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Original Article

Clinicopathologic profile of extranodal follicular dendritic cell sarcoma in the mesentery of small intestine: a study of two cases with literature review

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Abstract: Aim: To analyze the clinicopathologic profile of the extranodal follicular dendritic cell sarcoma (FDCS) in the mesentery of small intestine. Materials and methods: The clinical observations and histopathologic andimmunohistochemical features of FDCS were analyzed in the mesentery of small intestine. Results: FDCS had no characteristic clinical manifestations. Histology showed oval sarcoma cells forming fascicles of spindle cells and whorls. Sarcoma tissue was distributed pervasively and often mixed with some T lymphoid cells. Immunohistochemical markers showed the follicular dendritic sarcoma cells were positive for CD21, CD23, CD35, D2-40 and vimentin, and weakly positive for CD68, and S-100, while EMA, CD1a, desmin, AE1/AE3, CD45RO, CD3, LCA and MPO were negative. Conclusion: FDCS is a rare malignant tumor in the mesentery of small intestine.I Its diagnosis depends on the histopathology and immunohistochemical staining, and its prognosis is uncertain. Surgical operation is the first choice of treatment.

Keywords: Mesentery of small intestine, follicular dendritic cell sarcoma, immunohistochemistry, diagnosis

Introduction

Follicular dendritic cell sarcoma (FDCS) originates from follicular dendritic cells (FDC), which are accessory immune cells of the lymphoid system. The first case report of FDCS in tonsil was described in 1986 by Monda et al. [1]. Since then, FDCS has been reported increasingly. FDCS occurs most frequently in the lymph nodes of neck and axillary [2]. However, FDCS in extranodal sites is widespread throughout the body, presenting in urinary bladder [3], tonsil, palate [4], pancreas [5], pharynx, peritoneum, gastrointestinal tract (GIT) [6], spleen [7], mediastinum [8], thyroid, lung, oral cavity, and liver. Few articles about FDCS In the mesentery of small intestineare reported in the English literature [9, 10].

Therefore, an erroneous diagnosis is easily made. In order to facilitate the diagnosis and understanding of this rare cancer, we describe two cases of FDCS in the mesentery of small intestine, with clinicopathologic profile. In addition, we review the literature on FDCS.

Materials and methods

In our report, two cases diagnosed as FDCS in mesentery of small intestine were obtained from Department of Pathology, Tianjin Nankai Hospital. The detailed clinical data was gathered from the hospital records.

Affinity-purified mouse monoclonal antibodies for CD21, CD23, CD35, D2-40, Vimentin, CD68, EMA, CDla, desmin, AE1/AE3, CD45RO, CD3, LCA, MPO and S-100 were purchased from Beijing Zhongshan Golden Bridge Biotechnology Co. LTD. EliVisionTMplus Polyr HRP (Mouse/Rabbit) IHC kit (KIT-9901) were purchased from MAIXIN-BIO.

Pathologic materials were fixed in 10% buffered formalin and routinely processed for light microscopy. 4-μm-thick sections were cut from the tissue blocks and stained with hematoxylin and eosin (H&E). The avidin-biotin-peroxidase complex (ABC) method was used for immunohistochemical staining studies. In order to evaluate the specificity of the antibodies, serial sec-

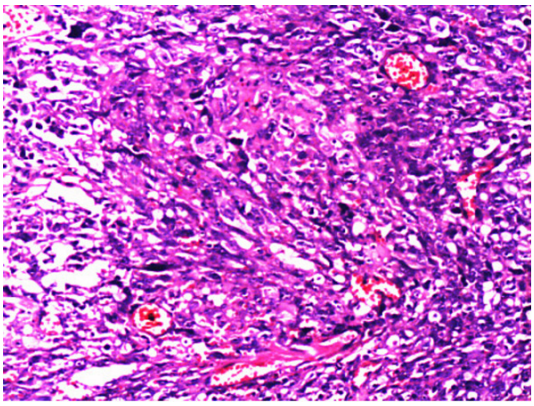

Figure 1. The tumor cells are composed of oval to spindle cells growing in fascicles, and whorls. A multinucleated giant cell can be seen. HE×100.

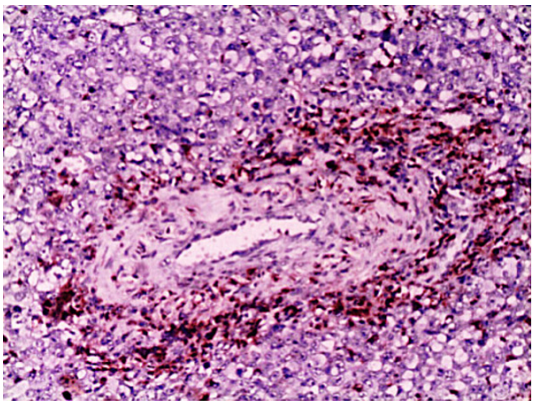

Figure 2. T lymphocytes around a vessel confirmed by CD3 SP×100.

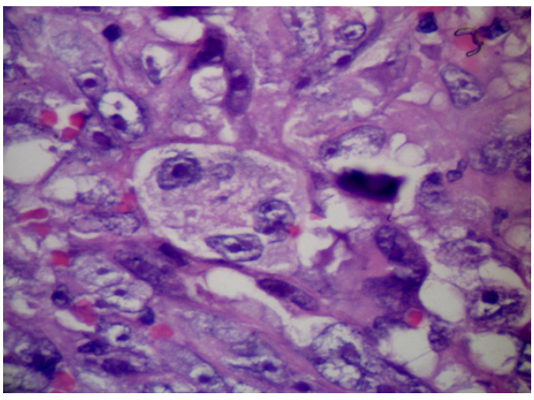

Figure 3. Multinucleated giant cell, HE×400.

tions were incubated with PBS displacing the primary antibody as negative controls, and known positive tissues were used as the positive controls.

Results

Clinical data

Case 1: A 74-year-old man presented at our hospital with a burst of epgastric pain after drinking. Ultrasound indicated that there were large amount of gas in gastrointestinal area, and suspicion of small intestine perforation. No obvious abnormity was found by laboratory examination. A perforation of 3 cm in diameter was found in the small intestine during emergency surgery, besides which a mass was seen in the mesentery, measuring 3.3×2.2 cm in diameter. The small intestine structure was almost completely destroyed by tumor cells. Consistent short fusiform to oval cells were found in the submucosa, with to moderate atypia and eosinophilic cytoplasm, arranged in bundle and storiform patterns (**Figure 1**). There were small amounts of lymphocytes scattered throughout the tumor cells and clustered around blood vessels, which were T cells as demonstrated by CD3 (**Figure 2**). Large geographic necrosis areas were seen. At high magnification, the tumor cells were spindle shaped with prominent eosinophilic cytoplasm. The nuclei were round or oval. A few mononuclear tumor giant cell or syncytial cells were seen (**Figure 3**). Mitotic rate was 10-13/10 HPF. Immunohistochemical analysis demonstrated that the tumor cells were positive with CD21, CD23, CD35, clusterin, vimentin, and D2-40, while focally positive for CD68, and S-100 (**Figure 4**). The patient received no other treatment except for surgery; he died of other disease one year after the diagnosis.

Case 2: Patient 2 was a 34-year-old woman. She complained about an abdominal mass during a physical examination a months before. She had no fever, cough, decompensation, or expectoration. Surgical examination confirmed her finding. The mass in the small intestine mesentery was found and removed surgically. The neoplasm measured 9×7×6 cm. The mass was excised and sectioned for histologic examination. The oval and fusiform tumor cells were slightly atypia with eosinophilic cytoplasm, and large nuclei with obvious nucleoli and mitoses (**Figure 5**). A mixture of lymphocytes and neutrophils patchily infiltrated among neoplastic cells. Immunohistochemical analysis showed that the tumor cells were positive with

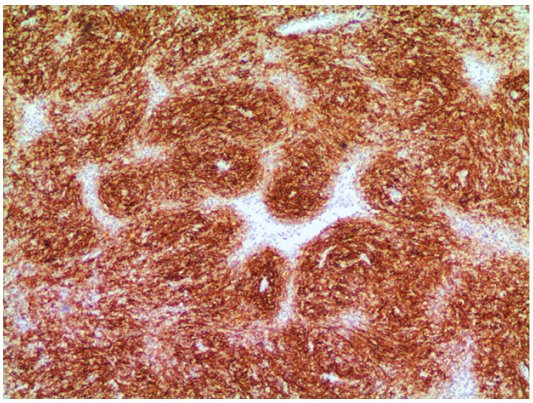

Figure 4. Neoplastic cells are immunoreactive for CD21. SP×100.

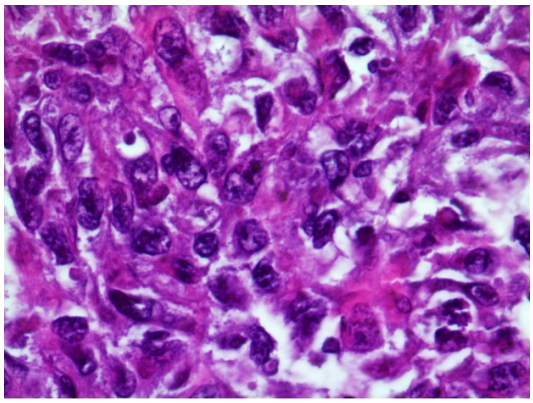

Figure 5. At high magnification, the tumor cells are round with prominent eosinophilic cytoplasm. HE×400.

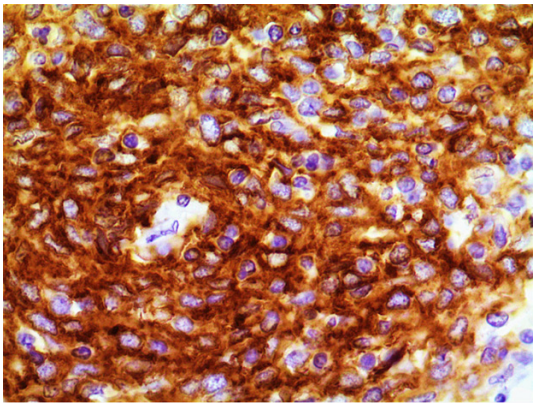

Figure 6. Neoplastic cells are immunoreactive for CD23 SP×200.

CD21, CD23, CD35, vimentin, and D2-40, and focally positive for CD68, and S-100 protein (**Figure 6**). Patient received radiotherapy after the operation for 3 months, and there was no evidence of recurrence for 8 years' follow up.

Criteria

Tumor cell membranes are positive for CD21, CD23, CD35, D2-40, and cytoplasm is positive for CD68, S-100 protein, and vimentin.

Discussion

FDCS is a very rare neoplasm, and its identity-remained unknown until recently. Therefore, perhaps there are more FDCS cases than those that have been reported. Although FDCS can occur at any age, it usually affects young adults [11]. A review of the literature showed a slight female predominance [12]. But another article indicated that there was no gender difference, in agreement with us [13]. Clinical presentation of intra-abdominal FDCS included abdominal pain, diarrhea, intestinal obstruction, melena, and dyspepsia [14]. However, none of our cases had the above symptoms. FDCS is traditionally regarded as an indolent tumor, with a tendency to local recurrence and little risk of distant metastasis. However, Chan et al. considered that high mitosis, nuclear pleomorphism, and lack of adjuvant therapy were predictors of higher recurrence rate [14].

The microscopic appearance of FDCS showed that the tumor cells were composed of oval to spindle cells in fascicles, whorls, sheets, and a storiform pattern, just as in our cases. The neoplastic cells were intimately admixed with small T lymphocytes as we described above. Necrotic areas usually can be seen. Yun-Chen Chang et al. considered that nuclear pleomorphism, brisk mitotic rate, coagulative necrosis, and atypical forms were aggressive featuresthat correlated with poor outcome [9].

FDCS are mesenchymal tumors. All the tumors in this series have similar morphologic features. It is often difficult in histology to differentiate them from inflammatory myofibroblastic tumour, smooth muscle, and fibrohistiocytic tumors, metastatic sarcoma, and undifferentiated carcinoma. Hence, Immunohistochemistry (IHC) is necessary for diagnosis of FDCS [15]. FDCS has distinct immunohistochemical features. The tumor cells are commonly positive for CD21, CD23, CD35, which aremarkers of normal follicular dendritic cells. Thus their diagnoses rely on these 3 markers in almost all reported cases. Few cases lack both CD21 and CD35 immunoreactivity. Xie et al. showed that D2-40 was superior or equal to CD21 for evaluating

the disrupted reactive FDC meshwork in a variety of lymphocytic disorders, and D2-40 was a sensitive and specific new marker for diagnosis of FDCS [16]. Some studies reported FDCS expressed FDCSP and SRGN, which probably can serve as novel follicular dendritic cell sarcoma markers [17]. The positive expression of D2-40, S-100 protein, CD68, vimentin and Ki-67 may be nonspecific. Our cases had the same immunohistochemical findings. Therefore, it is important to use a sufficient number of markers in the diagnosis of FDCS.

Surgical resection is considered to be an effective clinical treatment for FDCS. The survival rate of surgical resection for non-metastatic cases is significantly higher than non-operative therapy. The therapeutic effect of adjuvant chemotherapy and radiotherapy is unclear due to its rarity. Thus, FDCS treatment should be adjusted according to the specific invasion of the tumor, age of patient, location, size, mitotic count, presence of significant necrosis and cellular atypia of the tumor. Some articles reported that the chemotherapy regimens and radiotherapies commonly used for lymphomas or soft tissue sarcomas were suitable for some non-operative patients, but their roles remained unclear.

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Disclosure of conflict of interest

None.

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