

Cancer Association of South Africa (CANSA)



Fact Sheet on Follicular Dendritic Cell Sarcoma

Introduction

Follicular dendritic cell sarcoma (FDCS) is an extremely rare neoplasm. While the existence of FDC tumours was predicted by Lennert in 1978, the tumour was not fully recognized as its own cancer until 1986. It accounts for only 0.4% of soft tissue sarcomas, but has significant recurrent and metastatic potential and is considered an intermediate grade malignancy.

It presents as a slowly growing, painless mass with the histologic appearance of spindle-shaped cells in a whorled pattern. Several hundred cases have been reported since it was first described in 1986.

[Picture Credit: Tonsillar Follicular Dendritic Cell Sarcoma]



Follicular Dendritic Cell Sarcoma

Follicular dendritic cell sarcoma (FDCS) is a rare sarcoma, which commonly presents as a slow-growing, painless mass. Tumours of the follicular dendritic cells (FDC-Sarcoma) represent a rare entity with only about 200 annual cases reported worldwide.

Kaur, R., Mehta, J. & Borges, A. 2021.

“Follicular dendritic cell sarcoma (FDCS) is an intermediate-grade malignancy of follicular dendritic cells, which are derived from mesenchymal stem cells. Nodal FDCS is well-recognized. However, when it occurs at an extranodal site, it may not be recognized and is often misdiagnosed. These tumors exhibit a variable spindle to epithelioid cell morphology with a lymphocytic infiltrate and a distinct immunophenotype. The World Health Organization has classified this entity under tumors of hematopoietic and lymphoid tissue, that is, histiocytic and dendritic cell neoplasms. However, its occurrence at extranodal sites and its behavior and management more closely resemble that of a soft tissue tumor. Increased awareness about the existence of FDCS at extranodal sites may aid in the reduction of diagnostic errors. We wish to draw attention to this entity by reporting our experience of 54 cases of extranodal FDCS encountered over a period of 14 years and present a review of the literature of this underrecognized entity. We also describe the ontogeny and molecular pathogenesis of this uncommon lesion.”

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Incidence of Follicular Dendritic Cell Sarcoma (FDC)

The National Cancer Registry (2017) does not provide any information regarding the incidence of follicular dendritic cell sarcoma in South Africa.

Signs and Symptoms of Follicular Dendritic Cell Sarcoma (FDC)

Symptoms depends on where in the body Follicular Dendritic Cell Sarcoma develops, usually a painless swelling of a lymph node, usually in the neck. But a swelling can develop elsewhere in the lymphatic system.

Other symptoms may include:

- a cough
- a sore throat
- difficulty in swallowing
- weight loss
- tiredness
- a painless swelling
- tummy (abdominal pain)
- night sweats
- a raised temperature

Risk Factors for Follicular Dendritic Cell Sarcoma (FDC)

The cause of Follicular Dendritic Cell Sarcoma (FDC) is not known, however, it has been linked to Castleman's Disease – a condition where non-cancerous tumours develop in the lymph nodes. There has also been a link to Epstein Barr virus infection in cases of FDC sarcomas of the liver and spleen.

Diagnosis of Follicular Dendritic Cell Sarcoma (FDC)

A definitive diagnosis is made following a biopsy.

Other tests may include:

- CT scan
- PET scan
- chest x-ray
- blood tests (cancer markers)

Lu, Y., Liu, Q.L., Lu, T., Pang, J.Y., Huo, Z. 2020. Clinicopathological Features of Follicular Dendritic Cell Sarcoma. *Zhongguo Yi Xue Ke Xue Yuan Xue Bao*. 2020 Aug 30;42(4):504-512.

Objective To explore the clinicopathological and immunohistochemical characteristics of follicular dendritic cell sarcoma(FDCS)and the expressions of IgG and IgG4.

Methods We retrospectively analyzed the clinicopathological and immunohistochemical data of 9 pathologically confirmed FDCS cases in Peking Union Medical College Hospital from January 2005 to December 2018.Immunohistochemical staining of IgG and IgG4 were performed,and Epstein-Barr virus(EBV)-encoded RNA(EBER)in situ hybridization were carried out.

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Results Nine cases of FDCS included 4 men and 5 women aged 16-53 years [mean(38.2±9.7)years].The clinical manifestations included masses,lymph node enlargement,rash,and fever.The tumors were located in lymph node,retroperitoneal region,adrenal gland,neck,axillary region,and liver,respectively.Ultrasound showed clear boundary cystic or solid mass with maximum diameters of 1.5-15.0 cm.Microscopically,the spindle tumor cells were arranged in solid and storiform patterns with abundant and slightly stained cytoplasm,vacuolated nuclei,and small nucleoli.The mitosis was 1-3/10 high power fields,and necrosis was found in 5 cases.Immunohistochemically,the tumor cells were positive for CD21(6/9),CD35(6/9),and CD23(7/9).

Conclusions FDCS is a rare malignant tumor,which is easy to be missed.The combination of CD21,CD35,and CD23 is helpful for diagnosis.Hyaline-vascular type Castleman's disease may be the precursor of FDCS,and there may be only a small number of IgG4-positive plasma cells in FDCS.Surgical resection remains the main treatment for FDCS.

Asiry, S., Khade, S.N., Villanueva-Siles, E. & Hakima, L. 2021.

“Follicular dendritic cell sarcoma (FDCS) is a rare malignant neoplasm, which primarily arises in lymph nodes with occasional cases occurring in extranodal locations. The diagnosis is often challenging particularly on cytology fine needle aspiration due to overlapping morphologic and immunohistochemical features. We present a case of FDCS diagnosed in an otherwise asymptomatic 33-year old male. The aim of our case report is to highlight the key cytomorphologic features and discuss various differential diagnoses of this unusual entity.”

Treatment of Follicular Dendritic Cell Sarcoma

The main form of treatment includes complete surgical resection with or without radiation therapy.

The role of adjuvant chemotherapy or radiation is still unclear while the role of chemotherapy in advanced disease is not well established.

Shah, P., Shah, S. & Agostino, N. 2020. Disease response to Pazopanib in follicular dendritic cell sarcoma. *Case Rep Oncol.* 2020 Sep 21;13(3):1131-1135.

“Follicular dendritic cell sarcoma (FDCS) is a rare sarcoma, which commonly presents as a slow-growing, painless mass. There are only a few hundred reported FDCS cases, and the role for adjuvant chemo- or radiation therapy has not been established. Choosing an appropriate therapy in disseminated disease can therefore be challenging. A 26-year-old patient with FDCS was admitted with dyspnea, fever, and night sweats. He was found to have a large right hemothorax with compressive atelectasis on initial imaging. CT of the chest revealed multiple bilateral lung and pleural nodules with associated bilateral hilar adenopathy, a hypodense mass within the right hemithorax, and necrotic right external iliac and inguinal nodes. Inguinal node biopsy diagnosed FDCS. The patient was initially treated with cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy. Gemcitabine/Taxotere was given as second-line therapy and pembrolizumab as third-line therapy, with continued disease progression after 2 cycles of both regimens. The patient was switched to fourth-line therapy with pazopanib and had a partial response for 9 months. This case illustrates a successful FDCS treatment with pazopanib. Due to the rarity of FDCS, where large studies comparing treatment approaches are not available, recommendations for optimal treatment are not well defined. This case is in support of growing evidence suggesting that FDCS responds to systemic therapies that are used for soft tissue sarcoma, such as pazopanib.”

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The [South African National Clinical Trials Register](#) provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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Sources and References Consulted and/or Utilised

Asiry, S., Khade, S.N., Villanueva-Siles, E. & Hakima, L. 2021. Follicular dendritic cell sarcoma: cytomorphologic features and diagnostic challenges. *Diagn Cytopathol.* 2021 Mar;49(3):457-461.

Follicular Dendritic Cell Sarcoma

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Kaur, R., Mehta, J. & Borges, A. 2021. Extranodal follicular dendritic cell sarcoma – a review: “what the mind does not know the eye does not see”. *Adv Anat Pathol.* 2021 Jan;28(1):21-29.

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Tonsillar Follicular Dendritic Cell Sarcoma

<https://ijisrt.com/assets/upload/files/IJISRT21JUN690.pdf>

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