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Abdulkadir Yasir Bahar M.D.
Kahramanmaras Sutcu Imam University School Of Medicine, ayasirbahar@gmail.com

Ülkü Kazancı M. D.
Kahramanmaras Sutcu Imam University School Of Medicine, ulkukazanci@hotmail.com

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Cover Page Footnote
Corresponding Author: Abdulkadir Yasir BAHAR, MD Kahramanmaraş Sütcüimam University, School Of Medicine, Department of Pathology, Kahramanmaraş, TURKEY, ayasirbahar@gmail.com Co-author: Ülkü KAZANCI, MD Kahramanmaraş Sütcüimam University, School Of Medicine, Department of Pathology, Kahramanmaraş, TURKEY, ulkukazanci@hotmail.com

PRIMARY SPLENIC DENDRITIC CELL SARCOMA (FIBROBLASTIC RETICULAR CELL SARCOMA): CASE REPORT OF A VERY RARE MALIGNANCY

B Abdulkadir Yasir MD, K Ülkü MD

INTRODUCTION

Fibroblastic reticulum cell sarcoma may be defined as a malign

proliferation of the histiocytic and dendritic system or accessory immune system cells. This tumor can occur in lymph node, spleen or soft tissue (1-3). The cellular counterparts of this group of neoplasms consist of myeloid stem cell-derived macrophages and dendritic cells and; mesenchymal/stromal stem cell-derived dendritic cells (4).

Fibroblastic reticular cells ensheath the post-capillary venules in reticuloendothelial system and are involved in production and transport of cytokines and other mediators (5). Tumors of these system cells are rare, probably representing less than 1% of tumors presenting in lymph node or soft tissues (2,6)

Historically, some large B cell or T cell lymphomas were named histiocytic or reticulum cell sarcomas.

In this case, diagnosed with primary splenic fibroblastic reticulum cell sarcoma, We aimed to discuss the diagnosis and different diagnosis, histopathologically and immünohistochemically, to contribute to the literature by sharing this rarely seen case along with the literature.

CASE REPORT

A 23-years-old female patient admitted to hospital with the complaint of left side pain one year ago.

Abdominal ultrasound examination showed splenomegaly and a splenic mass.

Thereafter, this finding was confirmed by Computed Tomography (CT), and the whole spleen was filled with mass (Picture 1).

In January 2011, the patient underwent splenectomy. In the pathology report, it was seen as a spindle cell mesenchymal tumor with hematoxylin and eosin stain (Figure 2)

The diagnosis case was registered as fibroblastic reticulum cell sarcoma from the group of histiocytic and dendritic cell neoplasms with immunohistochemical stains.

In the immunohistochemical examination, the tumor cells were strongly positive for vimentin and CD31, weakly for CD68KP1 and CD68PGM1, focally for S100 and smooth muscle actin and, were negative for CD45, CD34, 117, 21, 35, 10, 43, pan-cytokeratin, desmin, chromogranin, synaptophysin, HMB45, and CD1a. (Figure 3,4). The Ki67 score was 40%.

Tomography (PET CT) examination was done for the evaluation of disease diffuseness. It was detected that 18F fluorodeoxyglucose (FDG) involvement increased in the right axillary lymph nodes (max SUV 3,2).

This finding was interpreted as histiocytic sarcoma metastasis. Based on these findings, ıt was administered chemotherapy to the patient. At the end of therapy, new PET CT did not show any pathology favoring the malignity. The patient is followed-up in the remission, and there is no recurrence In the three years.

DISCUSSION

Fibroblastic reticulum cells (FBRCs) are stromal support cells located in the parafollicular area and deep cortex of lymph nodes and also in the extrafollicular areas of the spleen and tonsils. FBRCs have myofibroblastic-like features, in that they are immunohistochemically reactive for vimentin, smooth muscle actin, and desmin and negative for CD21, CD35, and S-100 This immunophenotype differs from follicular dendritic cells (FDCs) and interdigitating dendritic cells (IDCs). FDCs are immunoreactive for CD21, CD35, R4/23, and Ki-M4p, whereas are IDCs immunoreactive for S-100 protein and variably immunoreactive for CD1a and histiocytic markers.

A subset of FBRCs that express cytokeratins 8 and 18 and various other epithelial markers have been identified. Although tumors derived from FBRCs are reasonably well documented, very few cases have been reported. The present case is distinguished from inflammatory pseudotumor by the predominance of spindle cell component with atypical features (1)

The morphological and immunophenotypical features of our present case are consistent with an FBRC origin.

World Health Organization (WHO) included classification of histiocytic and dendritic cell neoplasms the following entities: histiocytic sarcoma, Langerhans cell histiocytosis, Langerhans cell sarcoma, interdigitating dendritic cell sarcoma-tumor, follicular dendritic cell sarcoma-tumor, and dendritic cell sarcoma, not otherwise specified(4).

The rarity of FBRC tumors makes predicting their clinical behavior difficult (7) location has been found to correlate with a particularly aggressive clinical course(7,8).

Although splenectomy was performed in the patients with splenic fibroblastic reticulum cell sarcoma (with or without chemotherapy), fatal clinical outcomes were reported (4). In the cases reported in the literature, poor prognosis and short survival were detected despite chemotherapy. There is no clinical and radiological recurrence in our patient for one year.

FIGURES

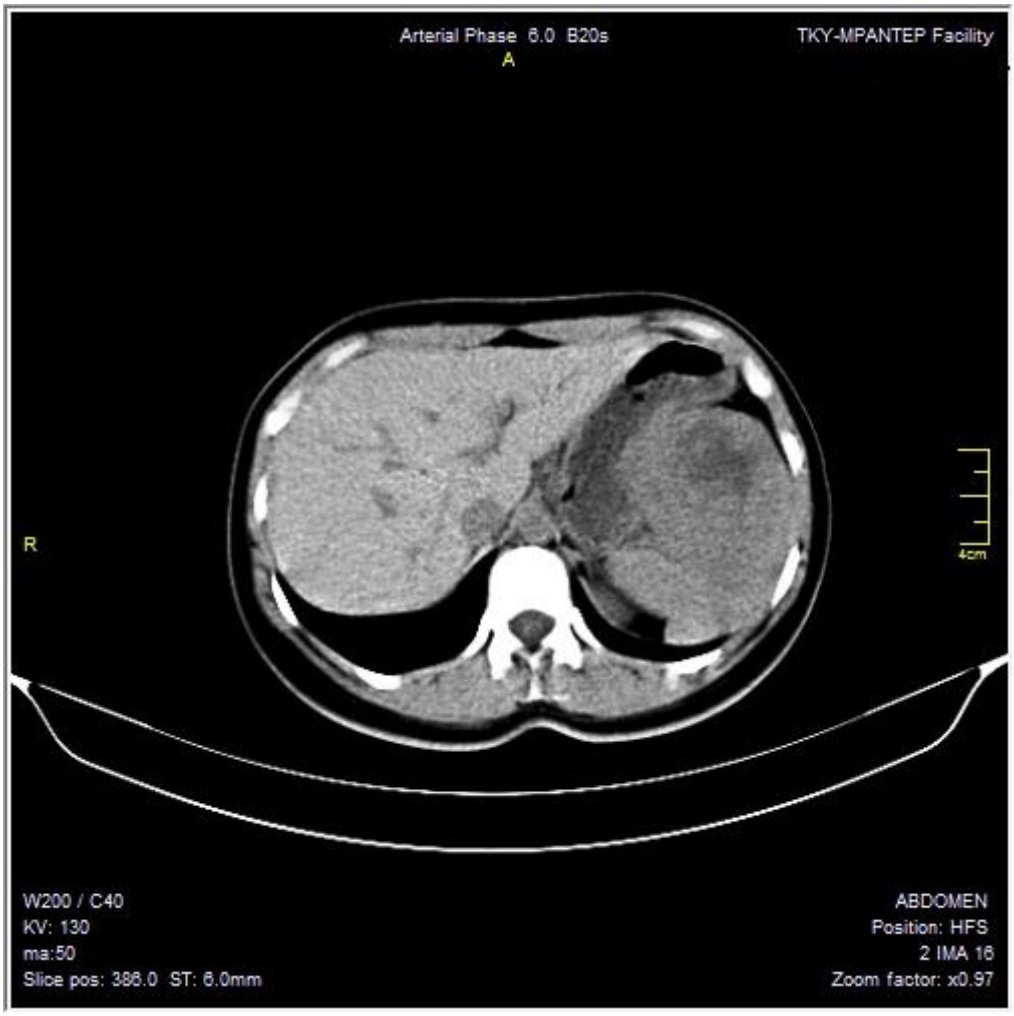

Figure 1: Abdominal CT shows a lobulated-contoured splenic mass including central cystic necrotic fields originating from upper half portion of the spleen extending to subdiaphragmatic and gastrosplenic areas.

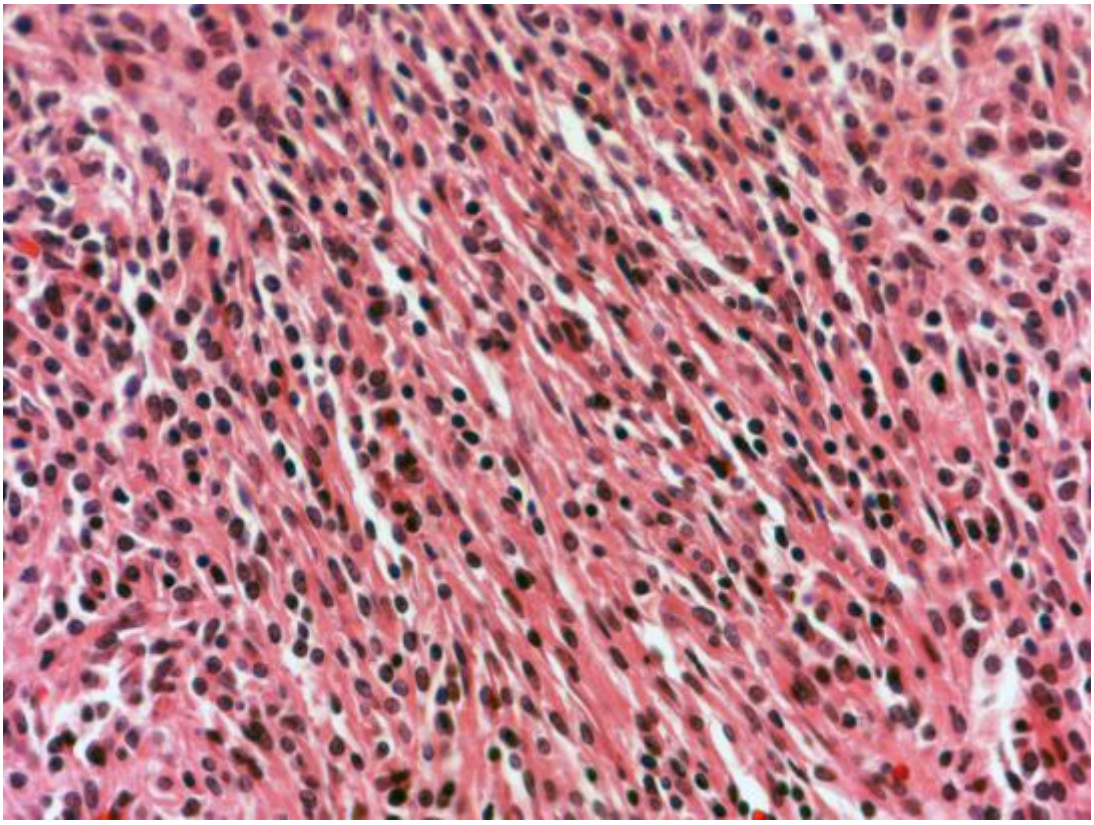

Figure 2: Atypical spindle and oval cells were seen (hematoxylin and eosin, original magnification x40).

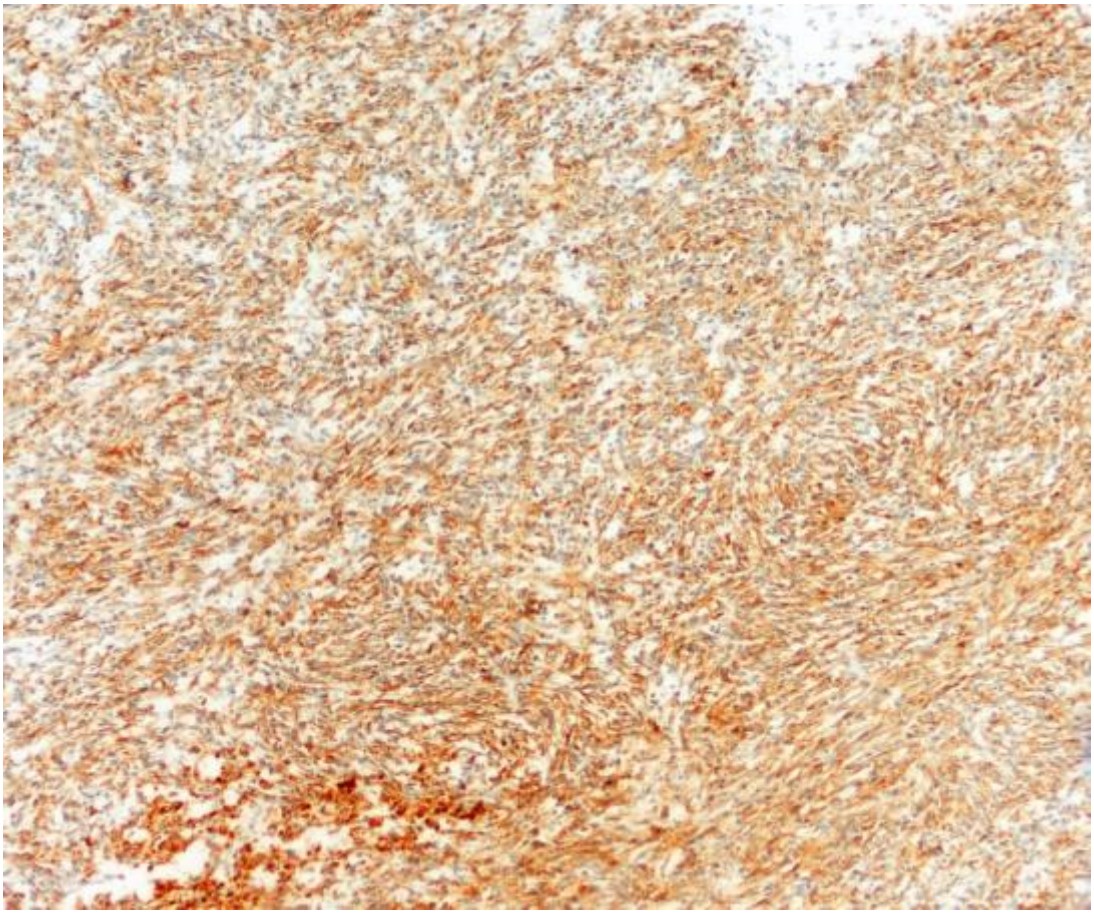

Figure 3: Weak and diffuse positive staining is observed immunohistochemical staining for CD68PGM1 (DAB, original magnification x100).

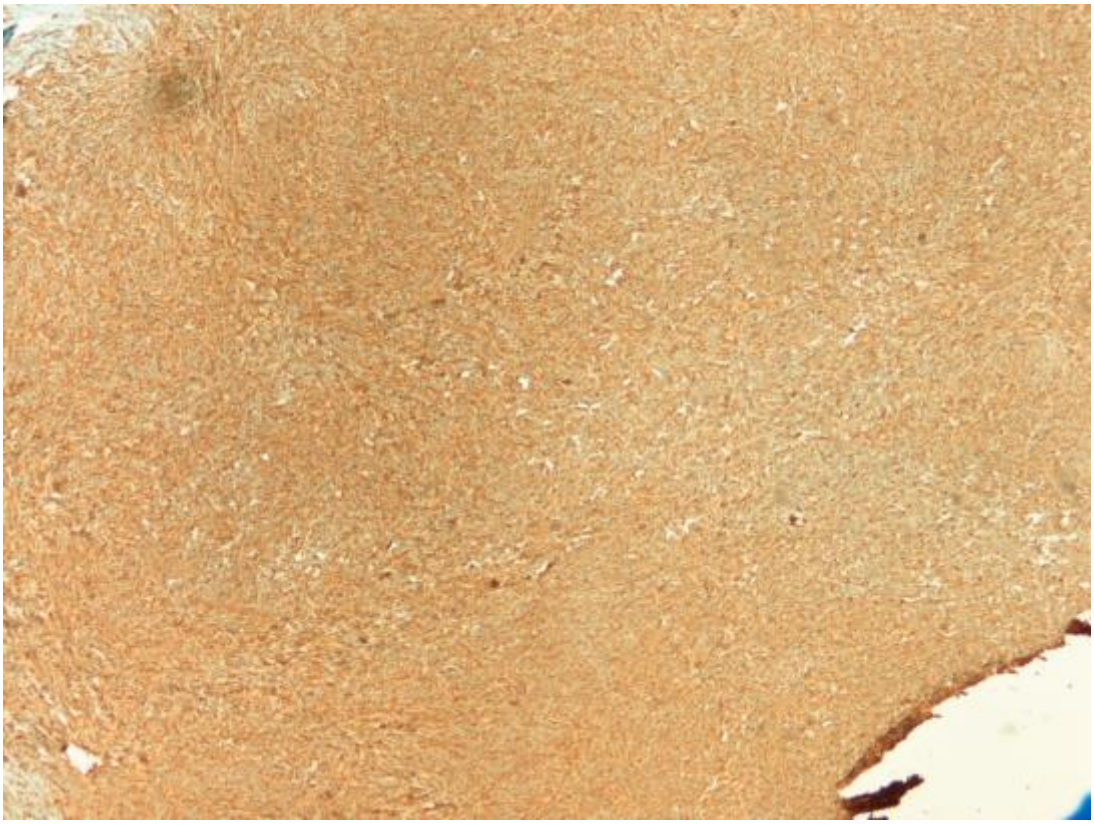

Figure 4: Immunohistochemical staining for vimentin is strongly positive (DAB, original magnification x100)

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