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Follicular Dendritic Cell Sarcoma: A Rare Case Report

Dr. Arya B Nair[1], Dr. Veena R [2] , Dr B S Ajai Kumar [3]
[1]Resident in Pathology, [2]Consultant Pathologists, [3]Chairman and Radiation Oncologist

Abstract
Follicular dendritic cell sarcoma (FDCS) is a rare neoplasm arising from stromal dendritic cells of lymphoid follicles. It can occur in nodal or extra nodal sites, and diagnosis relies on histopathological and immunohistochemical findings. We report a case of FDCS in a 34-year-old man presenting with a right iliac fossa mass, initially suspected to be an extratesticular germ cell tumor. Surgical excision and subsequent histopathological evaluation confirmed the diagnosis of follicular dendritic cell sarcoma. The case emphasizes the importance of considering FDCS in the differential diagnosis of soft tissue tumors.
Keywords: *Follicular dendritic cell sarcoma, lymphoid tissue, CD21, CD23, Vimentin, soft tissue tumor, case report*

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I. Introduction

Follicular dendritic cells (FDCs) are stromal cells of mesenchymal origin located in the lymphoid follicles of B-cell areas. They play a key role in organizing lymphoid tissue architecture, presenting antigens to B cells, clearing debris from germinal centers, and maintaining immune homeostasis (1). FDC proliferation is observed in several reactive and neoplastic conditions such as reactive follicular hyperplasia, follicular lymphoma, mantle cell lymphoma, nodular lymphocyte-predominant Hodgkin lymphoma, and angioimmunoblastic T-cell lymphoma (2). Follicular dendritic cell sarcoma (FDCS) is a neoplasm derived from FDCs and, according to the WHO classification, is grouped under stromal-derived neoplasms of lymphoid tissue (mesenchymal dendritic cell neoplasms) (3). Here, we present a rare case of FDCS in a young male patient.

II. Case Presentation

A 34-year-old man presented with a right iliac fossa mass, without pain, weight loss, or other systemic symptoms. He had no significant past medical history. On physical examination, a fixed, non-tender mass was palpable in the right inguinal region. Laboratory findings included: LDH – 143 U/L, β-hCG – <2.39 IU/L, Na^+ – 146 mmol/L, K^+ – 4.3 mmol/L, Cl^- – 110 mmol/L, Hb – 17.4 g/dL, and HCT – 50.4%.
Contrast-enhanced CT revealed a 6.7 × 5.6 cm heterogeneously enhancing, well-encapsulated soft tissue lesion with necrotic areas in the distal right external iliac region, initially suggestive of a primary extratesticular germ cell tumor.

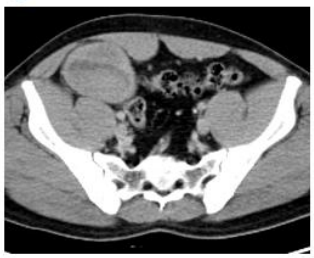

Figure 1. CT scan showing a well-encapsulated soft tissue lesion in the right iliac fossa.

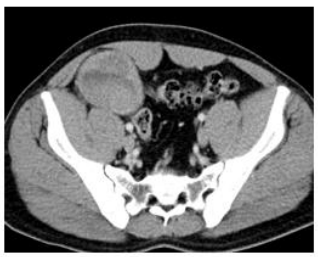

Figure 2. CT axial image showing heterogeneously enhancing soft tissue mass with necrotic areas.

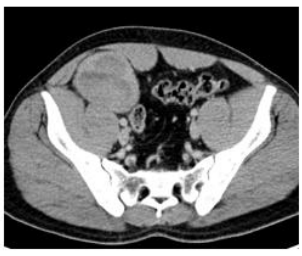

Figure 3. Another CT section showing the extent of the mass adjacent to pelvic structures.

A USG-guided biopsy indicated an undifferentiated malignant tumor. The patient underwent exploratory laparotomy with complete excision of the mass. Intraoperatively, a 6 × 7 cm firm tumor was found abutting the external iliac vein, artery, and genitofemoral nerve. The mass was excised completely while preserving the vas deferens and gonadal vessels.

Grossly, the resected specimen measured 7.5 × 7 × 6 cm, appearing grey-white with areas of hemorrhage and necrosis. Microscopically, sections showed diffuse sheets of spindle to ovoid pleomorphic cells with vesicular nuclei, prominent nucleoli, and moderate eosinophilic cytoplasm. Mitoses (24–25/10 HPF), multinucleated giant cells, and bizarre forms were noted. Thick-walled blood vessels and hemorrhagic areas were seen, with compressed lymphoid tissue at the periphery.

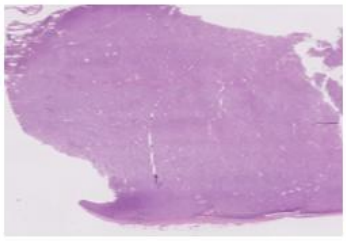

Figure 4. Low-power photomicrograph showing diffuse proliferation of spindle to ovoid cells (H&E, 4x).

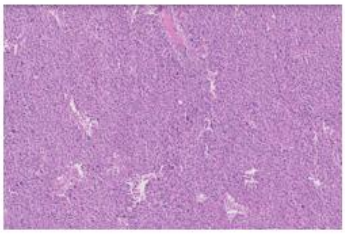

Figure 5. High-power photomicrograph demonstrating pleomorphic tumor cells with vesicular nuclei (H&E, 40x).

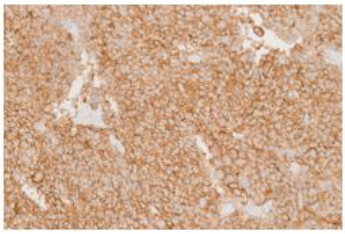

Figure 6. Immunohistochemistry showing diffuse membranous positivity for CD23 (IHC, 40x).

Immunohistochemistry revealed positivity for CD23, CD21, and Vimentin, and negativity for PanCK, SALL4, CD30, LCA, CD3, and CD20. Ki-67 labeling index was 35%. Based on these findings, a diagnosis of follicular dendritic cell sarcoma was made as per WHO classification. The patient received adjuvant radiotherapy and was maintained on pazopanib.

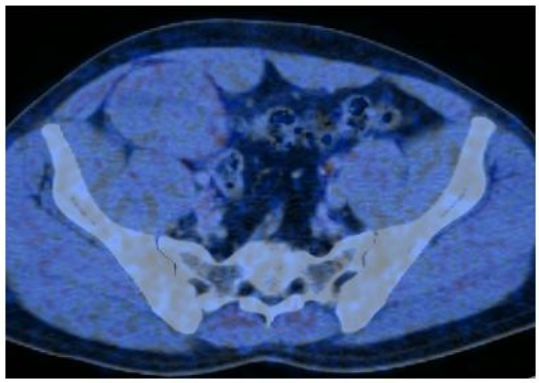

Figure 7. Postoperative PET-CT image demonstrating resolution of lesion post-surgery and adjuvant therapy.

III. Discussion

Follicular dendritic cell sarcoma is a rare malignant tumor exhibiting both morphological and immunophenotypic features of follicular dendritic cells (4). It can arise in lymph nodes or extra nodal sites such as the gastrointestinal tract, retroperitoneum, and upper aerodigestive tract (5,6). Patients commonly present with slow-growing, painless masses, with or without systemic symptoms. The etiology remains unclear, although an association with Epstein–Barr virus has been reported, particularly in inflammatory pseudotumor- like variants (7). Tumor size ranges from 1 to 15 cm, depending on location, with intra-abdominal and mediastinal tumors tending to be larger. There is no significant gender predilection, and FDCS can occur at any age, with a median around 50 years.

Macroscopically, tumors are usually well-circumscribed, solid, and tan-grey, with occasional hemorrhage and necrosis. Histologically, FDCS exhibits spindle to ovoid cells arranged in whorls, fascicles, or storiform patterns with vesicular nuclei, nucleoli, and moderate cytoplasm. Lymphoid aggregates at the periphery or around vessels and multinucleated giant cells may be seen (8). Immunohistochemically, FDCS shows positivity for CD21, CD23, CD35, Vimentin, Fascin, HLA-DR, and EMA (9). Due to its rarity, there are no standardized

treatment protocols. Surgical excision remains the mainstay, with adjuvant radiotherapy or chemotherapy reserved for incomplete excision, recurrence or metastasis (10, 11).

IV. Conclusion

Follicular dendritic cell sarcoma is an uncommon neoplasm with variable clinical and morphological presentations. Accurate diagnosis relies on histopathological and immunohistochemical correlation. Early recognition and complete surgical excision are crucial for favorable outcomes, though the role of adjuvant therapy remains to be clarified.

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