Abstract. Follicular dendritic cell sarcoma (FDGS) is an extremely rare tumor, which mainly originates from FDCs in the lymph nodes. Sometimes FDGS can arise from outside the lymph nodes due to the existence of acquired lymphoid tissue, which becomes the histological basis of the tumor. The diagnosis of FDGS, particularly extranodal FDGS, presents a challenge for pathologists and hematopathologists. The present study presents two cases of extranodal FDGS based on clinical features and histomorphology. Soft tissue of the chest wall was involved in case 1 and right tonsil tissue in case 2. Case 1 underwent surgery, and was in good health post-operatively. During the 5-month post-operative follow-up period, the patient was healthy in all respects. Case 2 received surgery combined with radiotherapy, and the follow-up data reported that the patient remained alive, without signs of recurrence or metastasis during the 4-month post-operative follow-up period. Additionally, a total of 102 cases of extranodal FDGS were retrieved from the literature, which were extracted and reviewed carefully. The rates of recurrence, metastasis and mortality were 14.63 (12/82), 17.07 (14/82) and 8.29% (15/82), respectively. The overall survival rates of the 102 cases, showing 2-year total survival rates, were 70%, the same with that of 5-year total survival rates. The 2-year tumor-free total survival rates were 68%, and the 5-year equivalents were 32%. Female patients had a poorer prognosis than male patients (P<0.05). Kaplan-Meier estimation presented no statistically significant differences between disease-free survival rates or overall survival rates and age, tumor size or treatment (P>0.05).

Introduction

Histiocytic neoplasms are a type of tumor that arise from the monocyte-macrophage system, including dendritic cells (DCs), macrophages and sometimes histiocytes (1,2). DC tumors are associated with certain types of antigen-presenting cells (2). Follicular DC sarcoma (FDGS) is extremely rare, so its epidemiology is unclear. There is a wide patient age range, with an adult predominance (median patient age, 50 years) and an almost equal sex distribution (2,3).

FDGS consists of a neoplastic proliferation of spindled to ovoid cells exhibiting morphological and immunophenotypic features of FDCs (1). Misdiagnosis occurs frequently, due to the fact that it has the same micromorphology as other common neoplasms, such as other types of sarcoma and lymphoma (2,3). In ~35% of cases, FDGS presents in the lymph nodes (commonly in cervical nodes), while ~65% of cases present with extranodal disease (1,4,5). A number of extranodal sites can be affected, such as tonsils, gastrointestinal tract, soft tissue, mediastinum, omentum and lungs (2,3). Surgical excision is the main treatment, sometimes combined with adjuvant radiotherapy or chemotherapy (3). The 2-year survival rates for early, locally advanced and distant metastatic diseases are 82, 80 and 42%, respectively (1,2). Some patients may die from refractory paraneoplastic pemphigus (2). Inflammatory pseudotumour-like FDGS or fibroblastic DCs often affect the liver or spleen in female patients (1,2). Histologically, the neoplastic-spindled cells are dispersed within a prominent lymphoplasmacytic infiltrate (1). In the present study, two cases of extranodal FDGS were reported, which affected the tonsils and soft tissue of the chest wall were collected. Additionally, an analysis of 102 cases of FDGS from the literature was performed to further explore the biological behavior of extranodal FDGS.

Case report

Samples. The present study retrospectively reviewed all cases of extranodal FDGS in the past 5 years, including excision surgery and biopsy, a total of two cases were collected from the pathological archives of the Affiliated Hospital of Chengde Medical College (Chengde, China) between January 2013 and January 2020. Two experienced pathologists reviewed all the
slides of the two cases independently, identifying the diagnosis. Case 1 was further reviewed and confirmed by the pathologists of the Department of Pathology of the China-Japan Friendship Hospital (Beijing, China). The case history was consulted to collect the corresponding clinical data. Additionally, two experienced radiologists were asked to confirm the preoperative imaging test. The two patients were recommended routine re-testing monthly and were followed up until now (followed for 4 and 5 months, respectively), and all the information was recorded.

**Histological examination.** Tissue specimens were collected for 10% buffered formalin fixation, grossing, routine dehydration, embedding into paraffin and sectioning into 4-µm-thick sections for hematoxylin and eosin staining (10% buffered formalin for 12 h at room temperature; hematoxylin for 5-15 min at room temperature; water for 1-3sec at room temperature; and eosin for 1-3 min, at room temperature). Microscopic and immunohistochemical phenotypes were observed to ensure the accuracy of the diagnosis, according to the diagnostic criteria of the 2017 World Health Organization (WHO) Classification of Tumors of Haematopoietic and Lymphoid Tissues (5).

**Immunohistochemistry (IHC).** IHC was performed on paraffin blocks, using the Leica automatic immunostaining device (Leica Microsystems, Inc.). The 4-µm-thick sections were fixed using 10% buffered formalin for 12 h at room temperature. All monoclonal antibodies used were purchased from OriGene Technologies, Inc., and are listed in Table I. The known positive tissue was used as the positive control, and PBS as the negative control. The scoring method based on both the intensity (0, no staining; 1, weak staining; 2, medium staining; and 3, strong staining) and proportion of positive cells (0, 0%; 1, 1-25%; 2, 26-75%; and 3, 76-100%). The final staining scores were calculated by multiplying the staining intensity score by the extent of staining score. A final staining score of ≥3 was considered positive, and others were classified as negative.

**Case description**

**Case 1.** A 63-year-old male patient visited the doctor presenting with a chest wall neoplasm without any complications in September 2019. He had no weight loss, pain, fever or other feeling of discomfort. Physical examination revealed a mass lesion measuring 5x4 cm under the skin of the chest wall. No palpable lymphadenopathy was found. The patient was suggested to undergo surgery to resect the mass. Grossly, the resected tissue with skin measured 6x5x1.5 cm. A white nodule measuring ~4.5x4 cm was found, with a complete capsule and firm quality. Some gray regions were observed on the cut surfaces, without necrosis and hemorrhage. Microscopically, the neoplasm was composed of ovoid to spindle-shaped tumor cells, arranged into whorls arrays. Collagen fiber and multinucleated cells were observed in the background, with areas showing clustering. Parts of the tumor were accompanied with hemosiderin. Necrosis and hemorrhage were not observed. Mitotic figures were rare (Ki-67 immunoproliferative index of ~10%; Fig. 1D). A total of 500-2,000 tumor cells were counted manually to evaluate the Ki-67 immunoproliferative index. The tumor was lightly infiltrated by small lymphocytes with lymphoid follicle formation (Fig. 1A).

IHC revealed that the tumor cells were positive for D2-40 (Fig. 1G), Vimentin, CD21, CD23, CD117, EGFR, ALK and HMB45, and negative for cytokeratin (CK), actin smooth muscle (SMA), Actin, P53, Desmin, MelanA, CD1a and CD34 (Fig. 1D and G).

The diagnosis of FDCS was made based on the morphological and immunological characteristics. In view of its rarity, the patient was suggested to further consult the pathologists of the Department of Pathology of the China-Japan Friendship Hospital, and the diagnosis was determined again. The postoperative recovery of the patient was rapid without complications. During the 5-month post-operative follow-up period, no recurrence, metastasis or other signs of discomfort were observed.

**Case 2.** A 70-year old male patient checked into the Oncology Department of the Affiliated Hospital of Chendge Medical College, with an abnormal sensation in the throat for 3 months. The patient had no dysphagia, fever or dyspnea. Upon ultrasound examination, a suspicious mass was revealed at the right tonsil. The cervical lymph node was not enlarged. A right tonsillectomy was performed under general anesthesia. Grossly, fragmentary tissue measuring 2x2x0.5 cm was observed. Microscopically, the biggest part of the tonsil was covered by mature squamous cells, while an ulcer was found on the surface. Under the squamous cells, an ovoid to round-shaped tumor formed solid or nested patterns. Some tumor cells exhibited prolonged nuclei and obvious nucleoli, as well as an eosinophilic cytoplasm. Significant cytological atypia, focal coagulative necrosis and hemorrhage were identified. Lymphocytes, eosinophils and plasmocytes were also observed under light microscopy (Fig. 1B and C). Immunohistochemically, the tumor cells were positive for CD21, epithelial membrane antigen (EMA) and CD35 (Fig. 1E, F and H, respectively), weakly positive for CD68, and negative for CD20, CD3, CD30, S-100, Lyso, CD1a, ALK and leukocyte common antigen (LCA). The patient underwent postoperative radiotherapy as well. During the 4-month follow-up period after undergoing tonsillectomy, no local recurrence or distant metastasis was observed.

**Literature review**

**Literature review.** The literature was reviewed by searching the key words ‘extranodal follicular dendritic cell’ on PubMed (www.ncbi.nlm.nih.gov/pubmed), and the Chinese literature was searched on the China National Knowledge Infrastructure (http://www.cnki.net/) between 2008 and 2019. All the cases of FDCS, except those in lymph nodes, were collected. As much of the literature as possible was consulted to ensure each case was reported only once and discern the renewed information about the cases.

**Statistical analysis.** SPSS 17.0 (SPSS, Inc.) was used for statistical analysis. The Life Tables method was used to calculate the overall and tumor-free survival rates. The association between clinicopathological parameters and prognosis were calculated using Kaplan-Meier estimation and the log-rank test. P<0.05 was considered to indicate a statistically significant difference.
Clinical features of extranodal FDCS. A total of 102 cases of extranodal FDCS were retrieved from the literature (Table II) (1-55). The clinicopathological features, management and clinical outcomes were extracted and recorded. Of the 102 cases, 55 were male and 47 female (male to female ratio, 1.17:1) and the age range was 16-79 years at the time of diagnosis (mean age, 48.87 years; median age, 47.5 years). The tumors were located in different regions, including the abdominal cavity (n=46), tonsil (n=20), oropharynx (n=15), mediastinum (n=12), neck soft tissue (n=2), pelvic cavity (n=2), bone (n=1), lower limbs (n=1), intracranium (n=1), esophagus (n=1) and thyroid (n=1). The tumor longitudinal diameter measured 1-20 cm, with a mean diameter of 6.63 cm, an abdominal cavity diameter of 8.33 cm and a diameter of the outside of the abdominal cavity of 4.74 cm. A total of 8 cases were coexistent with other diseases, including 1 with Castleman disease, 3 with paraneoplastic pemphigus, 2 with chronic hepatitis B virus infection and 2 with carcinoma.

Pathological features of extranodal FDCS. Most of the cases from the literature were well circumscribed, with grey white-red cutting surfaces. Focal coagulative necrosis (n=20) and hemorrhage (n=14) were observed in some cases. Mitotic figures were rarely observed [range, rare-30/high power field (HPF)]. Generally, extranodal FDCS is positive for specific markers for DCs, such as CD23, D2-40, CD35 and CD21. One case of urinary bladder FDCS was negative for BRAF. EBV-encoded RNA (EBER) was detected using in situ hybridization (ISH) in 53 cases and it was positive in 20 and negative in 23 cases, most located in the liver and spleen. A total of 19 cases were misdiagnosed to other tumors or inflammatory lesions (Table II).

Management and clinical outcomes of extranodal FDCS. The data of the management of the extranodal FDCS cases were available for 95 cases. A total of 50 cases underwent surgery to resect the neoplasm. A total of 1 case received...
chemotherapy procedures, while 44 cases came to the clinic for adjuvant treatment (17 cases for radiation treatment, 16 for chemotherapy and 11 for both). Complete follow-up information was accessible in 82 cases. The follow-up duration was 1-108 months, with an average of 24.31±22.98 months (Table II). The 2- and 5-year total survival rates were 70 and 70%, respectively. The 2- and 5-year tumor-free total survival rates were 68 and 32%, respectively (Table III and Fig. 2). The rates of recurrence, metastasis and mortality were 14.63 (12/82), 17.07 (14/82) and 18.29% (15/82), respectively (Table III). The median survival time was 108 months.

Prognostic factors of extranodal FDCS. The Kaplan-Meier method was used to analyze the association between clinico-pathological features and prognosis (Table III and Figs. 3-6). Upon this analysis, it was found that sex was associated with disease-free survival (P=0.032; Table III), with female patients having a poorer prognosis than male patients.
Table II. Clinical characteristics of 102 patients with extranodal FDCS from the literature.

| Case no. | Age, years | Sex | Size, cm | Mitotic Counts (/10 HPF) | Site | Necrosis | Initial diagnosis | EBER | Treatment | Follow-up, months | Recurrence/metastasis, months | Status (Refs.) |
|----------|------------|-----|----------|--------------------------|------|----------|------------------|------|------------|------------------|---------------------------|---------------|
| 1        | 62         | M   | NA       | NA                       | Mediastinal | Yes       | FDCS              | NA   | Surg+ChT   | 24               | No                        | STD (1)       |
| 2        | 46         | M   | NA       | NA                       | Mediastinal | Yes       | FDCS              | NA   | Surg+RT    | 12               | No                        | NED (1)       |
| 3        | 31         | M   | NA       | NA                       | Mediastina  | No        | FDCS              | NA   | Surg       | 10               | No                        | NED (1)       |
| 4        | 43         | M   | 5        | Obvious                  | Mouth, tongue | No       | Paraneoplastic pemphigus | NA   | NA         | NA               | No                        | NA (2)        |
| 5        | 36         | F   | 3        | NA                       | Tonsil      | NA       | Non-specific inflammation | -    | Surg       | 15               | Recurrence, 6 | AWD (3)       |
| 6        | 59         | F   | 4.5      | NA                       | Tonsil      | NA       | Benign tumor       | -    | NA         | 24               | Recurrence, 17 | STD (3)       |
| 7        | 64         | F   | 6.0      | NA                       | Oropharyngeal | NA       | Squamous Carcinoma | -    | Surg+ChT   | 7                | No                        | STD (3)       |
| 8        | 59         | M   | 4.6      | NA                       | Right tonsil | NA       | Lymphoma           | NA   | Surg+RT    | 44               | No                        | NED (4)       |
| 9        | 31         | F   | 3.5      | NA                       | Liver       | NA       | FDCS              | +    | Surg       | 10               | No                        | NED (5)       |
| 10       | 48         | M   | 10.0     | NA                       | Liver       | NA       | FDCS              | +    | Surg       | 2                | No                        | NED (5)       |
| 11       | 54         | F   | 3.5      | NA                       | Spleen      | NA       | FDCS              | NA   | Surg       | 10               | No                        | NED (6)       |
| 12       | 79         | M   | 6.0      | NA                       | Spleen      | NA       | FDCS              | NA   | Surg       | 18               | No                        | NED (6)       |
| 13       | 46         | M   | 12.0     | <10                      | Abdominal   | Yes      | FDCS              | NA   | Surg       | 12               | No                        | NED (7)       |
| 14       | 60         | F   | 2.0      | Obvious                  | Stomach     | NA       | FDCS              | NA   | Surg       | 8                | Recurrence, 8 | STD (8)       |
| 15       | 47         | M   | 4.5      | Few                      | Hepatogastric Ligament | NA       | FDCS              | NA   | Surg       | 3                | No                        | NED (9)       |
| 16       | 46         | F   | 8.6      | 12                       | Retroperitoneal | Yes      | FDCS              | NA   | Surg+ChT+RT| 36               | Metastasis, 36 | AWD (10)      |
| 17       | 67         | F   | 4.0      | NA                       | Liver       | NA       | FDCS              | +    | Surg       | 36               | No                        | NED (11)      |
| 18       | 50         | M   | 3.1      | NA                       | Liver       | NA       | FDCS              | +    | Surg       | 6                | Metastasis, 6 | AWD (12)      |
| 19       | 39         | M   | 18.0     | Rare                     | Infratemporal fossa | Yes      | FDCS              | -    | Surg       | 1                | No                        | STD (13)      |
| 20       | 16         | F   | 8.0      | Rare                     | R Posterior | Yes      | FDCS              | -    | Surg       | 24               | No                        | NED (14)      |
| 21       | 60         | M   | 5.0      | <1                       | Tonsil      | NA       | Granuloma          | NA   | Surg+RT    | 86               | No                        | NED (15)      |
| 22       | 35         | F   | 5.0      | 10                       | Parapharyngeal Space | NA       | Nasopharyngeal carcinoma | NA   | Surg       | 12               | Recurrence, 2 | AWD (15)      |
| 23       | 63         | M   | 4.0      | 1                        | Infratemporal fossa | NA       | PNET              | -    | Surg+RT+ChT| 72               | No                        | NED (15)      |
| 24       | 30         | F   | 5.0      | 9                        | Pyform Sinus | NA       | FDCS              | -    | Surg       | 25               | Metastasis, 25 | AWD (15)      |
| 25       | 23         | M   | 8.0      | 3                        | Mediastinum  | NA       | Malignant nerve sheath tumor | -    | Surg+RT+ChT| 45               | Metastasis, 45 | AWD (15)      |
| 26       | 45         | M   | 14.5     | <1                       | Liver       | NA       | FDCS              | +    | Surg       | 27               | No                        | NED (15)      |
| 27       | 36         | F   | 15.0     | 7                        | Mesentery   | NA       | Malignant GIST    | -    | Surg       | 27               | Metastasis, 4 | AWD (15)      |
| Case no. | Age, years | Sex | Size, cm | Mitotic Counts (/10 HPF) | Site                      | Necrosis | Initial diagnosis | EBER      | Treatment | Follow-up months | Recurrence/metastasis months | Status (Refs.) |
|---------|------------|-----|----------|--------------------------|---------------------------|----------|------------------|-----------|-----------|----------------|-----------------------------|----------------|
| 28      | 28         | F   | 6.0      | 3                        | Parapharyn-Geal Space     | NA       | FDCS             | -         | Surg+RT+ChT | 22             | Metastasis, 14              | AWD (15)       |
| 29      | 55         | M   | 2.0      | 9                        | Tonsil                    | NA       | FDCS             | NA        | Surg+RT    | 21             | Recurrence, 18               | AWD (15)       |
| 30      | 63         | F   | 4.0      | Obvious                  | Urinary bladder           | Yes      | Infiltrating Urothelial Carcinoma | -         | Surg+RT    | 24             | Metastasis, 24               | AWD (16)       |
| 31      | 66         | F   | 2.3      | NA                       | Liver                     | NA       | FDCS             | +         | Surg+RT+ChT | 12             | Metastasis, 12               | AWD (17)       |
| 32      | 65         | M   | 1.0      | 16                       | Tonsil                    | NA       | FDCS             | NA        | Surg+RT    | 24             | No                          | NED (18)       |
| 33      | 19         | F   | 4.0      | >10f                     | Small intestine           | Yes      | FDCS             | -         | Surg+ChT   | 8              | No                          | NED (19)       |
| 34      | 72         | F   | 4.3      | Obvious                  | Middle Mediastinum        | NA       | FDCS             | NA        | RT         | 12             | No                          | STD (20)       |
| 35      | 51         | F   | 9.1      | Obvious                  | Middle Mediastinum        | NA       | FDCS             | NA        | Surg+RT    | 10             | Metastasis, 10               | STD (20)       |
| 36      | 53         | F   | 10.3     | NA                       | Anterior Mediastinum      | NA       | FDCS             | NA        | Surg       | 18             | No                          | NED (20)       |
| 37      | 72         | M   | 3.5      | NA                       | Tonsil                    | NA       | FDCS             | NA        | Surg+ChT   | 12             | No                          | STD (21)       |
| 38      | 43         | M   | 20.0     | NA                       | Mesentry                  | Yes      | FDCS             | NA        | Surg       | 18             | No                          | NED (22)       |
| 39      | 22         | M   | 2.0      | NA                       | Parapharyngeal Space      | NA       | FDCS             | -         | Surg+RT    | 26             | No                          | NED (23)       |
| 40      | 74         | M   | 3.3      | 10-13                    | Small intestine Mesentery | Yes      | FDCS             | NA        | Surg       | 12             | No                          | STD (24)       |
| 41      | 34         | F   | 9.0      | Obvious                  | Small intestine Mesentery | NA       | FDCS             | NA        | Surg+RT    | 96             | No                          | NED (24)       |
| 42      | 31         | M   | 4.7      | 0-1                      | L Parapharynx             | No       | FDCS             | NA        | Surg+RT    | NA             | NA                          | NA (25)        |
| 43      | 73         | M   | 30       | NA                       | Urinary bladder           | No       | FDCS             | NA        | Surg+ChT   | 1.5            | Recurrence, 1.5              | AWD (26)       |
| 44      | 64         | F   | 19.0     | NA                       | Spleen                    | NA       | FDCS             | NA        | Surg       | 36             | No                          | NED (27)       |
| 45      | 63         | M   | NA       | NA                       | L Tonsil                  | NA       | Paraganglioma     | NA        | Surg+RT    | 52             | Recurrence, 52               | AWD (28)       |
| 46      | 26         | M   | NA       | NA                       | Nasopharynx               | NA       | Poorly Differentiated Carcinoma | NA        | Surg+RT+ChT | 44             | Recurrence, 44               | AWD (28)       |
| 47      | 64         | M   | NA       | NA                       | Hypopharynx               | NA       | Poorly Differentiated Carcinoma | NA        | Surg+RT    | 39             | Recurrence, 39               | AWD (28)       |
| 48      | 28         | M   | NA       | NA                       | R Tonsil                  | NA       | FDCS             | NA        | Surg+RT+ChT | NA             | NA                          | NA (28)        |
| 49      | 66         | M   | NA       | NA                       | R Tonsil                  | NA       | FDCS             | NA        | Surg+RT    | 31             | No                          | NED (28)       |
| 50      | 68         | F   | NA       | L Tonsil                  | NA                       | NA       | FDCS             | NA        | Surg       | 19             | Recurrence, 19               | AWD (28)       |
| 51      | 65         | F   | L Tonsil  | NA                       | L Tonsil                  | NA       | FDCS             | NA        | Surg+ChT   | 47             | Recurrence, 47               | AWD (28)       |
Table II. Continued.

| Case no. | Age, years | Sex | Size, cm | Mitotic Counts (/10 HPF) | Site | Necrosis | Initial diagnosis | EBER | Treatment | Follow-up, months | Recurrence/ metastasis, months | Status | (Refs.) |
|----------|------------|-----|----------|-------------------------|------|----------|------------------|------|-----------|-----------------|--------------------------------|--------|--------|
| 52       | 40         | M   | NA       | NA                      | L Tonsil | NA       | FDCS             | NA   | Surg+ChT   | NA              | NA                             | NA     | (28)   |
| 53       | 51         | F   | NA       | NA                      | L Tonsil | NA       | FDCS             | NA   | Surg+ChT   | NA              | NA                             | NA     | (28)   |
| 54       | 38         | M   | NA       | NA                      | R Tonsil | NA       | FDCS             | NA   | Surg+RT+ChT | 45              | Recurrence, 45                | AWD    | (28)   |
| 55       | 24         | M   | NA       | NA                      | Bone    | NA       | FDCS             | -    | ChT       | 108             | No                             | NED    | (29)   |
| 56       | 24         | F   | NA       | NA                      | Pelvic, abdominal Cavity | NA       | FDCS             | -    | Surg+ChT   | 5               | No                             | STD    | (29)   |
| 57       | 61         | F   | NA       | Obvious Intracranial    | NA       | Yes      | FDCS             | NA   | Surg+ChT   | 12              | No                             | STD    | (30)   |
| 58       | 67         | F   | 1.5 15   | Esophagus               | NA       | FDCS     | -               | Surg+RT+ChT | 26              | Metastasis, 24                | STD    | (31)   |
| 59       | 42         | M   | 5.0      | Neck                    | NA       | FDCS     | NA               | Surg | 12        | No              | No                             | NED    | (32)   |
| 60       | 24         | M   | 6.5 8    | Ileocecal region        | NA       | FDCS     | -               | Surg | 12        | No              | No                             | NED    | (33)   |
| 61       | 27         | M   | 2.8 Rare | Tonsil                  | NA       | FDCS     | NA               | Surg+RT | 6              | No               | No                             | NED    | (34)   |
| 62       | 67         | F   | 4.5 Obvious | Pancreas              | Yes      | FDCS     | -               | Surg | NA        | NA              | NA                             | NA     | (35)   |
| 63       | 63         | F   | 13.4 3   | R Liver                 | Yes      | FDCS     | +               | Surg | 48        | No              | No                             | NED    | (36)   |
| 64       | 46         | F   | 11.0     | NA                      | Posterior | Fibrosing | -               | Surg+ChT+RT | 27              | Metastasis, 24                | STD    | (37)   |
| 65       | 44         | F   | NA       | NA                      | Thyroid  | NA       | FDCS             | NA   | Surg+RT    | NA              | NA                             | NA     | (38)   |
| 66       | 59         | F   | 4.7      | NA                      | Soft palate | FDCS     | NA               | Surg+RT | 12              | No               | No                             | NED    | (39)   |
| 67       | 78         | F   | 3.9 Rare | Colonic                 | NA       | FDCS     | +               | Surg | 5         | No              | No                             | NED    | (40)   |
| 68       | 22         | M   | 10       | Mesentery               | NA       | High-Risk GIST | NA       | Surg+ChT | NA | NA  | NA | NA | NA | (41) |
| 69       | 28         | M   | NA       | NA                      | mesentery | GIST     | NA               | NA   | 60        | Metastasis, 48                | AWD    | (42)   |
| 70       | 63         | M   | 12.0     | NA                      | Retroperitoneal | GIST    | NA               | Surg+ChT+RT | 60              | Metastasis, 48                | AWD    | (42)   |
| 71       | 59         | F   | NA       | Obvious Thigh           | Yes      | FDCS     | NA               | NA   | NA        | NA              | NA                             | NA     | (43)   |
| 72       | 70         | M   | 14.0     | NA                      | Pancreas Spleen | Yes | FDCS             | NA       | NA        | NA              | NA                             | NA     | (44)   |
| 73       | 37         | M   | 11.0     | NA                      | Mediastinum | Yes | FDCS             | NA   | Surg+ChT   | 6               | No                             | NED    | (45)   |
| 74       | 39         | M   | 6.0      | NA                      | Mediastinum | NA       | FDCS             | NA   | Surg+RT    | 96              | No                             | NED    | (46)   |
| 75       | 61         | M   | 10.0 Rare | Spleen                 | Yes      | FDCS     | +               | NA   | 12        | No               | No                             | NED    | (47)   |
| 76       | 19         | F   | 6.0      | NA                      | Liver     | NA       | FDCS             | +    | Surg       | 12              | No                             | NED    | (48)   |
| 77       | 60         | M   | 4.5      | NA                      | Neck      | NA       | FDCS             | NA   | Surg       | NA              | No                             | NED    | (49)   |
| 78       | 53         | M   | 1.0      | NA                      | Tonsil    | NA       | FDCS             | NA   | Surg       | NA              | NA                             | NA     | (49)   |
| 79       | 43         | F   | 2.0      | NA                      | Tonsil    | NA       | FDCS             | NA   | Surg       | 51              | No                             | NED    | (49)   |
| 80       | 42         | M   | 12.0     | NA                      | Omentum   | NA       | FDCS             | NA   | Surg+ChT   | 10              | No                             | STD    | (49)   |
| 81       | 45         | M   | 7.5      | NA                      | Posterior | NA       | FDCS             | NA   | Surg       | 36              | No                             | NED    | (49)   |
| 82       | 60         | M   | 2.2      | NA                      | Spleen    | NA       | FDCS             | NA   | Surg       | 5               | No                             | NED    | (49)   |
Table II. Continued.

| Case no. | Age, years | Sex | Size, cm | Mitotic Counts (/10 HPF) | Site       | Necrosis | Initial diagnosis | EBER | Treatment | Follow-up, months | Recurrence/metastasis, months | Status (Refs.) |
|----------|------------|-----|----------|--------------------------|------------|----------|-------------------|------|-----------|------------------|---------------------------|---------------|
| 83       | 46         | F   | 6.0      | NA                       | Liver      | NA       | FDCS              | NA   | Surg      | 2                | No                        | NED (49)      |
| 84       | 71         | M   | 5.5      | NA                       | Spleen     | NA       | FDCS              | +    | Surg      | 26               | No                        | NED (50)      |
| 85       | 32         | M   | 3.0      | NA                       | Liver      | NA       | FDCS              | +    | Surg      | 19               | No                        | NED (50)      |
| 86       | 69         | F   | 9.0      | Yes                      | Spleen     | Yes      | FDCS              | +    | Surg      | 1                | No                        | NED (50)      |
| 87       | 59         | F   | 15.0     | >10                      | Spleen     | NA       | FDCS +            | +    | Surg      | 48               | No                        | NED (51)      |
| 88       | 71         | F   | 4.5      | >10                      | Spleen     | NA       | FDCS              | +    | Surg      | 24               | No                        | NED (51)      |
| 89       | 77         | M   | 4.6      | >10                      | Spleen     | NA       | FDCS              | +    | Surg      | 12               | No                        | NED (51)      |
| 90       | 45         | F   | 1.8      | >10                      | Liver      | NA       | FDCS              | +    | Surg      | 5                | No                        | NED (51)      |
| 91       | 30         | F   | 1.0      | >10                      | Tonsil     | NA       | FDCS              | -    | Surg      | 3                | No                        | NED (51)      |
| 92       | 62         | M   | 12.0     | >3                       | Mesentery  | Yes      | FDCS              | -    | Surg+ChT  | 2                | No                        | NED (52)      |
| 93       | 26         | M   | NA       | NA                       | Spleen     | NA       | FDCS              | -    | Surg      | NA               | NA                        | NA (53)       |
| 94       | 57         | M   | NA       | NA                       | Retroperitoneal | NA | FDCS              | -    | Surg      | NA               | NA                        | NA (53)       |
| 95       | 24         | M   | NA       | NA                       | Pelvis     | NA       | FDCS              | -    | Surg      | 10               | No                        | NED (53)      |
| 96       | 70         | M   | 3.9      | 10                       | Pharyngeal | NA       | FDCS              | -    | Surg      | NA               | NA                        | NA (54)       |
| 97       | 40         | F   | 2.0      | 2                        | Pharyngeal | NA       | Malignant Fibrous | NA   | Surg      | NA               | NA                        | NA (54)       |
| 98       | 38         | M   | 5.0      | 20                       | Pharyngeal | NA       | FDCS              | NA   | Surg      | NA               | NA                        | NA (54)       |
| 99       | 45         | M   | 2.0      | NA                       | Nasal cavity | NA | FDCS              | +    | Surg      | NA               | NA                        | NA (55)       |
| 100      | 59         | F   | 19.0     | NA                       | Small intestine | NA | FDCS              | NA   | Surg+ChT  | NA               | NA                        | NA (55)       |
| 101      | 37         | F   | 3.5      | >30                      | Tonsil     | NA       | FDCS              | NA   | Surg+ChT  | 28               | No                        | STD (55)      |
| 102      | 31         | F   | 10.5     | NA                       | Mesojejenum | NA | FDCS              | NA   | Surg      | NA               | NA                        | NA (55)       |

M, male; F, female; R, right; L, left; NA, not available; Surg, surgery; RT, radiotherapy; ChT, chemotherapy; NED, no evidence of disease; AWD, alive with disease; STD, succumbed to disease; FDCS, follicular dendritic cell sarcoma; HPF, high power field.
Table III. Survival rates.

| Characteristic | Total, n | OS rate, % | DFS rate, % | Recurrence rate, % (n/total) | Metastasis rate, % (n/total) | Mortality rate, % (n/total) |
|----------------|----------|------------|-------------|-------------------------------|------------------------------|-------------------------------|
|                |          | 2-year     | 5-year      | P-value                       | 2-year                      | 5-year                       | P-value                       |
| Available total| 82       | 70         | 70          | 68                            | 32                           | 14.63 (12/82)                | 17.07 (14/82)                | 18.29 (15/82)                |
| Sex            |          |            |             |                               |                              |                              |                              |
| Male           | 42       | 82         | 82          | 0.103                         | 91                           | 0.032                        | 14.29 (6/42)                 | 9.52 (4/42)                  | 11.90 (5/42)                 |
| Female         | 40       | 57         | 57          | 47                            | 34                           | 15.00 (6/40)                 | 25.00 (10/40)                | 25.00 (10/40)                |
| Age, years     |          |            |             |                               |                              |                              |                              |
| <50            | 41       | 78         | 78          | 0.274                         | 74                           | 0.623                        | 9.76 (4/41)                  | 17.07 (7/41)                 | 12.20 (5/41)                 |
| ≥50            | 41       | 64         | 64          | 63                            | 30                           | 19.51 (8/41)                 | 17.07 (7/41)                 | 24.39 (10/41)                |
| Size, cm       |          |            |             |                               |                              |                              |                              |
| <4             | 20       | 36         | /           | 0.119                         | 45                           | 0.235                        | 15.00 (3/20)                 | 15.00 (3/20)                 | 25.00 (5/20)                 |
| ≥4             | 46       | 87         | 77          | 69                            | 47                           | 4.35 (2/46)                  | 21.74 (10/46)                | 15.22 (7/46)                 |
| Treatment      |          |            |             |                               |                              |                              |                              |
| Surgery        | 40       | 89         | /           | 0.109                         | 80                           | 0.567                        | 10.00 (4/40)                 | 7.50 (3/40)                  | 7.50 (3/40)                  |
| Surgery + RT/ChT| 37      | 63         | 63          | 70                            | 24                           | 18.91 (7/37)                 | 24.32 (9/37)                 | 27.30 (10/37)                |

RT, radiotherapy; ChT, chemotherapy.
Kaplan-Meier estimation exhibited no other statistically significant differences between disease-free survival rates or overall survival rates and age, tumor size or treatment (Table III).

**Discussion**

FDCS is an extremely rare tumor that affects the lymphoid tissues and mostly presents in the lymph nodes, while the
extranodal type of the disease accounts for only one-third of cases (5). Due to the limited number of reported cases, a proportion of FDCSs, particularly extranodal FDCSs, has been difficult to recognize, especially on purely morphological grounds (11). FDCS has been proven to derive from DCs or macrophages, making it similar to diffuse large B-cell lymphoma or anaplastic large cell lymphoma, and therefore complicating its diagnosis (6,9). Additionally, the diagnosis of extranodal FDCS is even more challenging (2,5,10). Currently, to the best of our knowledge, the clinical manifestations and prognosis-associated factors of extranodal FDCS have not been statistically described. The present study presented two cases of extranodal FDCS affecting the tonsil and soft tissue of the chest wall, respectively. Additionally, 102 cases of extranodal FDCS from the literature were analyzed (1-55).

The existence of FDC tumors (FDCTs) was first described by Lennert in 1978 (56), but it was Monda et al (57) who in 1986 recognized and characterized this type of tumor. As antigen-presenting cells, DCs can be found in various sites and participate in multiple types of activations (20,22). Langerhans cells are specialized dendritic cells in mucosal sites and skin that upon activation become specialized for antigen presentation to T cells, and then migrate to the lymph node through lymphatics (14,15). In contrast to other types of myeloid-derived DCs (such as Langerhans cells, interdigitating DCs and dermal/interstitial DCs), FDCs seem to stem from bone marrow stromal cells, with myofibroblasts as a characteristic (35,42). FDCs are located in primary and secondary follicles, trapping and presenting antigens to B cells, and storing immune complexes for long periods of time on the cell surface (58,59). The cause of FDCT remains unknown; potential risk factors may be Epstein Barr Virus (EBV) infection or Castleman disease (2,26), which may be found concurrently with FDCS or may precede the latter by several years (27). EBV is suspected to carry a viral oncopogene-latent membrane protein 1 that may encourage transformation, often detected in the spleen and liver (30,34).
Among the 102 cases from the literature, EBER was positive mostly in the liver and spleen, except for one case in the colon. A number of cases appeared to be associated with autoimmune diseases, such as paraneoplastic pemphigus and myasthenia gravis. It has been suggested that FDCS encourages aberrant immune system activation, given that patients often demonstrate immature T cells (8,15).

The epidemiology of FDCS is unclear. A wide age range has been reported, but FDCS was most common in adults (50). Similarly, the mean age of the patients in the present study was 48.87 years (range, 16-79 years), and the median age was 47.5 at initial presentation. The sex distribution was similar, and the overall male to female ratio was 1.17:1. In the present study, FDCS was slightly more common in males, which was inconsistent with the results of Shaw et al (8). The mean diameter of the tumors was 6.63 cm (range, 1-20 cm). Tumor size was closely associated with the primary site and was larger in the inner abdomen compared with other sites. Most FDCS present as lymphadenopathy, but a number of extranodal regions, such as the soft tissue, tonsil, stomach and intestines, were found to be the primary sites. Systemic symptoms were uncommon. Sometimes patients complained of a slow-growing, painless lump, while others visited the doctor presenting with abdominal pain, which was usually due to an abdominal tumor; rarely patients had paraneoplastic pemphigus (such as 2 cases in the present literature review).

The gross observation and histopathology are manifold. Overall, the cut surface of most extranodal FDCS had a yellowish white appearance, was circumscribed and caused extrinsic compression in some cases. Microscopically, the neoplastic cells were arranged in a fascicular pattern and had a storiform structure, with an ovoid-round shape. Similar to meningioma, a whorl pattern was observed in certain areas. At high power, tumor cells with a slightly eosinophilic cytoplasm, distinct elongated nuclei and cell membrane were observed. Lymphocytes were dispersed characteristically in the background. The mitotic rate was 0-10/HPF, with a higher rate of >30/HPF in pleomorphic cases, with easily seen coagulative necrosis and pathological karyokinesis. FDCS is classified into two types, the classic FDCS and inflammatory pseudotumor (IPT)-like FDCS (35,48). IPT-like FDCS is rarer than the classic type and typically presents as a renal and hepatic lump (35,51). Histologically, lymphoplasmacytic spindle cells infiltrate the tumor, mainly including plasma cells, lymphocytes and a small number of neutrophils, sometimes with a lymphoid follicle formation (51). In the 2 cases of the present study, tumor cells were ovoid-to-spindle-shaped, forming solid or nested patterns or whorls arrays. The tumor was lightly infiltrated by small lymphocytes and multinucleate cells. IHC and ISH are essential for the diagnose of FDCS. No single marker is able to identify all DC subsets; as described in the WHO classification (58,60), important markers include D2-40, CD23, CD21 and CD35 (3). In the two cases of the present study, the tumor cells were both positive for CD21.

The diagnosis of extranodal FDCS depends mainly on pathology; therefore, due to its infrequency and non-specific histopathological features, misdiagnosis occurs frequently. The most common reason for misdiagnosis is failure to consider FDCS at the initial pathological evaluation. By reviewing the aforementioned literature, it was found that some cases were misdiagnosed as non-specific inflammation, benign tumor, carcinoma, lymphoma, granuloma, pancreatic neuroendocrine tumour and gastrointestinal stromal tumour (GIST). In the study by Hu et al (3), the misdiagnosis rate was 57%, higher than that in the present study. Carcinoma and FDCS cells are all ovoid cells with avascular nuclei; however, carcinomas are positive for CK and negative for CD21, CD23 and CD35 (21,53). Similarly to FDCS, GISTs exhibit fascicles, storiform arrays and whorls patterns, and are negative for CK, but immune histochemical markers are positive for Dog-1, CD34 and CD117, and negative for specific markers for FDCS, which may be used to distinguish GIST from FDCS (5,19).

The limited cytogenetic data exhibit complex karyotypes. A targeted next-generation sequencing study indicated frequent function loss alterations in tumor suppressor genes, negative regulation of NF-xB and cell-cycle progression involvement (60).

The use of the genomic sequencing approach enhanced the understanding of genomic features of FDCS in the thyroid (38). Extensive mutations were detected, including VEGFR1, CLTCL1 and TP53 mutations and hepatoma-derived growth factor related protein 3 (HDGFRP3) and Src homology 2 domain containing family member 4 (SHC4) (10,19). SHC4 is associated with the EGFR signaling pathway, from which it was deduced that this pathway may serve a role in the etiopathogenesis of FDCS (25). The BRAF V600E mutation was also reported in 0-19% of cases (61,62).

The treatment of FDCS has not been standardized, as there is no worldwide consensus due to the rarity of the reported cases and limited prospective research on prognosis. The shortage of medical molecular genetics hindered the development of targeted treatments. In most cases, patients with FDCS receive surgery and adjuvant radiotherapy or chemotherapy (33,52). Radical dissection is an important treatment of regional lumps, particularly tumors appearing to have clear boarders (63). Postoperative radiotherapy is recommended, with total doses of 6,000-7,000 cGy in the head and neck region (3.8). With regards to chemotherapy, the options targeting non-Hodgkin’s lymphoma are most commonly used (64). However, it remains controversial whether it is beneficial to administer radiotherapy or chemotherapy post-surgically. In the present study, only 50 cases underwent surgery, and combination therapy was administered post-operatively to 45 cases; however, a comparison of prognosis between the surgery only and the adjuvant treatment groups did not yield any significant results (P>0.05).

FDCS is a type of low-intermediate grade malignant tumor. Due to a shortage of cases, the prognosis and predictive factors are not definite. Saygin et al (65) reported 2-year survival rates for early stage, local infiltration and distant metastasis stage of 82, 80 and 42%, respectively. By reviewing the data of 42 FDCS cases in the tonsils, Lu et al (4) revealed that the 3-year overall survival rate was 86.5%, a little higher than the 5- and 8-year rates (both 77.8%). By reviewing 32 subjects of mediastinal FDCS, Wu et al (1) identified that the 1-year total and tumor-free survival rates were 80.4 and 76.9%, respectively, the 3-year total and tumor-free survival rates were 68.5 and 51.7%, respectively, and the 5-year rates were 58.8 and 32.3%, respectively.

According to the investigation of WHO, prognostic analysis of extranodal FDCS is scarce. In the present
study, the follow-up duration was 1-108 months, with an average of 24.31±22.98 months, and the 2- and 5-year total survival rates were both 70%. The 2- and 5-year disease-free total survival rates were 68 and 32%, respectively. Domínguez-Malagón et al (66) demonstrated that FDCS originating from the pharyngeal region had low recurrence (25%), metastasis (25%) and mortality rates (5%), similar to those of Duan et al (67) (23, 21 and 3%, respectively). In the present study, the rates of recurrence, metastasis and mortality were 14.63 (12/82), 17.07 (14/82) and 18.29% (15/82), respectively. These different results may be due to the limitation of the tumor sites. FDCS in the parapharyngeal space exhibited poorer outcomes, while intra-abdominal tumors are more likely to recur (40,42). However, in the present study, the follow-up data available for analysis were scarce, the follow-up time was short and the survival curves were founded on a small number of cases, affecting the availability and effectiveness of the present study. The prognostic factors of extranodal FDCS remain unclear, and may include tumor diameter, necrosis and mitotic count (65). Lu et al (4) reported that a large tumor size resulted in a poor prognosis, and Hu et al (3) detected that combined treatment improved survival rates. The current study revealed that sex was a significant prognostic factor. However, Kaplan-Meier estimation exhibited no other statistically significant differences between disease-free survival rates or overall survival rates and age, tumor size or treatment. Due to the scarcity of the follow-up data available for analysis, the current data are insufficient, and more data and further analyses are urgently required.

In conclusion, two rare cases of primary extranodal FDCS were presented, and 102 cases from the literature were reviewed. The present study described the known biological behavior of extranodal FDCS. The confirmation of pathology of extranodal FDCS is challenging, leading to further delays in diagnosis. Surgical resection remains essential for definitive treatment. Further research into the pathogenesis and therapy of FDCS is required to improve the outcomes of this rare disease.

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Availability of data and materials
The datasets used and/or analyzed during the present study are available from the corresponding author upon reasonable request.

Authors' contributions
XZ collected and analyzed the data, GZ made substantial contributions to the acquisition of data, analysis and interpretation of data and DS made substantial contributions to conception and design. All authors wrote the manuscript. All authors have read and approved the final manuscript.

Ethics approval and consent to participate
The study was approved by the Ethics Committee of the Affiliated Hospital of Chengde Medical College (Chengde, China; approval no. LL049). Written informed consent was obtained from the patients for the storage of samples and data, follow-up contact, and further use of samples and data for research purposes.

Patient consent for publication
Not applicable.

Competing interests
The authors declare that they have no competing interests.

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