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Case study

Follicular dendritic cell sarcoma with immature T-cell proliferation ☆

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Summary Follicular dendritic cell sarcoma is characterized by proliferation of spindled to ovoid cells reminiscent of follicular dendritic cells. However, the association of follicular dendritic cell sarcoma with a dense infiltration of immature T cells has not hitherto been reported. We report an unusual case of follicular dendritic cell sarcoma of the mesentery with immature T-cell proliferation in a 68-year-old man. The infiltrating immature T cells demonstrated expression of CD3, CD1a, TdT, and coexpression of CD4 and CD8 by immunohistochemistry. In addition, the patient was subsequently diagnosed with myasthenia gravis and paraneoplastic pemphigus and died of distant metastasis within 2 years after initial diagnosis of follicular dendritic cell sarcoma. The aggressive clinical course of this case contrasts with the indolent course of follicular dendritic cell sarcomas, and thus, the prognostic implications of follicular dendritic cell sarcoma with immature T-cell proliferation require clarification. The complication of myasthenia gravis and paraneoplastic pemphigus may suggest that immature T-cell proliferation has an autoimmunity-related systemic influence.

1. Introduction

Follicular dendritic cell (FDC) sarcoma is a rare tumor characterized by proliferation of spindled to ovoid cells recapitulating the morphologic and immunophenotypic features of follicular dendritic cells [1,2]. The neoplastic cells form sheets, fascicles, storiform patterns, and sometimes whorls, and the tumor is usually lightly infiltrated by small lymphocytes, although more prominent lymphoplasmacytic infiltration is seen in the less common inflammatory pseudotumor-like variant [1,3]. However, the association of FDC sarcoma with a dense infiltration of immature T cells has not been previously reported in the literature.

Here, we report an unusual case of FDC sarcoma of the mesentery, which demonstrated large numbers of immature T cells. Interestingly, this case was also associated with myasthenia gravis (MG) and paraneoplastic pemphigus (PNP) and pursued an aggressive course.

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2. Case report

A 68-year-old man presented with a palpable mass of the midabdomen. Computed tomography (CT) revealed a mass of probable small bowel mesentery origin (Fig. 1A). Chest CT showed no abnormal findings. The mass was completely excised by explorative laparotomy without complications. Grossly, a 9-cm-sized, well-circumscribed lobulated mass was found in the small bowel mesentery (Fig. 1B). The mass had a grayish white cut surface and a firm fleshy texture. Microscopically, a diffuse and vaguely fascicular proliferation of spindled to ovoid cells with mild to moderate nuclear atypia was observed, and notably, numerous small lymphoid cells were scattered throughout the tumor (Fig. 2A, B). The spindled to ovoid cells had eosinophilic cytoplasm with indistinct cytoplasmic borders, large, vesicular nuclei, and centrally located small but prominent nucleoli. Mitoses were frequently encountered at up to 5/10 high-power fields. An infiltrative growth into adjacent mesenteric fat tissue and multifocal necrosis was also observed. Most infiltrating small lymphoid cells had nuclei with dark chromatin, indistinct nucleoli, and scant cytoplasm. They exhibited the so-called starry-sky pattern with occasional tingible body macrophages (Fig. 2A). Some plasma cells and a few immunoblastic cells were also present. Immunohistochemical studies were performed using antibodies listed in Table 1. The tumor cells were positive for CD21, CD35, D2-40, fascin, epidermal growth factor receptor, and vimentin, and were negative for all other markers (Fig. 2C, D). Ultrastructural examination revealed shallow or deep cytoplasmic indentations of tumor cell nuclei (Fig. 2E, inset). The infiltrating lymphoid cells within the tumor were mainly $CD3^+$ T cells (>80%) with multifocal small aggregates of $CD20^+$ B lymphocytes. Unexpectedly, the $CD3^+$ T cells were also positive for TdT, CD1a, and βF1 and showed dual CD4 and CD8 expression, characteristic of immature T cells (Fig. 3A-E). The Ki-67 labeling index of these cells reached approximately 80% (Fig. 3F). A few scattered cells showed equivocal positivity for CD30. In situ hybridization using EBER1/2 oligonucleotide probes (Novocastra, Newcastle, UK) revealed no evidence of Epstein-Barr virus infection in both tumor cells and

Table 1 Antibodies used in this case

Antibody	Source	Antibody	Source
CD1a	Novocastra (Newcastle, UK)	CD68	Dako
CD3	Dako (Copenhagen, Denmark)	D2-40	Dako
CD4	Thermo Scientific (Freemont, CA, USA)	Pancytokeratin	Dako
CD8	Neomarkers (Freemont, CA, USA)	Vimentin	Dako
CD15	Dako	S-100 protein	Dako
CD20	Dako	Smooth muscle actin	Dako
CD21	Dako	Ki-67	Dako
CD30	Dako	Clusterin	Millipore (Bedford, MA, USA)
CD35	Novocastra	Fascin	Dako
β F1			Endogen (Woburn, MA, USA)
Terminal deoxynucleotide transferase (TdT)			Dako
Anaplastic large-cell lymphoma kinase (ALK)			Dako
Epithelial membrane antigen (EMA)			Dako
Epidermal growth factor receptor (EGFR)			Zymed (San Francisco, CA, USA)

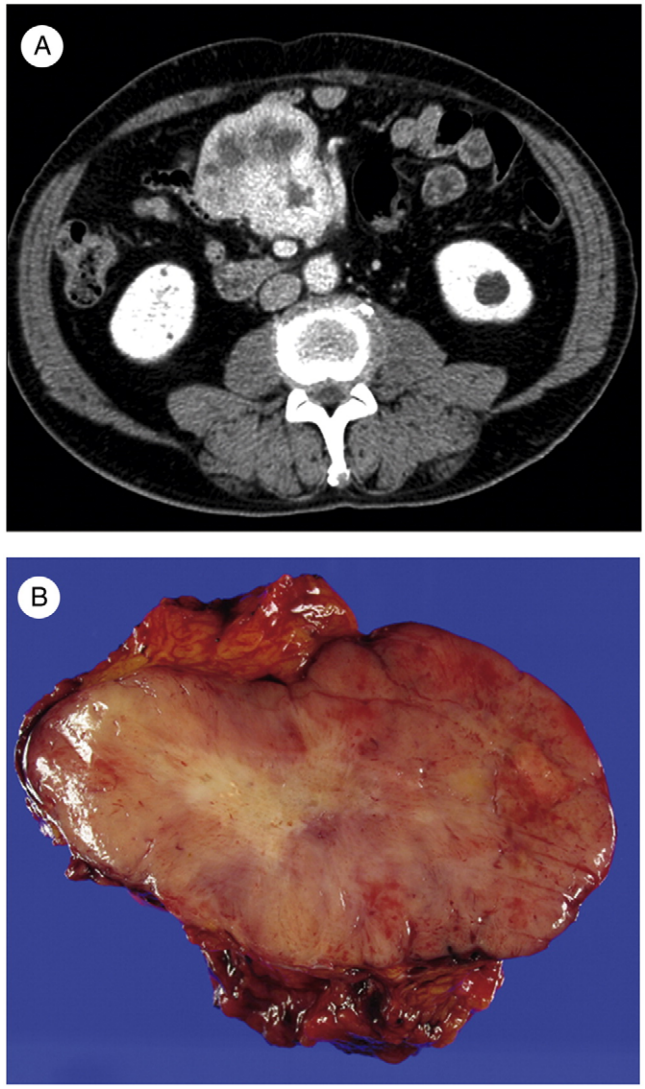

Fig. 1 Radiologic and gross features of the tumor. A, An abdominal CT scan showing an irregular peripherally enhancing mass of mesenteric origin. B, The mass was 9-cm-sized, firm, grayish white, and well circumscribed.