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RESEARCH ARTICLE
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CASE REPORT: FOLLICULAR DENDRITIC CELL SARCOMA OF NECK.A RARE HISTOPATHOLOGICAL VARIANT.ITS PRESENTATION & MANAGEMENT.

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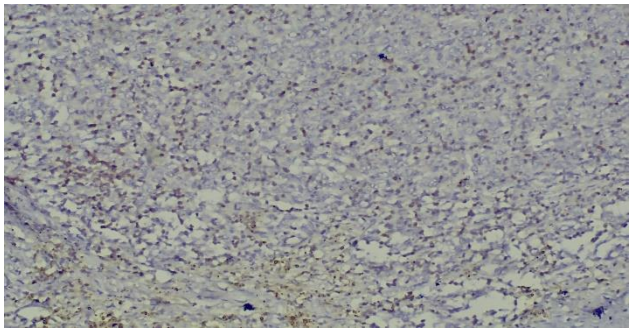
Introduction:
Follicular dendritic cell sarcoma (FDCS), is a rare entity originating from reticular dendritic origin. Dendritic cells are non-lymphoid accessory immune cells which help in captivating and displaying antigens to T & B cells. There are four types of dendritic cells. (a) Follicular dendritic cells (b) Interdigitating dendritic cells. (c) Langerhans cells (d) Fibroblastic cells. It resembles other lymph proliferative disorders especially in view of clinical presentation.it was first reported in 1986 by Monda *et al*.[1] FDCS is characterized as intra-nodal or extra-nodal. Extra-nodal sites include oropharynx, parapharyngeal space, thyroid, neck soft tissue, kidney, axial skeleton, liver and intraabdominal tumors.[2,.] FDCS diagnosis is challenging as approximately 50% of cases are misdiagnosed as undifferentiated carcinomas.Histopathologically , these malignancies are characterized by ovoid, polygonal, and spindle shaped cells arranged in awhorling-type stori-form pattern with admixed lymphocytes.[10] further complicating the issue, fine needle aspirates and biopsies contain necrotic or reactive lymphoid tissue, making the diagnosis more challenging.FDCS is considered to be low grade neoplasm with majority of patients surviving up to 10 years and beyond in most published reports.[2].We report a case of 50 year old male having FDCS of neck ,its presentation, management and outcome of treatment.

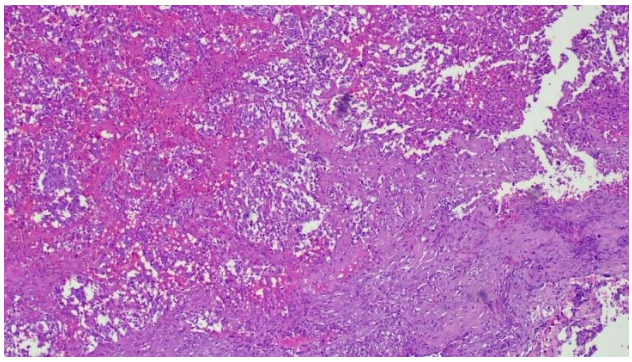
Case report:
A 50 year old male, resident of Nowshehra (KP-PAKISTAN) presented with history of bilateral neck masses, progressively enlarging, firm to hard in consistency, there were no associated B-symptoms. Contrast enhanced MRI Neck revealed level II & III Nodes measuring 7.5x7x4cm deep to Sternocleidomastoid muscle. Contrast enhanced CT Chest abdomen was unremarkable. Biopsy of the mass revealed FOLLICULAR DENDRITIC CELL SARCOMA.A panel of IHC Stains were applied as following:

S.no	Stain	Result
	CK	Negative
	LCA	Positive in few large cells
	CD138	Negative
	SOX10	Negative
	ALK	Negative
	CD3	Negative

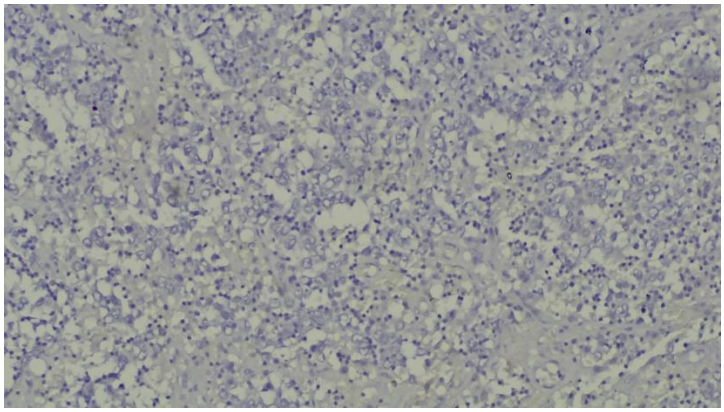
	CD20	Negative
	PAX5	Negative
	KI67	Positive in 90% of tumor cells
	CD68	Highlight histiocytes
	CD34	Negative
	S100	Negative
	CD79a	Negative
	CD35	Focal Positive in tumor cells.
	CD23	Diffuse Positive in tumor cells
	CD21	Diffuse Positive in tumor cells
	CD24	Focal Positive
	Muramidase	Focal Positive in tumor cells
	MUM1	Positive in large cells



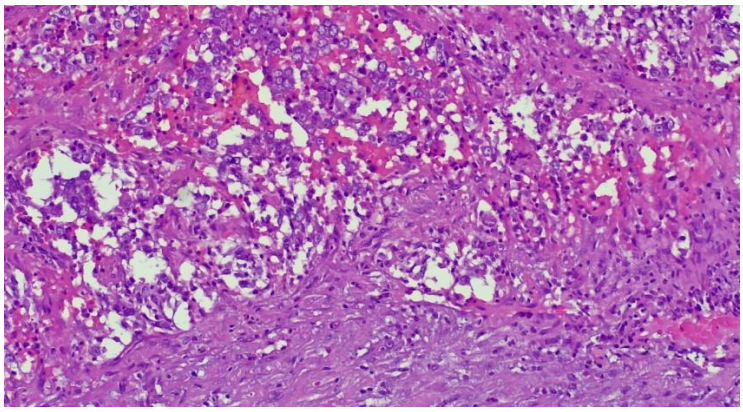
H & E 4x



H &E 10x



H & E 20x



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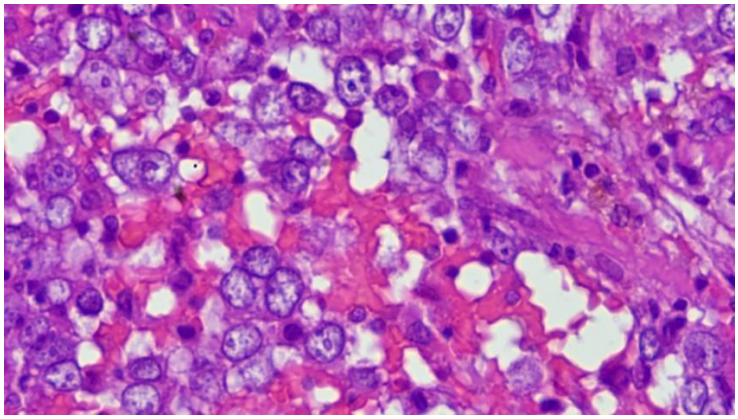
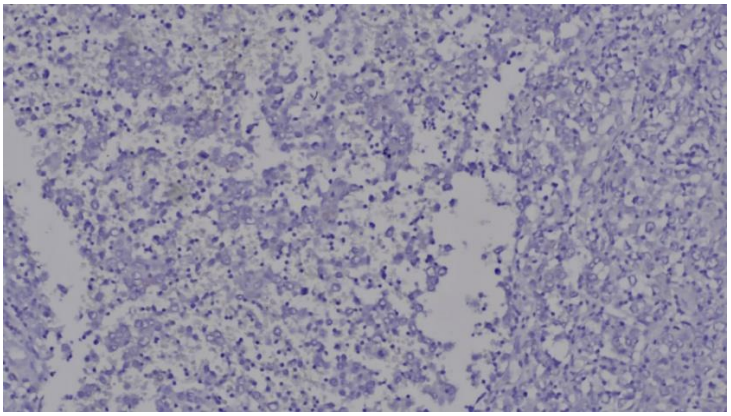
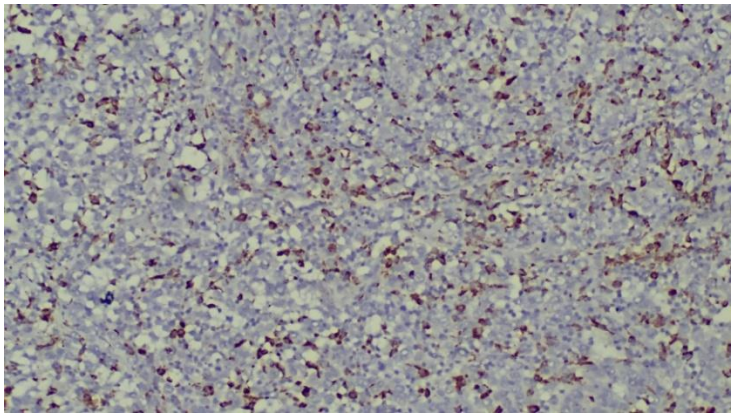


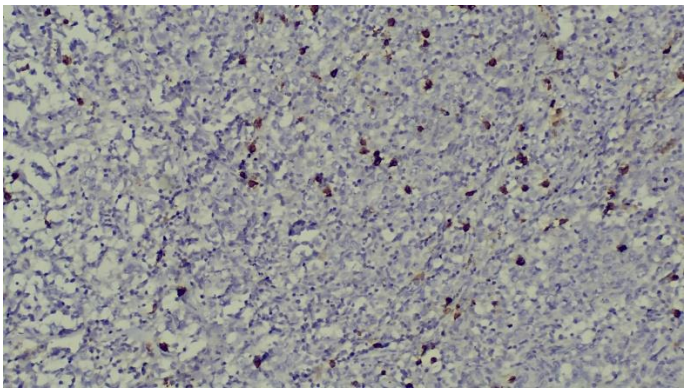
CD 3 (10x)



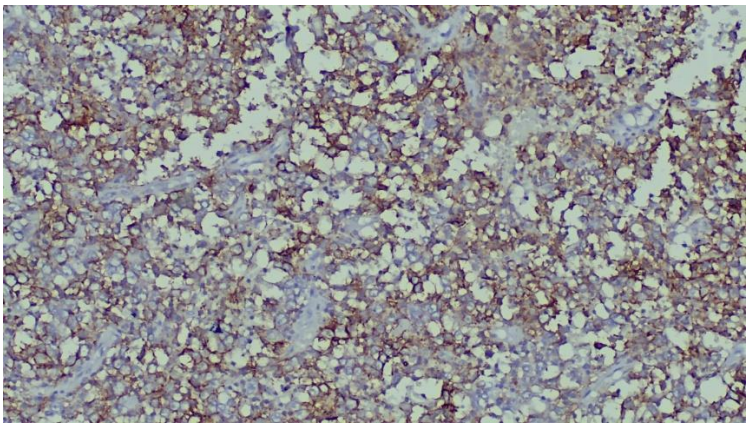
Tdt



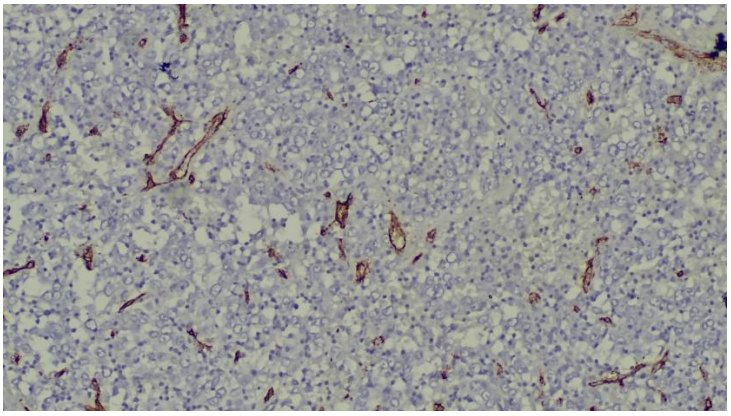
CD68



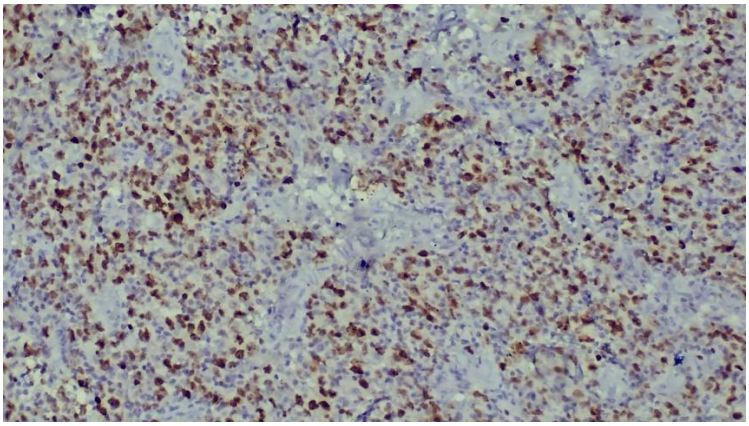
CD 23



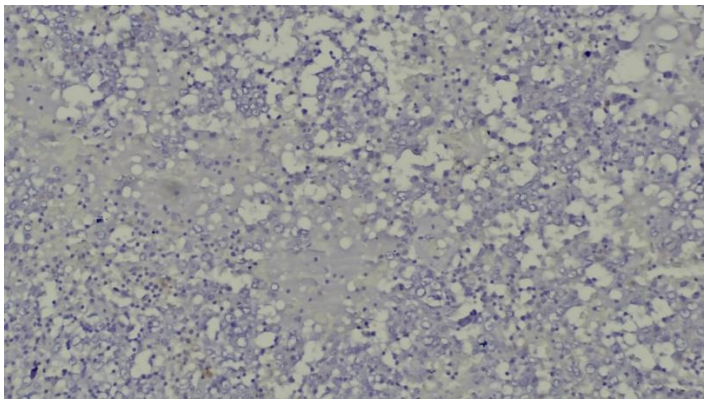
CD 34



MUM 1



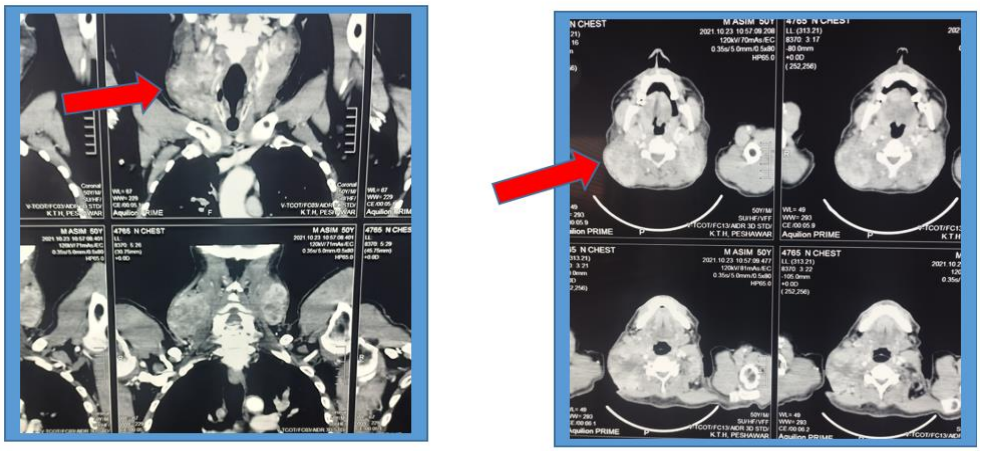
S 100



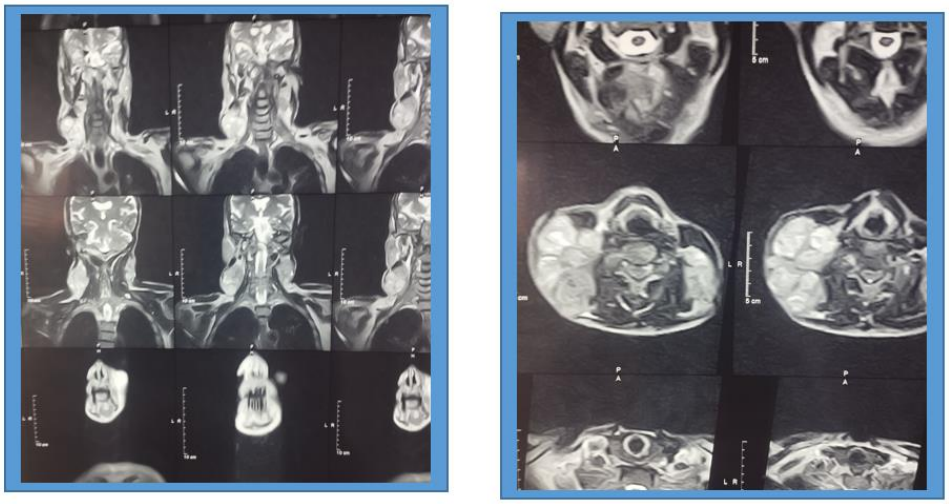
Ki 67

Patient was started on neo-adjuvant chemotherapy ifosfamide and doxorubicin.
After four cycles assessment was done which showed marked clinical response.6 cycles of chemotherapy were completed. Patient was in complete remission. He was given definitive radiotherapy 6,000cGy.Patient is on follow up since 1.8 years.

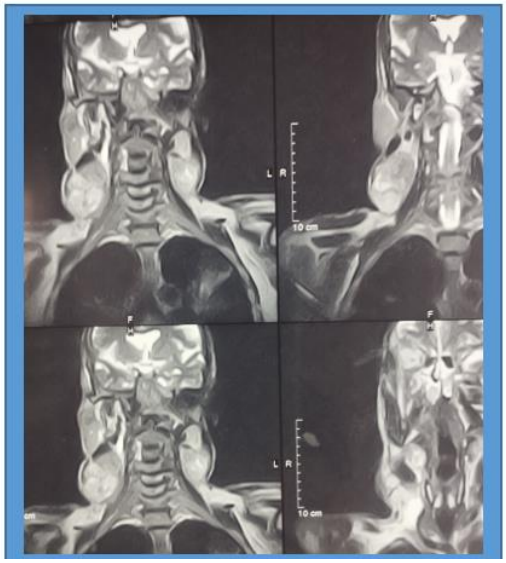
Pre-chemotherapy scans

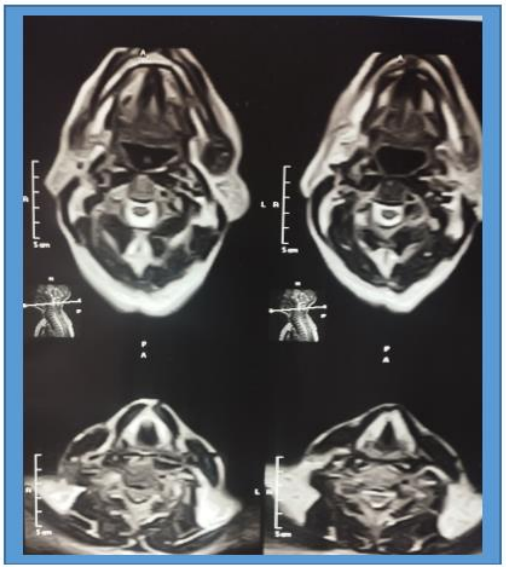


Interim scans



Post chemotherapy scans





Discussion:

Etiology of FDCS is known very little. Very few cases show association with EBV, but it's applicable only to cases of liver and spleen and is associated with an inflammatory peudotumor-like histologic appearance. Approximately 15% of cases reported show association with castle man disease, in that both components are identified histologically at presentation, or the later develops at the same site of a previously diagnosed hyaline-vascular Castle man disease. The evolution from hyaline-vascular Castle man disease to FDCS appears to pass through a phase of extra follicular overgrowth of FDCs that lack significant atypia.[7] In conclusion, for tumors occurring outside mediastinum that demonstrate features suggestive of thymoma or thymic carcinoma (including Castle), the possibility of FDCS has to be seriously considered, especially if a preliminary panel of immunostains fail to reveal cytokeratin expression.[8]

FDCS is grouped with histiocytic and dendritic cell neoplasm in WHO Classification of tumors.FDCS typically occurs in young and middle age adults. Mean age -43 years (range 17-75 years) with no predilection for either gender.[3],[4]FDCS are most common in cervical nodes just as with our case.[4]

S.no	Site	Number of cases
1	CERVICAL NODES	40%
2	OROPHARYNX	24%
3	SOFT TISSUE OF NECK	10%
4	NASOPHARYNX	10%
5	PARAPHARYNGEAL SPACE	08%
6	PAROTID	5%

Currently no consensus exist on standard treatment protocols due to clinical obscurity. But still it is unanimously acceptable that if the lesion is surgically resectable, surgical resection is the mainstay of treatment for all FDCS sites. Patients who have primary in parapharyngeal, oral cavity, neck dissection is preferable. John pang *et al*[4], found in their pooled analysis that chemo radiotherapy should be considered for receiving surgery with adjuvant radiotherapy in contrast to patients with surgery alone. Because they noted that patients receiving surgery plus radiotherapy had a significantly lower loco regional recurrence rate than patients receiving surgery alone (15% vs. 45%;

p=019).therefore it seems that chemo radiotherapy should be considered for patients with FDCS.although it is difficult to make strong recommendations about high risk groups like tumor>4cm, positive margins resection.

Radiotherapy may also be considered in patients with close positive margins <1cm which is recommendation based on National Comprehensive Cancer Network Clinical Practice Guideline for Soft tissue Sarcomas.[5] Approach to post-op radiotherapy is similar to that of carcinoma.60,000-70,000cGy to involved sites and elective nodal radiotherapy.[6]

Chemotherapy can be offered in metastatic setting and as a neoadjuvant chemotherapy. No unified chemotherapy regimen is recommended. Ifosfamide, Carboplatin, Etoposide, Epirubicin, Cyclophosphamide, have been used.however recent studies have elucidated that follicular dendritic cells have a vascular stromal origin rather than hematopoetic origin, a finding that is consistent with their clinical

TUMORS >4CM had worse DFS.FDCS is an intermediate grade malignancy with over all recurrence, metastasis, mortality rate as 43%, 24% and 17% respectively.

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