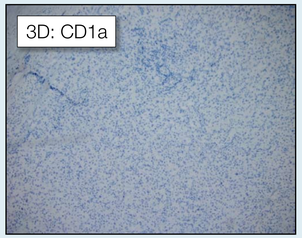












DISCUSSION

Follicular dendritic cell sarcoma (FDCS) is a neoplastic proliferation of spindled to ovoid follicular dendritic cells. This neoplasm is rare with wide age spectrum. The median age of patients according to one study was 48 years, and they had an equal sex distribution. Cervical lymph nodes are the most common site. However disease can present extranodally. Patients usually present with slowly growing painless mass. Systemic symptoms are uncommon. The morphology is that of spindle to ovoid cells forming fascicles, storiform arrays / whorls around blood vessels, diffuse sheets and vague nodularity. The mitotic rate is usually between 0 and 10/ 10 high-power fields (HPF). The more pleomorphic cases demonstrate a higher mitotic rates (>30/ 10 HPF). Nuclear atypia is highly variable. Necrosis and hemorrhage can be seen. The Immunophenotype is always positive for one or more of the FDC markers such as CD21, CD45, CD23 and CD35. The tumor is variably positive for CD68, S100 and negative for CD1a. The behavior is usually indolent. High-grade features such as overt cytologic atypia, extensive coagulative necrosis and a high proliferative index, and large tumour size or intraabdominal location are associated with an adverse prognosis. No prospective studies of treatments and outcomes in patients with FDCS have been reported which make it difficult to provide firm treatment recommendations. However, most patients are treated by complete surgical excision, with or without adjuvant radiotherapy or chemo -therapy. Local recurrences occur in more than 50% of cases, and metastases occur in about 25% of patients. Our case presented with unifocal disease and therefore was treated with excision and radiation therapy. No further clinical follow up on this patient as of this time to evaluate further response.

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