

# Les sarcomes folliculaires dendritiques

# Les critères de Mulrow

Pour être qualifiée de systématique, la revue doit répondre aux **critères de Mulrow** :

1. spécification de l'objectif de la revue
2. source des données: éviter les biais liés à la sélection des publications
3. sélection des données: critères (inclusion/ exclusion) utilisés pour la revue
4. évaluation de la validité et de la qualité des études (critères à définir au départ)
5. synthèse des données: qualitative et quantitative (méta-analyse)
6. résumé et discussion des investigations futures à mener

**Tableau 1** Classification des histiocytoses<sup>a</sup>.

*Histiocytoses langerhansiennes (HL)<sup>b</sup>*

Formes localisées

Granulome éosinophile

HL pulmonaire

HL cutanée isolée

Formes systémiques

Sans atteinte d'organe à risque (syndrome de Hand-Schüller-Christian)

Avec atteinte d'organes à risque<sup>c</sup> (syndrome de Letterer-Siwe)

*Histiocytoses non langerhansiennes (HNL)*

HNL d'origine monocytaire/macrophagique

HNL d'origine exogène

HNL métaboliques

HNL infectieuses

HNL hémophagocytaires

Maladie de Destombes-Rosai-Dorfman

Réticulo-histiocytose multicentrique

HNL à cellules dendritiques (dendrocyte interstitiel) :

HNL de type xanthogranulome juvénile (XGJ)

HNL de type XGJ à prédominance cutanéomuqueuse

HNL de type XGJ atteignant la peau avec une prédominance extra-cutanée

HNL de type XGJ essentiellement extra-cutanée

Maladie d'Erdheim-Chester<sup>b</sup>

Histiocytose à cellules indéterminées

*Histiocytoses malignes (HM)*

HM d'origine monocytaire/macrophagique

Sarcomes histiocytaires/macrophagiques<sup>b</sup>

Leucémies myélonocytaires

HM d'origine dendritique

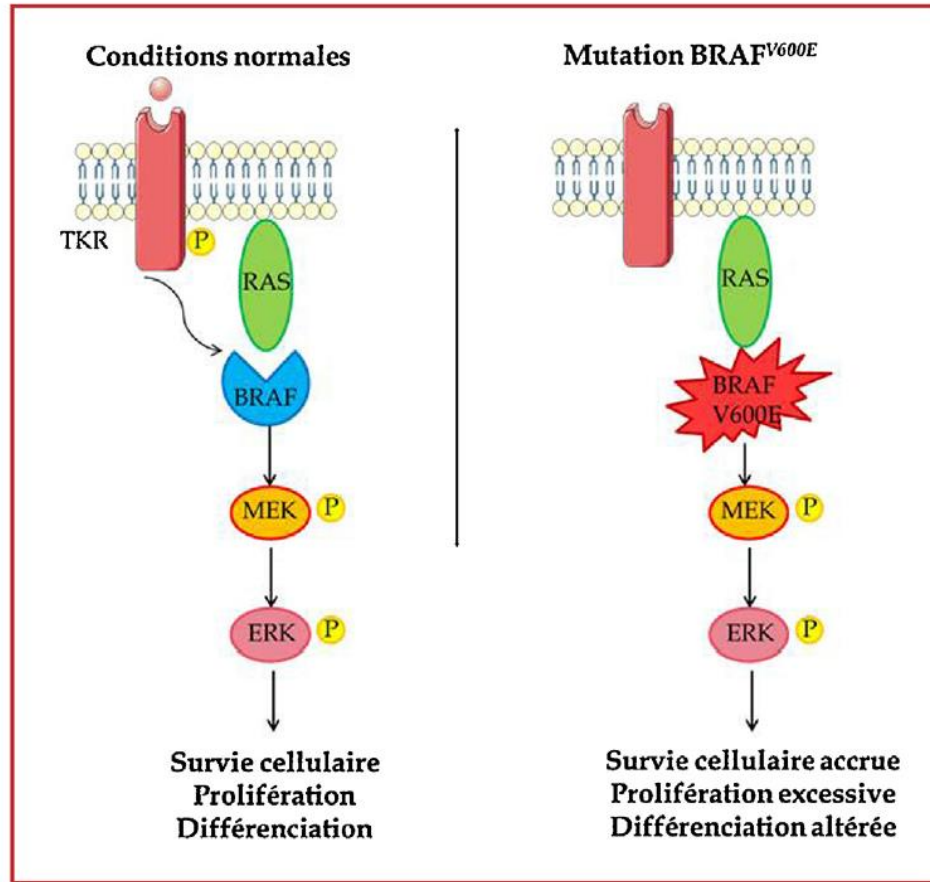
Sarcomes à cellules de Langerhans<sup>b</sup>

Sarcomes à cellules interdigitées

<sup>a</sup> Adaptée de la référence [1]. Des formes de passage ou mixtes existent.

<sup>b</sup> Pathologies dans lesquelles la mutation BRAF<sup>V600E</sup> a été décrite.

<sup>c</sup> Organes à risque : foie, rate, atteinte hématologique.



**Figure 4.** Représentation schématique de la voie de signalisation RAF-MEK-ERK en situation normale et cas de mutation BRAF<sup>V600E</sup>. Adaptée à partir de la référence [96].

# Deux formes de sarcomes dendritiques

- à cellules folliculaires dendritiques
- à cellules inter-digitées dendritiques

# Interdigitating and Follicular Dendritic Cell Sarcomas A SEER Analysis

Stephanie M. Perkins, MD and Eric T. Shinohara, MD, MSCI

**Objectives:** Follicular dendritic cell sarcoma (FDCS) and interdigitating dendritic cell sarcoma (IDCS) are rare neoplasms of dendritic cell origin. Because of the rarity of these diagnoses, optimal management is unclear.

**Methods:** In this study, we reviewed the data on FDCS and IDCS available in the Surveillance, Epidemiology, and End Results database. Fifty-four patients with FDCS and 20 with IDCSs were identified between the years 2001 and 2008.

**Results:** Median follow-up was 28 months. Sixty-one percent of FDCS patients and 55% of IDCS patients presented with localized disease. Of the FDCS patients with localized disease, 31/33 (94%) underwent surgical resection. Fifty-five percent (6/11) of localized IDCS patients underwent surgical resection. Radiation therapy was given to 30% of patients. Overall survival was significantly better for patient with FDCS compared to those with IDCS. Median survival was 35 months in patients with IDCS and was not reached in patients with FDCS. There was a trend toward improved overall survival in FDCS patients with localized disease. IDCS patients with localized disease had a significantly improved overall survival compared with those with distant disease with 2-year overall survival of 72% versus 33%, respectively ( $P=0.05$ ).

**Conclusions:** These data demonstrate that most patients with localized disease are treated similar to a soft tissue sarcoma with primary surgical resection with or without radiation. No chemotherapy data were available in the Surveillance, Epidemiology, and End Results database. The role of chemotherapy and radiation therapy remains unclear.

**Key Words:** SEER, dendritic cell sarcoma, follicular

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reported in the literature, and the clinical course and optimal treatment for these neoplasms is unclear.

FDCS most commonly presents in the head and neck at both nodal and extranodal sites. These tumors typically occur during middle age but have been reported from childhood to late adulthood.<sup>1,4</sup> IDCS is rarer than FDCS, and previous reviews have indicated that this variant may have a poorer outcome.<sup>1</sup> Treatment for both IDCS and FDCS is often approached similar to that of sarcoma with surgical excision, followed by either observation, radiation, chemotherapy, or a combination of chemotherapy and radiation. Surgery is potentially curable in the setting of localized disease; however, these tumors have the potential to recur both locally and/or distantly.<sup>1,5</sup> The efficacy of adjuvant radiation therapy or chemotherapy is unclear.<sup>6</sup>

Because of the rarity of both IDCS and FDCS, prospective data on optimal management of these patients do not exist. The present study was designed to evaluate patient characteristics, treatment modalities, and outcome using the Surveillance, Epidemiology, and End Results (SEER) database.

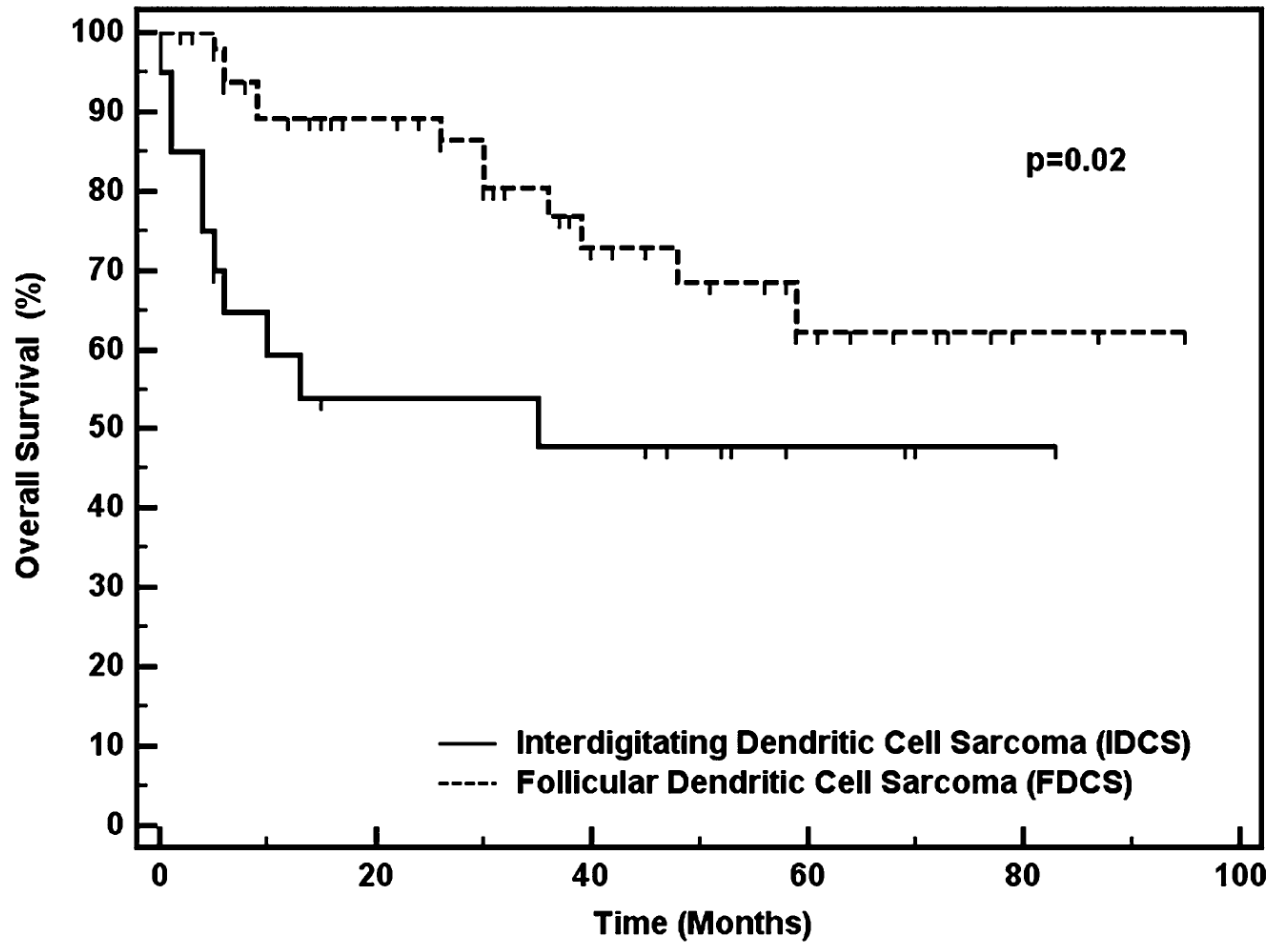
## MATERIALS AND METHODS

The present study is a retrospective review of data extracted from the National Cancer Institute's SEER database. Patients from 1973 to 2008 provided by 17 registries were identified (November 2010 submission).

Cases of interdigitating dendritic sarcomas were identified using the International Classification for Childhood Cancer site recode extended ICD-0-3 histology code 9757/3 (IDCS, Inter-

**TABLE 1.** Patient Demographics, Tumor Characteristics, and Treatment Information

| <b>Variables</b>     | <b>Categories</b>  | <b>Follicular<br/>Dendritic Cell<br/>Sarcomas,<br/>N (%)</b> | <b>Interdigitating<br/>Dendritic<br/>Cell Sarcomas,<br/>N (%)</b> |
|----------------------|--------------------|--------------------------------------------------------------|-------------------------------------------------------------------|
| Age                  | <40                | 16 (29)                                                      | 2 (10)                                                            |
|                      | 40-49              | 9 (17)                                                       | 1 (5)                                                             |
|                      | 50-59              | 14 (26)                                                      | 3 (15)                                                            |
|                      | > 60               | 15 (28)                                                      | 14 (70)                                                           |
| Sex                  | Male               | 27 (50)                                                      | 12 (60)                                                           |
|                      | Female             | 27 (50)                                                      | 8 (40)                                                            |
| Race                 | White              | 45 (83)                                                      | 16 (80)                                                           |
|                      | Black              | 4 (7)                                                        | 3 (15)                                                            |
|                      | Other              | 3 (6)                                                        | 1 (5)                                                             |
|                      | Unknown            | 2 (4)                                                        | 0 (0)                                                             |
| Grade                | II                 | 3 (6)                                                        | 2 (10)                                                            |
|                      | III                | 5 (9)                                                        | 2 (10)                                                            |
|                      | IV                 | 5 (9)                                                        | 3 (15)                                                            |
|                      | Unknown            | 41 (76)                                                      | 13 (65)                                                           |
| Stage                | Local              | 33 (61)                                                      | 11 (55)                                                           |
|                      | Distant            | 17 (32)                                                      | 9 (45)                                                            |
|                      | Unknown            | 4 (7)                                                        | 0 (0)                                                             |
| Year<br>of diagnosis | 2001-2004          | 22 (41)                                                      | 10 (50)                                                           |
|                      | 2005-2008          | 32 (59)                                                      | 10 (50)                                                           |
| Marital status       | Yes                | 30 (56)                                                      | 15 (75)                                                           |
|                      | No                 | 20 (37)                                                      | 5 (25)                                                            |
|                      | Unknown            | 4 (7)                                                        | 0 (0)                                                             |
| Radiation            | No                 | 37 (68)                                                      | 13 (65)                                                           |
|                      | Yes                | 16 (30)                                                      | 6 (30)                                                            |
|                      | Unknown            | 1 (2)                                                        | 1 (5)                                                             |
| Surgery              | Yes                | 44 (81)                                                      | 9 (45)                                                            |
|                      | No                 | 8 (15)                                                       | 9 (45)                                                            |
|                      | Unknown            | 2 (4)                                                        | 2 (10)                                                            |
| Primary site         | Head and<br>neck   | 16 (30)                                                      | 3 (15)                                                            |
|                      | Abdomen/<br>pelvis | 19 (35)                                                      | 2 (10)                                                            |
|                      | Thorax             | 10 (19)                                                      | 6 (30)                                                            |
|                      | Other              | 4 (7)                                                        | 5 (25)                                                            |
|                      | Extremity          | 5 (9)                                                        | 4 (20)                                                            |



**FIGURE 1.** Kaplan-Meier curve of the overall survival of patients with follicular dendritic cell sarcoma and interdigitating dendritic cell sarcoma.



# Dendritic Cell and Histiocytic Neoplasms: Biology, Diagnosis, and Treatment

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**Background:** Dendritic and histiocytic cell neoplasms are rare malignancies that make up less than 1% of all neoplasms arising in lymph nodes or soft tissues. These disorders have distinctive disease biology, clinical presentations, pathology, and unique treatment options. Morphology and immunohistochemistry evaluation by a hematopathologist remains key for differentiating between these neoplasms. In this review, we describe tumor biology, clinical features, pathology, and treatment of follicular dendritic cell sarcoma, interdigitating dendritic cell sarcoma, indeterminate dendritic cell sarcoma, histiocytic sarcoma, fibroblastic reticular cell tumors, and disseminated juvenile xanthogranuloma.

**Methods:** A literature search for articles published between 1990 and 2013 was undertaken. Articles are reviewed and salient findings are systematically described.

**Results:** Patients with dendritic cell and histiocytic neoplasms have distinct but variable clinical presentations; however, because many tumors have recently been recognized, their true incidence is uncertain. Although the clinical features can present in many organs, most occur in the lymph nodes or skin. Most cases are unifocal and solitary presentations have good prognoses with surgical resection. The role of adjuvant therapy in these disorders remains unclear. In cases with disseminated disease, prognosis is poor and data on treatment options are limited, although chemotherapy and referral to a tertiary care center should be considered. Excisional biopsy is the preferred method of specimen collection for tissue diagnosis, and immunohistochemistry is the most important diagnostic method for differentiating these disorders from other entities.

**Conclusions:** Dendritic cell and histiocytic cell neoplasms are rare hematological disorders with variable clinical presentations and prognoses. Immunohistochemistry remains important for diagnosis. Larger pooled analyses or clinical trials are needed to better understand optimal treatment options in these rare disorders. Whenever possible, patients should be referred to a tertiary care center for disease management.

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**Table 1. — Differential Diagnosis of Dendritic Cell Sarcomas**

| <b>FDCS and IDCS</b>                                                                                                                                                                                                               | <b>FRCT</b>                                                       | <b>HS</b>                                                                                                                                                                | <b>INDCS</b>                                                                                                   | <b>JXG</b>                                                                                                                                  |
|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------------|
| Anaplastic large cell lymphoma<br>IDCS (for FDCS) or FDCS<br>(for IDCS)<br>Inflammatory pseudotumors<br>Intranodal myofibroblastoma<br>LCH<br>Non-Hodgkin lymphoma<br>Peripheral nerve sheath tumors<br>True histiocytic lymphomas | FDCS<br>IDCS<br>INDCS<br>Palisaded<br>myofibroblastoma<br>Sarcoma | Anaplastic large cell<br>lymphoma<br>Diffuse large B-cell<br>lymphoma<br>Hemophagocytic<br>lymphohistiocytosis<br>LCH<br>Lymphoma<br>Metastatic carcinoma<br>or melanoma | LCH<br>Pityriasis rosea<br>Scabies<br>T-cell lymphomas<br>(cutaneous T-cell hyperplasia,<br>mycosis fungoides) | Dermatofibroma<br>Eruptive xanthomas<br>LCH<br>Mastocytoma<br>Papular xanthoma<br>Spitz nevus<br>Tuberous xanthoma<br>Xanthoma disseminatum |

FDCS = follicular dendritic cell sarcoma, FRCT = fibroblastic reticular cell tumor, HS = histiocytic sarcoma, IDCS = interdigitating dendritic cell sarcoma, INDCS = indeterminate dendritic cell sarcoma, JXG = juvenile xanthogranuloma, LCH = Langerhans cell histiocytosis.

**Table 2. — Clinical and Pathological Findings of Dendritic Cell Sarcomas**

|                                               | <b>FDGS</b>                                                             | <b>IDCS</b>                                                 | <b>INDCS</b>                                                                        | <b>HS</b>                                                                            | <b>FRCT</b>                                                                                                               | <b>JXG</b>                                                                                          |
|-----------------------------------------------|-------------------------------------------------------------------------|-------------------------------------------------------------|-------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------|
| <b>Clinical Findings (usual presentation)</b> | Slow growing mass, usually a lymph node                                 | Asymptomatic solitary lymph node mass                       | Papules, nodules, or plaques on the skin                                            | Solitary mass with systemic symptoms<br>Can have skin lesions (rash-like)            | Asymptomatic mass                                                                                                         | Small solitary papule                                                                               |
| <b>Cytomorphology</b>                         | Spindle to ovoid cells with whorls                                      | Spindle to ovoid cells with whorls                          | Resembles Langerhans cells with irregular nuclear grooves and clefts                | Large and round to oval shape with focal areas of spindling                          | Spindle to ovoid cells with whorls in paracortical areas                                                                  | Small and oval with a bland round to oval nucleus without grooves                                   |
| <b>Immunophenotypical Markers</b>             | CD4 (+)<br>CD21 (+)<br>CD34 (-)<br>CD35 (+)<br>CD68 (+/-)<br>Fascin (+) | CD4 (+)<br>CD45 (+/-)<br>CD68 (+)<br>Fascin (+)<br>S100 (+) | CD1a (-)<br>CD4 (+)<br>Fascin (+)<br>S100 (+)<br>CD68 (+/-)<br>Birbeck granules (-) | CD163 (+)<br>CD68 (+)<br>Lysozyme (+)<br>CD1a (-)<br>CD21 (-)<br>CD35 (-)<br>CD33(-) | Vimentin (+)<br>Desmin (+)<br>Smooth muscle actin (+)<br>Factor XIIIa (+)<br>CD21 (-)<br>CD35 (-)<br>S100 (-)<br>CD1a (-) | Vimentin (+)<br>sCD14(+)<br>CD68 (+)<br>Stabilin-1 (+)<br>CD163 (+)<br>Factor XIIIa (+)<br>CD1a (-) |
| <b>Treatment for Limited Disease</b>          | Surgical resection ± adjuvant chemotherapy or RT                        | Surgical resection or RT                                    | Surgical excision                                                                   | Surgical resection ± RT                                                              | Surgical resection ± RT                                                                                                   | None needed for localized asymptomatic lesion                                                       |
| <b>Treatment for Disseminated Disease</b>     | Lymphoma-type chemotherapy                                              | Lymphoma-type chemotherapy                                  | Multimodality                                                                       | Lymphoma-type chemotherapy                                                           | Participation in a clinical trial                                                                                         | Langerhans histiocytosis–based treatment                                                            |

FDGS = follicular dendritic cell sarcoma, FRCT = fibroblastic reticular cell tumor, HS = histiocytic sarcoma, IDCS = interdigitating dendritic cell sarcoma, INDCS = indeterminate dendritic cell sarcoma, JXG = juvenile xanthogranuloma, RT = radiation therapy.

# Dendritic cell sarcoma: A pooled analysis including 462 cases with presentation of our case series

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Table 3  
Demographic and clinical features of our and reported cases.

|                                  | FDCS (343 cases)<br>[10–190] | IDCS (100 cases)<br>[191–253] | FRCT (19 cases)<br>[254–264] |
|----------------------------------|------------------------------|-------------------------------|------------------------------|
| Age                              |                              |                               |                              |
| Range                            | 9–90 years                   | 1.8–88 years                  | 13–80 years                  |
| Median                           | 50 years                     | 56.5 years                    | 61 years                     |
| Gender                           |                              |                               |                              |
| Male:female                      | 169:172 (1:1.01)             | 58:42 (1.38:1)                | 11:8 (1.38:1)                |
| Ethnicity                        |                              |                               |                              |
| Asian                            | NA                           | 16/40                         | 4/13                         |
| USA, white                       | NA                           | 11/40                         | 6/13                         |
| USA, black                       | NA                           | 2/40                          | –                            |
| Caucasian                        | NA                           | 8/40                          | 2/13                         |
| Hispanic                         | NA                           | 2/40                          | 1/13                         |
| Other                            | NA                           | 1/40                          | –                            |
| Presentation                     |                              |                               |                              |
| Nodal                            | 105/334                      | 45/95                         | 16/19                        |
| Extranodal                       | 194/334                      | 23/95                         | 3/19                         |
| Both                             | 35/334                       | 27/95                         | –                            |
| Involved sites (number of cases) |                              |                               |                              |
| Lymph nodes                      |                              |                               |                              |
| Cervical                         | 77                           | 41                            | 9                            |
| Axillary                         | 22                           | 22                            | 3                            |
| Mediastinal                      | 21                           | 9                             | 8                            |
| Abdominal                        | 24                           | 18                            | 5                            |
| Inguinal                         | 5                            | 10                            | –                            |
| Epirochlear                      | –                            | –                             | 1                            |
| Extranodal sites                 |                              |                               |                              |
| Liver                            | 45                           | 13                            | 2                            |
| Lung                             | 31                           | 10                            | 2                            |
| Tonsil                           | 27                           | 3                             | –                            |
| Spleen                           | 25                           | 10                            | 2                            |
| Soft tissue                      | 24                           | 1                             | 1                            |
| Mediastinum                      | 20                           | –                             | –                            |
| GIT                              | 18                           | 11                            | –                            |
| Retroperitoneum                  | 17                           | –                             | –                            |
| Mesentery                        | 13                           | 1                             | –                            |
| Bone                             | 11                           | 5                             | 1                            |
| Nasopharynx                      | 10                           | 3                             | –                            |
| Parapharyngeal space             | 10                           | –                             | –                            |
| Oral cavity                      | 9                            | 1                             | –                            |
| Breast                           | 8                            | 2                             | –                            |
| Pancreas                         | 5                            | –                             | –                            |
| Thyroid                          | 4                            | –                             | –                            |
| Bone marrow                      | 2                            | 11                            | –                            |
| Skin                             | 2                            | 7                             | –                            |
| Chest wall                       | 2                            | 3                             | –                            |
| Kidney                           | 2                            | 2                             | 1                            |
| Pleura                           | 1                            | 3                             | –                            |
| Testis                           | –                            | 2                             | –                            |
| Bladder                          | –                            | 2                             | –                            |
| CNS                              | 1                            | 2                             | –                            |
| Other                            | 26 <sup>a</sup>              | 6 <sup>b</sup>                | 1 <sup>c</sup>               |

FDCS, follicular dendritic cell sarcoma; IDCS, interdigitating dendritic cell sarcoma; FRCT, fibroblastic reticular cell tumor; GIT, gastrointestinal tract; CNS, central nervous system; NA, not available.

<sup>a</sup> Other rare extranodal sites of FDCS included omentum, peritoneum, ovary, thymus, adrenal gland, abdominal wall, diaphragm, infratemporal fossa, piriform sinus, oropharynx, hypopharynx, ethmoid/sphenoid sinus, muscle, lip and dura mater.

<sup>b</sup> Less common extranodal sites of IDCS included nose, ovary, cervix, eyelid and heart.

<sup>c</sup> Adrenal gland was involved in our case 1.

Table 4

Univariate analysis of factors associated with adverse outcome (i.e. local recurrence, distant metastasis or death) (chi-square method).

|                                | FDCS (number with positive events/total number of patients) | <i>p</i> value | IDCS (number with positive events/total number of patients) | <i>p</i> value | FRCT (number with positive events/total number of patients) | <i>p</i> value |
|--------------------------------|-------------------------------------------------------------|----------------|-------------------------------------------------------------|----------------|-------------------------------------------------------------|----------------|
| Age                            |                                                             |                |                                                             |                |                                                             |                |
| ≤40 years                      | 48/79                                                       |                | 20/27                                                       |                | 2/6                                                         |                |
| >40 years                      | 68/157                                                      | 0.016          | 21/47                                                       | 0.027          | 5/10                                                        | 0.633          |
| Gender                         |                                                             |                |                                                             |                |                                                             |                |
| Male                           | 51/113                                                      |                | 26/47                                                       |                | 4/10                                                        |                |
| Female                         | 65/123                                                      | 0.292          | 15/27                                                       | 0.823          | 3/6                                                         | 1.000          |
| Tumor type                     |                                                             |                |                                                             |                |                                                             |                |
| CIRC                           | NR                                                          |                | NR                                                          |                | 5/11                                                        |                |
| CNRC                           | NR                                                          |                | NR                                                          |                | 2/5                                                         | 1.000          |
| Presentation                   |                                                             |                |                                                             |                |                                                             |                |
| Nodal                          | 33/58                                                       |                | 10/29                                                       |                | 5/14                                                        |                |
| Extranodal                     | 75/149                                                      |                | 7/18                                                        |                | 2/2                                                         |                |
| Nodal and extranodal           | 11/30                                                       | 0.198          | 22/24                                                       | <0.001         | -                                                           | 0.175          |
| Intraabdominal involvement     |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 40/78                                                       |                | 18/21                                                       |                | 3/5                                                         |                |
| Not present                    | 76/158                                                      | 0.748          | 23/53                                                       | 0.002          | 4/11                                                        | 0.596          |
| Classical morphology*          |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 79/159                                                      |                | 12/31                                                       |                | 2/9                                                         |                |
| Not present                    | 15/25                                                       | 0.457          | 13/22                                                       | 0.235          | 2/3                                                         | 0.236          |
| Lymphoplasmacytic infiltration |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 71/153                                                      |                | 21/43                                                       |                | 4/11                                                        |                |
| Not present                    | 6/6                                                         | 0.011          | 6/13                                                        | 0.883          | 1/1                                                         | 0.416          |
| Epithelioid cells              |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 12/25                                                       |                | 3/4                                                         |                | 1/3                                                         |                |
| Not present                    | 87/169                                                      | 0.912          | 24/28                                                       | 0.343          | 4/9                                                         | 1.000          |
| Giant cells                    |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 18/42                                                       |                | 8/13                                                        |                | 2/4                                                         |                |
| Not present                    | 81/152                                                      | 0.306          | 19/40                                                       | 0.575          | 3/8                                                         | 1.000          |
| Tumor size                     |                                                             |                |                                                             |                |                                                             |                |
| ≥6 cm                          | 37/71                                                       |                | 8/14                                                        |                | 1/2                                                         |                |
| <6 cm                          | 27/79                                                       | 0.040          | 12/29                                                       | 0.519          | 4/10                                                        | 1.000          |
| Necrosis                       |                                                             |                |                                                             |                |                                                             |                |
| Present                        | 28/62                                                       |                | 8/14                                                        |                | 3/6                                                         |                |
| Absent                         | 30/48                                                       | 0.106          | 7/22                                                        | 0.247          | 2/3                                                         | 1.000          |
| Mitosis                        |                                                             |                |                                                             |                |                                                             |                |
| ≥5/10 HPF                      | 35/55                                                       |                | 6/11                                                        |                | 3/5                                                         |                |
| <5/10 HPF                      | 25/61                                                       | 0.024          | 7/14                                                        | 0.859          | 1/5                                                         | 0.524          |

FDCS, follicular dendritic cell sarcoma; IDCS, interdigitating dendritic cell sarcoma; FRCT, fibroblastic reticular cell tumor; CIRC, cytokeratin positive interstitial reticular cell tumor; CNRC, cytokeratin negative interstitial reticular cell tumor

NR, not relevant as this classification is only for FRCT.

\* Histological pattern common to all three subtypes includes spindled to ovoid cells forming fascicles, whorls, sheets, or nodules.

Table 5  
Multivariate Cox regression analysis for prognostic factors.

|                                           | FDCS ( <i>n</i> = 50 cases) |                |
|-------------------------------------------|-----------------------------|----------------|
|                                           | HR (95% CI)                 | <i>p</i> value |
| Age ( $\leq 40$ years)                    | 0.907 (0.318–2.586)         | 0.855          |
| Intraabdominal involvement                | 0.613 (0.184–2.047)         | 0.426          |
| Absence of lymphoplasmacytic infiltration | 6.330 (1.592–25.165)        | 0.009          |
| Tumor size ( $\geq 6$ cm)                 | 5.263 (1.483–18.675)        | 0.010          |
| Necrosis                                  | 0.638 (0.199–2.045)         | 0.450          |
| Mitosis ( $\geq 5/10$ HPF)                | 1.461 (0.461–4.625)         | 0.519          |

FDCS, follicular dendritic cell sarcoma; HR, hazard ratio; CI, confidence interval.

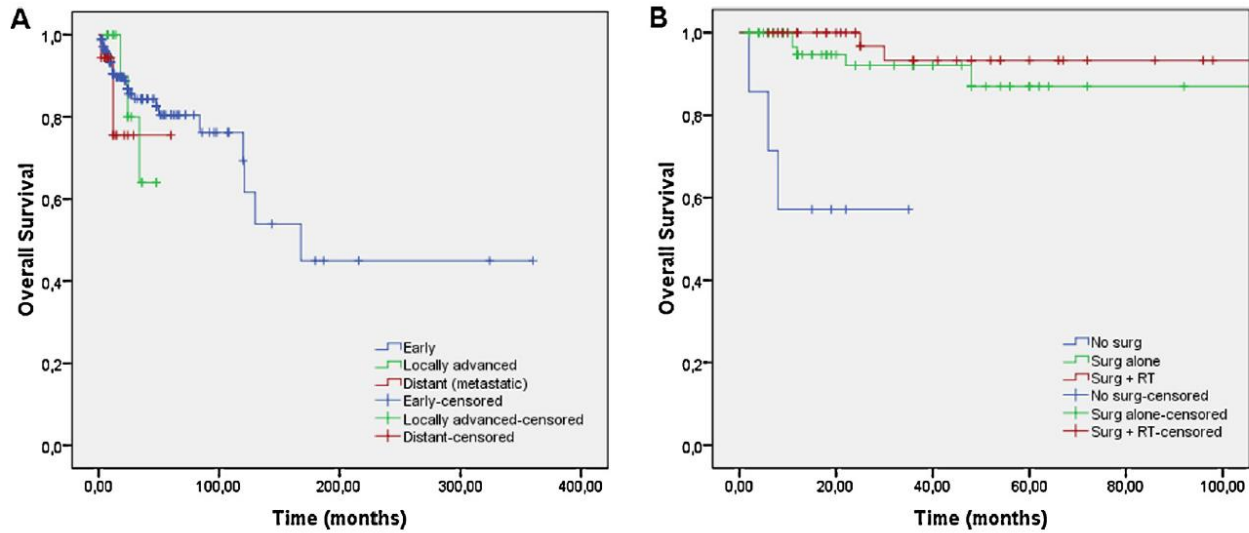


Fig. 1. Kaplan–Meier curve of overall survival (OS) of patients with follicular dendritic cell sarcoma (FDCCS). (A) Stage at presentation (early, locally advanced, or with distant metastasis) had no significant influence on OS of cases ( $p = 0.44$ ). (B) Localized FDCCS cases who received surgery had a significantly better OS when compared to patients who had other treatment modalities ( $p < 0.001$ ). There was no significant difference between OS of patients who received adjuvant radiotherapy and surgery alone ( $p = 0.474$ ).



# Etiologie et associations

- Étiologie virale :
  - EBV
  - herpès virus humain 8 (HHV-8)
- Maladie de Castleman (précurseur?): pathologie lymphoproliférative atypique d'étiologie indéterminée
- Autoimmunité: pemphigus, myasthénie