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Case Report

Follicular dendritic cell sarcoma: Rare presentation of incidental large hepatic mass

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Follicular dendritic cell sarcoma (FDCS) is a rare tumor, and it is even rarer for FDCS, to affect liver. FDCS is recognized as a distinct entity by the World Health Organization (WHO). Diagnosis is usually post resection and based on typical immunohistochemical stains. Resection with histologic negative margins is proposed, as a mainstay of therapy. We report a 63-year-old woman with large FDCS of the liver, managed with right hemihepatectomy. She remains disease-free at 48 months. **(Ann Hepatobiliary Pancreat Surg 2019;23:74-76)**

Key Words: Hepatectomy; Sarcoma

INTRODUCTION

Follicular dendritic cell sarcoma (FDCS) is a slow-growing low grade sarcomatous tumor, of mesenchymal dendritic cell origin.[1] It is a rare disorder, and accounts for 0.4% of all soft tissue sarcomas with peak incidence in the fifth decade of life, and equal gender predisposition.[2] Liver and spleen involvement are even rarer, with only a handful of case reports.[3] Due to slow growth and absence of pain, it is common for FDCS to present with large size, and symptoms due to size.[1,3]

CASE

A 63-year-old woman was presented to the hospital with fever and lethargy. She had no significant medical history. On examination, a firm non-tender mass extending 5 cm below the right costal margin was palpable. There were no stigmata of chronic liver disease. Routine hematology and serum biochemistry was unremarkable. Ultrasound of the abdomen showed a heterogeneous hypodense mass measuring 13.4×12.6×10.3 cm, in the right lobe of the liver with vascularity, and compressing the gall bladder. Hepatitis B and C viral serology were negative. Alpha-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen 19-9 were normal. Computerized tomography (CT) scan of the thorax-abdomen-pelvis confirmed sonography findings. Also, the mass was exophytic, centered at the gallbladder fossa, and had areas of hypervascularity and necrosis within it (Fig. 1A). The mass appeared to displace portal inflow medially (Fig. 1B). The gall bladder could not be delineated from the mass. A right hemihepatectomy was performed. Intra-operatively, the gallbladder was noted as displaced, but normal. The tumor was noted to have a pseudocapsule without invasion. Patient recovered uneventfully, and was discharged well, on post-operative day 6. On macroscopic examination, the resected specimen measured 13.4×13×11 cm and had a gross weight of 991 g. Cut surface of the tumor revealed a well circumscribed, firm, gray tan fleshy tumor with areas of hemorrhage and necrosis (Fig. 2).

On microscopic examination, the tumor was composed of spindle cells arranged in storiform pattern and whorls; as well as forming fascicles and sheets (Fig. 3A). There was dense interspersed lymphoplasmacytic infiltrate. Spindle cells showed oval to spindle vesicular nuclei, with prominent nucleoli. Large areas of coagulative necrosis were observed. Mitotic activity was 3/10 high power field.

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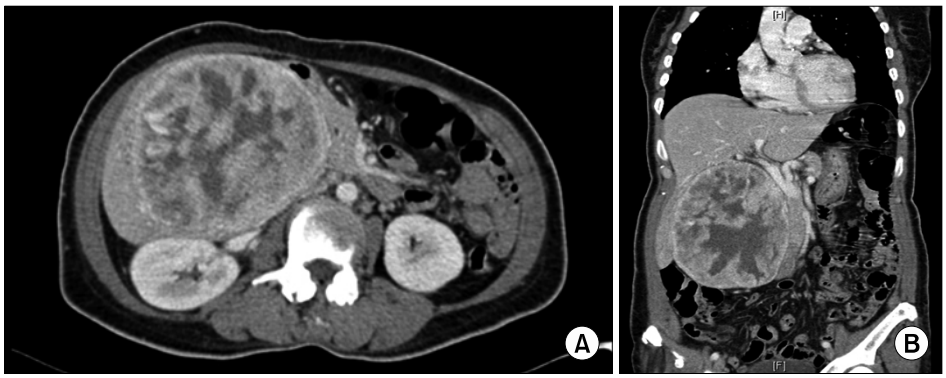

Fig. 1. Computed tomography scan of the hepatic mass: Axial (A) and coronal sections (B).

Fig. 2. Gross photograph showing the cut surface of resected tumour showing circumscribed fleshy tumor with focal necrosis.

Adjacent liver parenchyma did not show evidence of fibrosis or cirrhosis. Special stains for fungal (PASD & GMS) and mycobacterial (Ziehl Neelson) organisms are negative. On immunostaining, spindle cells show diffuse strong positive staining for CD21 (Fig. 3B). CD23 was also positive in large areas (Fig. 3C). CD3 highlights interspersed T lymphocytes and CD20 some scattered B lymphocytes without sheet like positive staining. ALK1, Desmin, Caldesmon, CD34, s100, DOG1 and Pancytokeratin AE1/3 were negative. SMA shows focal positive staining. Ki67 index was 20%.

In-situ hybridization (ISH) performed on a paraffin-embedded section reveals diffuse positive staining of tumor cells with an oligonucleotide probe specific for EBV encoded RNA (EBER) (Fig. 3D). In this setting of IMT (Inflammatory myofibroblastic tumor) like morphology in liver with positive follicular dendritic cell markers, diffuse and strong EBER (EBV encoded RNA) in situ hybridization positivity, indicates inflammatory pseudotumor like variant of follicular dendritic cell sarcoma (FDCS). Patient was further discussed at a cancer multidisciplinary meeting, and no adjuvant treatment was proposed. She remains well, and disease-free at 48 months.

DISCUSSION

Follicular dendritic cell sarcoma (FDCS) is an extremely rare tumor. It was first described by Monda et al.[4] in 1986. The majority of cases present as a painless slow-growing mass from cervical, mediastinal or axillary lymph node origin. The head and neck region are the more common sites for extra nodal presentation, especially in tonsils.[5] FDCS may also occur in the liver or spleen. Diagnoses of these are usually retrospective, after surgery has been performed.

In this case, a unique characteristic was presentation of an inflammatory pseudotumor (IPT) like variant of FDCS, of which only a handful of cases has been reported.[3] This variant is almost exclusively found within the liver and spleen, and has been recognized as a distinct entity by the World Health Organization (WHO).[6] They are related to the Epstein - Barr virus (EBV) proliferation. FDCS present equally in both genders and in young adulthood (40-50 years).[2] IPT like variant of FDCS has female predilection (2.2:1).[3] IPT is a benign lesion of a solid organ with inflammatory pathology, while IPT like variant of FDCS is a sarcomatous tumor, and is a malignant disease. It is speculated that involvement of EBV or hyaline vascular type Castleman's Disease (HVCD), can increase risk.

Diagnosis of FDCS is based on very distinct, immunophenotypical and histological findings. Tumors may exhibit spindle-shaped cells in a storiform arrangement, with indistinct cell borders and a background of lympho-

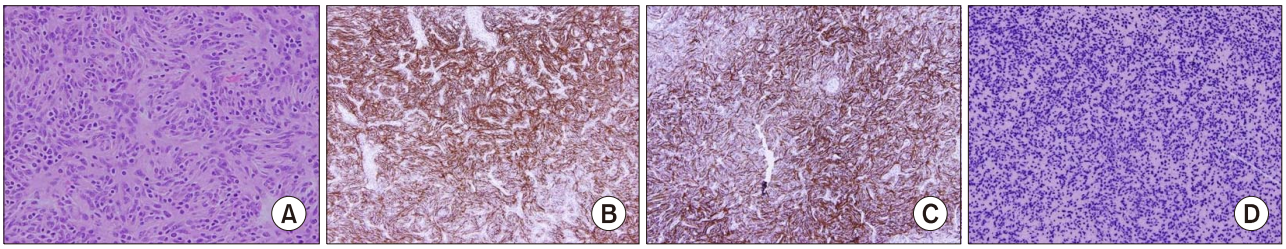

Fig. 3. Microscopic findings. (A) Spindle cells in storiform pattern and whorls with dense interspersed lymphoplasmacytic infiltrate (H&E ×400). (B) Diffuse and strong positive immunohistochemical staining for CD21. (C) Large areas of positive immunohistochemical staining for CD23. (D) Positive immunohistochemical staining for Epstein-Barr virus-encoded RNA.

cytes.[1,7] Immunohistochemical studies with FDC markers show strongly positive for CD21, CD35, Ki-MP4, FDRC1P, clustering.[1,7] Tumors may also commonly test positive for CD23, desmoplakin, NGFR, HLADR, vimentin and fascin. Immunohistochemistry staining pattern is essential and enough to conclude diagnosis of FDCS in patients with histological features of spindle-cell tumor.

A large retrospective review suggested curative surgery as standard treatment, with no adjuvant chemo radiation. Post-operative radiotherapy is currently advocated, for lesions incompletely resected.[8] Some reports also claim on possible metastatic potential of these tumors.[7] Our patient presented with a large palpable mass in the right upper abdomen, and did not show evidence of metastatic disease. Hence, surgical resection was planned. Pre-operatively we did not suspect diagnosis of FDCS, and we do not advocate liver biopsy for tissue diagnosis, as it doesn't alter the management plan i.e. surgical resection; and also poses risk of bleeding and tumor seeding along the biopsy track. FDCS is a rare case of solid liver lesion, and diagnosis is usually after resection. In patients with histological negative margins, surveillance is adequate. However, it is essential to be vigilant about FDCS as a distinct entity, due risk of recurrence, or metastases. FDCS are shown to be hypermetabolic and observed as avid spots on positron emission tomography scan.[9] Rarity of FDCS to affect extranodal solid organs, serves as an obligation to report experience of single cases, as this adds to the understanding of natural history and behavior of these rare neoplasms.

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