Interdigitating Dendritic Cell Sarcoma

A Report of Four Cases and Review of the Literature

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Abstract

To better define the clinical and pathologic features of interdigitating dendritic cell sarcoma (IDCS), we report 4 cases, including the first reported in the tonsil. There were 2 male and 2 female patients (mean age, 70 years). Sites of tumor included 1 case each in the right cervical lymph node, left axillary lymph node, right tonsil, and right inguinal lymph node. Histologically, all showed diffuse effacement of the lymphoid tissue by pleomorphic round to spindled cells with convoluted nuclei and abundant eosinophilic cytoplasm. All were immunoreactive for S-100, CD68, lysozyme, and vimentin. CD45 was positive in 3 cases and CD1a in 1 case. Fascin was positive in 3 cases. Other immunostains, including CD3, CD20, CD21, CD30, actin, cytokeratin, and HMB-45, were negative. Ultrastructurally, the tumor cells were elongated and showed indented nuclei, variable numbers of lysosomes, and interdigitating cytoplasmic processes. Follow-up was available for all cases. One patient died of widespread disease 2 months after diagnosis. One was alive with metastatic lung disease at 12 months. Two patients were disease free at 5 and 9 months.

Interdigitating dendritic cells are arachnoid nonlymphoid accessory cells found in the T-cell areas of peripheral lymphoid tissue, including the paracortex and deep cortex of lymph nodes, the splenic periarteriolar lymphoid sheaths, and the interfollicular areas of mucosa-associated lymphoid tissue.[1-4] They are potent antigen-presenting cells responsible for initiating primary T-lymphocyte immune responses.[2,3,5] Interdigitating dendritic cells are immunoreactive with S-100–like Langerhans cells and have a similar light microscopic appearance. However, in contrast with Langerhans cells, they lack Birbeck granules ultrastructurally and consistent CD1a immunostaining.[1,6-26] Neoplasms derived from interdigitating dendritic cells are rare, with only 25 cases previously reported in the literature. Their pathologic diagnosis often is challenging because they mimic other primary and metastatic spindle cell neoplasms of lymph nodes. The reported cases highlight this rare malignant neoplasm; describe the morphologic, immunohistochemical, and ultrastructural features; and expand the known sites of occurrence.

Materials and Methods

H&E-stained slides, paraffin-embedded tissue blocks, and a clinical history were available for each case. For case 1, small pieces of tissue were fixed in glutaraldehyde for electron microscopic evaluation. Immunohistochemical analysis was performed using the standard avidin-biotin-peroxidase method with a panel of antibodies[27] **Table 1**. When necessary, predigestion with 0.04% pepsin at pH 2.0 for 20 minutes at 40°C was performed. Microwave antigen retrieval involved heating sections in a microwave oven in a 1-mmol/L concentration of EDTA buffer solution at pH 8.0 for 10 minutes.

Table 1
Antibodies Used and Sources

Antibody (Clone)	Type	Source	Dilution	Pretreatment
Cytokeratin (AE1/AE3)	M	Boehringer, Indianapolis, IN	1:400	Enzyme
Vimentin (V9)	M	DAKO, Carpinteria, CA	1:80	Enzyme
S-100	R	DAKO	1:100	—
HMB-45	M	ENZO BIOCHEM, Farmingdale, NY	1:2,000	—
Alpha$_1$-antitrypsin	R	DAKO	1:1,000	Enzyme
Lysozyme	R	DAKO	1:1,000	Enzyme
Smooth muscle actin (1A4)	M	Sigma, St Louis, MO	1:1,200	—
CD1a (010)	M	Immunotech, Miami, FL	1:4	Microwave
CD3	R	DAKO	1:500	Enzyme
CD20 (L26)	M	DAKO	1:200	—
CD21 (1F8)	M	DAKO	1:50	Enzyme
CD30 (Ber H2)	M	DAKO	1:100	Enzyme
CD35	M	DAKO	1:100	Enzyme
CD45RB (PD7/26, 2B11)	M	DAKO	1:200	—
CD45RO (UCHL-1)	M	DAKO	1:200	—
CD68 (KP-1)	M	DAKO	1:500	Enzyme
Fascin (55K2)	M	CELL MARQUE, Austin, TX	1:20	Microwave
Desmin (D33)	M	DAKO	1:100	Enzyme
Factor XIII	R	CALBIOCHEM, San Diego, CA	1:800	Enzyme
p53	M	DAKO	1:100	Microwave

M, mouse monoclonal; R, rabbit polyclonal.

Electron microscopy was performed on 3 cases (1-3). Poor fixation precluded ultrastructural analysis of case 4. For case 1, fresh tissue fixed in 2.5% glutaraldehyde was used. For cases 2 and 3, formalin-fixed tissue was removed from the paraffin block, deparaffinized in xylene, and postfixed in glutaraldehyde. All tissue was postfixed in buffered osmium tetroxide, dehydrated in ethanol, and embedded in Maraglas-655 (Ladd Research Industries, Burlington, VT). Ultrathin sections were prepared, stained with uranyl nitrate–lead citrate, and examined using a transmission electron microscope (CM10, Philips).

Case Reports

Case 1

A 61-year-old man sought care because of a 5-month history of a painless, gradually enlarging mass on the posterior right side of the neck. He was examined in the surgery clinic with a presumed diagnosis of lipoma vs epidermal inclusion cyst. His medical history included hypertension, multiple colonic polyps, and aortic valvular stenosis. He had a distant history of tobacco use. The physical examination revealed a 2-cm subcutaneous nodule on the posterior right side of the neck without appreciable regional adenopathy. The mass was nontender and mobile. A 1-cm, mobile, nontender nodule also was identified on the left side of the chest in proximity to the nipple. No other abnormalities were identified.

An excisional biopsy of the neck mass was performed, resulting in an initial diagnosis of malignant spindle cell neoplasm. A chest computed tomography (CT) scan revealed a 4.7-cm suprahilar lung mass, a 9-cm mass along the posterior costophrenic sulcus, and a 1.2-cm subcutaneous nodule in the left side of the chest, and marked mediastinal and subcarinal lymphadenopathy was identified. An abdominal CT scan revealed an enlarged spleen measuring 19 cm. Laboratory studies showed mild anemia and thrombocytopenia, with a hematocrit of 32.6% (0.33) and a platelet count of $82 \times 10^3/\mu L$ ($82 \times 10^9/L$), and an elevated alkaline phosphatase level (284 U/L). Results of all other laboratory studies were within the reference ranges.

The patient underwent wide local excision of the neck mass for therapeutic and diagnostic purposes. The patient received 4 cycles of CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine [Oncovin], and prednisone) without significant response. The patient was alive with persistent disease at 1.5 years.

Case 2

A 70-year-old man was examined because of a slowly growing left axillary mass for 3 months. He had undergone a low anterior resection for colon cancer 6 years earlier, which was complicated by local tumor recurrence. The patient received chemotherapy (leucovorin and 5-fluorouracil) and radiation. There had been no evidence of colon cancer for 2 years at the last follow-up for that diagnosis.

Physical examination revealed that the left axillary mass was soft, mobile, and nontender, measuring 6.5 cm in greatest dimension. After a diagnosis of interdigitating dendritic cell sarcoma (IDCS), further radiologic and clinical workup did not identify any other sites of disease, and the

patient opted for no further therapy. The patient was alive and free of disease at 9 months.

Case 3

A 77-year-old woman sought care because of unilateral right tonsillar enlargement. She had a 1-year history of stage IV B-cell small lymphocytic lymphoma and had completed 6 cycles of chemotherapy (cisplatin, vincristine, prednisone) with regression of disease. The patient was otherwise without complaints, except for a stable postchemotherapy distal neuropathy. Excision of the affected tonsil yielded a 2.5 × 1.8 × 0.6-cm tonsil with a tan, polypoid mass measuring 1.5 × 1.2 × 0.5 cm. No other sites of disease were identified. After the diagnosis of IDCS, the patient was treated with local radiation and was free of disease at 5 months of follow-up.

Case 4

A 73-year-old woman without clinically significant medical history findings was examined because of an enlarged right inguinal lymph node. She denied systemic symptoms. Excision of the enlarged lymph node revealed a 3.5 × 3.0 × 1.8-cm node with central necrosis. She refused additional therapy and died of widespread disease 2 months after diagnosis.

Results

Macroscopically, the neoplasms were firm, relatively circumscribed, and white to tan. Tumor size ranged from 1.5 to 6.0 cm. Necrosis was identified in 1 case (case 4).

Microscopically, lymph nodes (cases 1, 2, and 4) were diffusely effaced by a proliferation of medium to large spindle-shaped cells with a fascicular or whorled growth pattern **Image 1** and **Image 2**. The individual neoplastic cells had round to ovoid, vesicular nuclei, often with deeply cleaved, irregular nuclear membranes **Image 3**. Nucleoli were generally prominent. Case 3, involving the tonsil, showed ulceration of the overlying epithelium and infiltrates of discohesive large cells with lobated nuclei and abundant eosinophilic cytoplasm **Image 4** and **Image 5**. Multinucleated cells and large pleomorphic cells up to 60 µm were prominent in case 2. Cells with a xanthomatous appearance were also present in case 2. The maximum mitotic rate was 9 mitoses per 10 high-powered fields. In all cases, small lymphocytes and plasma cells were intermixed with neoplastic cells. Collagen deposition was identified primarily between fascicles in all cases and was most prominent in case 1. Eosinophils were not observed in any of the cases.

The immunohistochemical results are summarized in **Table 2**. All cases were positive for S-100, CD68, and lysozyme. CD68 showed a granular cytoplasmic staining pattern and accentuated the dendritic cellular morphologic features **Image 6**, while lysozyme showed focal (case 1) to diffuse (cases 2-4) cytoplasmic staining. Fascin was positive in 3 of 4 cases with diffuse cytoplasmic staining. CD45RB was positive in 3 cases, with the malignant cells showing less intense staining than the background lymphocytes **Image 7**. CD1a showed weak patchy staining in 1 case **Image 8**. Strong nuclear staining was demonstrated for p53 in the neoplastic cells in 3 of 4 cases evaluated (1, 2, and 4). The tumor cells were negative for CD3, CD20 (in the 1 case evaluated), CD21, CD30, cytokeratin, smooth muscle actin,

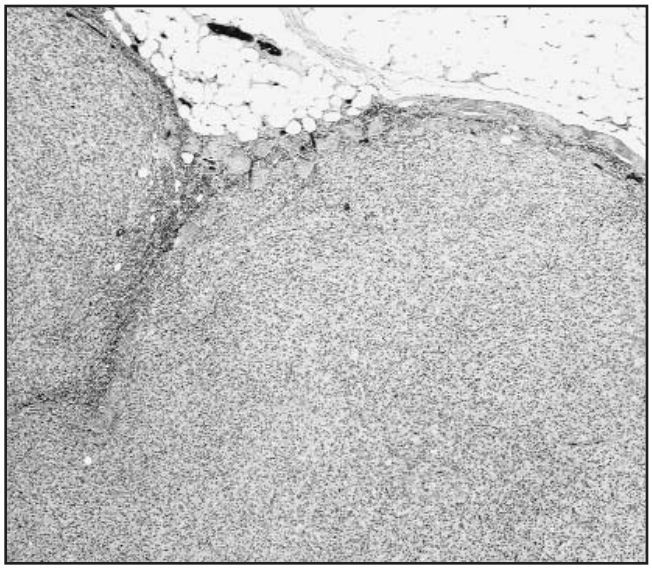

Image 1 Lymph node specimen showing diffuse architectural effacement (H&E, ×37).

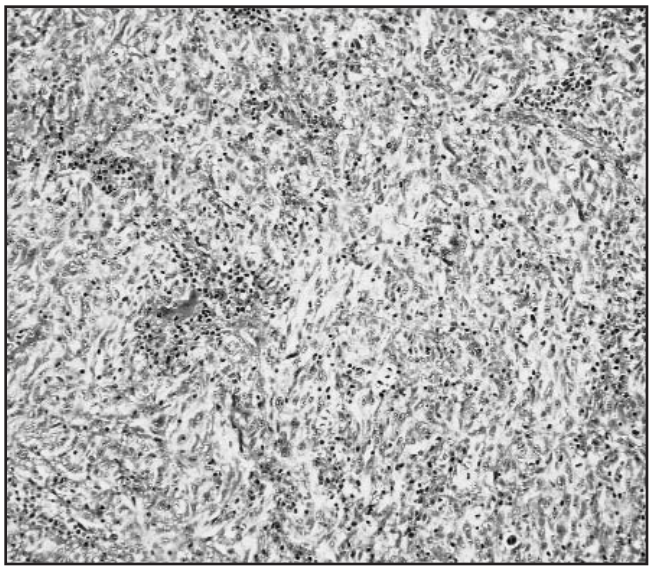

Image 2 Tumor cells arranged in ill-defined whorls (H&E, ×100).

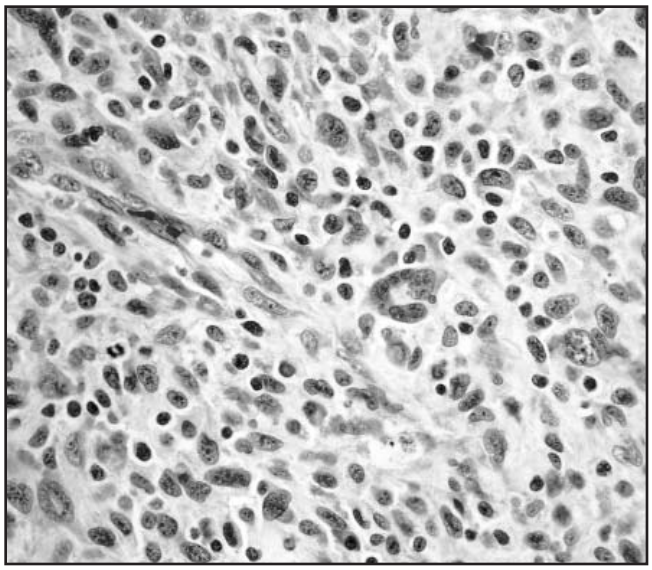

Image 3 Spindle cells with vesicular nuclei and ill-defined cytoplasmic borders. Note scattered multinucleated giant cells (H&E, ×385).

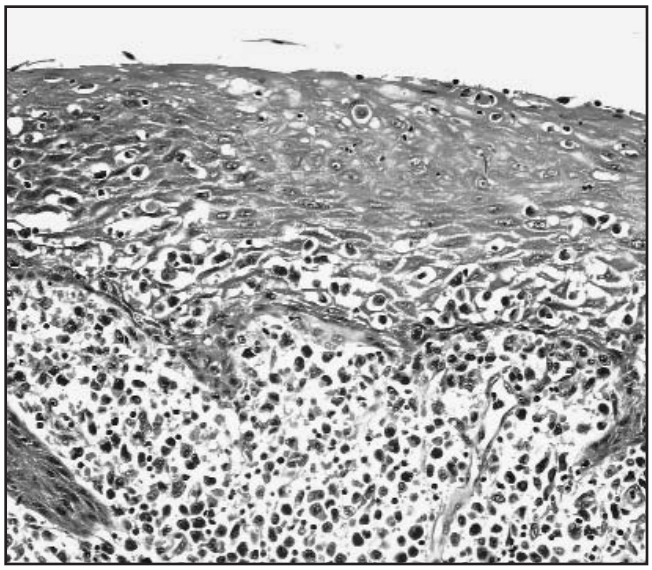

Image 4 Tonsil showing specimen ulceration and dense subepithelial infiltrates (H&E, ×112).

desmin (2 cases evaluated), HMB-45, and factor XIII. In addition, the tumor cells were negative for CD35 in 2 cases evaluated (cases 1 and 3).

Ultrastructural evaluation showed elongated tumor cells with prominent interdigitating processes **Image 9**. The nuclei were indented and irregularly shaped. The majority of tumor cells demonstrated rough endoplasmic reticulum and lysosomes and a sparse number of mitochondria **Image 10**. Occasional cells showed rare cytoplasmic filaments, short strands of basal lamina at the cell periphery, and fusiform densities. No melanosomes or Birbeck granules were identified.

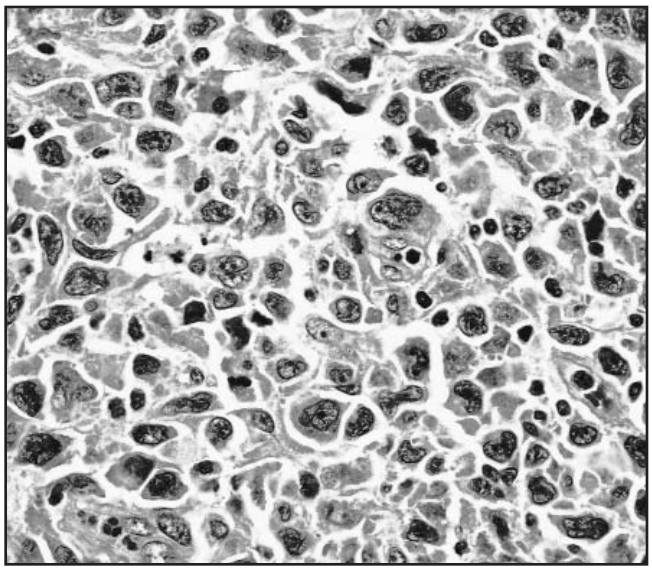

Image 5 Pleomorphic large cells with indented nuclei and abundant cytoplasm (H&E, ×640).

Discussion

The 4 cases presented show clinical and histologic similarity to the 25 previously reported cases of interdigitating dendritic sarcoma **Table 3**. IDCS generally manifests with lymphadenopathy, rarely with associated systemic symptoms including fever and weight loss. A widely affected age range, from 8 to 77 years, has been reported, with a median age of 52 years and a slight male predominance (approximately

Table 2
Immunohistochemical Findings

	Case			
Antibody	**1**	**2**	**3**	**4**
Cytokeratin	–	–	–	–
S-100	+	+	+	+
HMB-45	–	–	–	–
Lysozyme	+	+	+	+
CD1a	–	–	+	–
CD3	–	–	–	–
CD20	ND	–	ND	ND
CD21	–	–	–	–
CD30	–	–	–	–
CD35	–	ND	–	ND
CD45RB (leukocyte common antigen)	+	–	+	+
CD45RO	–	–	ND	–
CD68	+	+	+	+
p53	+	ND	+	+
Fascin	+	+	–	+
Factor XIII	–	–	–	–
Vimentin	+	+	+	+
Smooth muscle actin	–	–	–	–
Desmin	–	ND	ND	–

ND, not done; +, positive; –, negative.

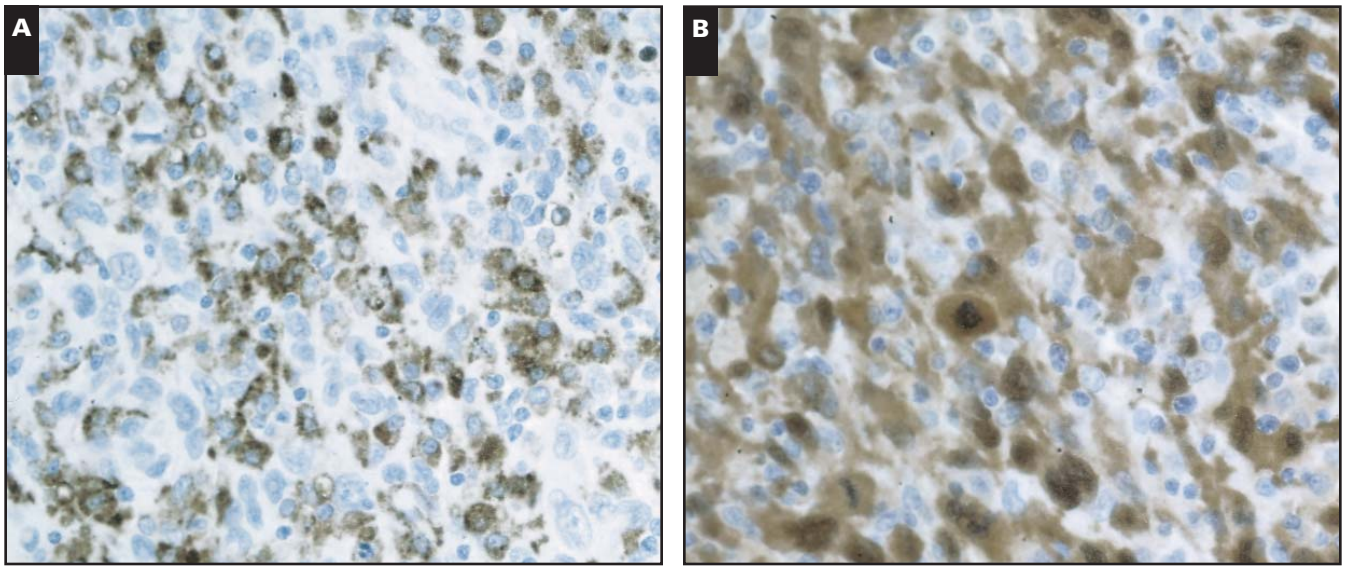

Image 6 Positive staining for S-100 (**A**, ×348) and CD68 (**B**, ×348).

l.5:1). Four cases without documented nodal involvement manifested with mass lesions in the nasopharynx,[12] small intestine and mesentery,[14,17] and testicle.[25] Liver and/or spleen involvement has been described in approximately one third of the cases,[6,12,16,17,21] whereas bone marrow involvement is uncommon.[6,16] Of interest, 2 patients had clinical manifestations consistent with superior vena cava syndrome.[7-9] Similar to case 3 in the present series, a history of previous or concurrent hematologic malignant neoplasm was reported in 6 patients: 1 with mycosis fungoides,[13] 1 with precursor T-lymphoblastic leukemia,[18] 2 with follicle center lymphoma, and 2 with B-cell small lymphocytic lymphoma/chronic lymphocytic leukemia.[20,22]

Histologically, IDCS shows a diffuse proliferation of oval to spindled cells with a variety of described growth patterns, including sinusoidal, nesting, fascicular, and storiform.[1,6-25] The proliferation can partially or completely replace the affected tissue. A collagenous or hyalinized background, with increased reticulin, may be present.[10,16] Microscopically, individual cells have a slender, spindled to plump (histiocytoid) appearance with ill-defined cell borders, abundant eosinophilic cytoplasm, and enlarged indented nuclei. Scattered multinucleated cells are common. Necrosis is unusual. The mitotic rate is variable, but generally moderate, with maximum reported rate of 10 per 10 high-powered fields.[25] A characteristic feature is the presence of lymphocytes and plasma cells sprinkled throughout the neoplasm.[7,8,19,21]

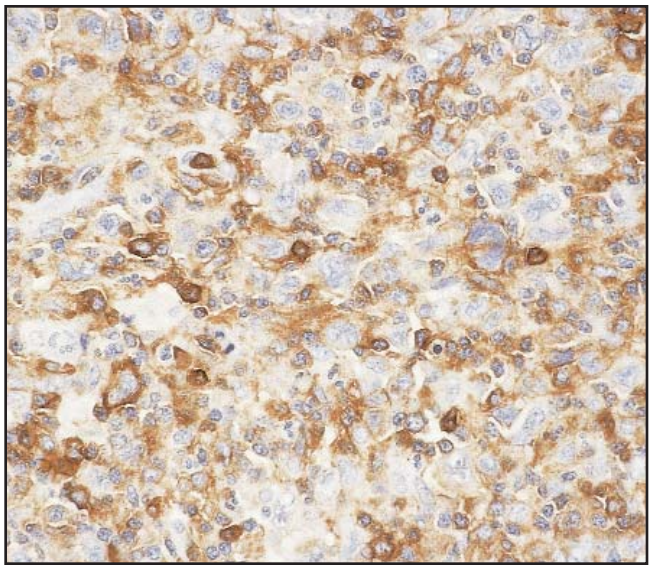

Image 7 Positive staining for CD45RB immunostain (×400).

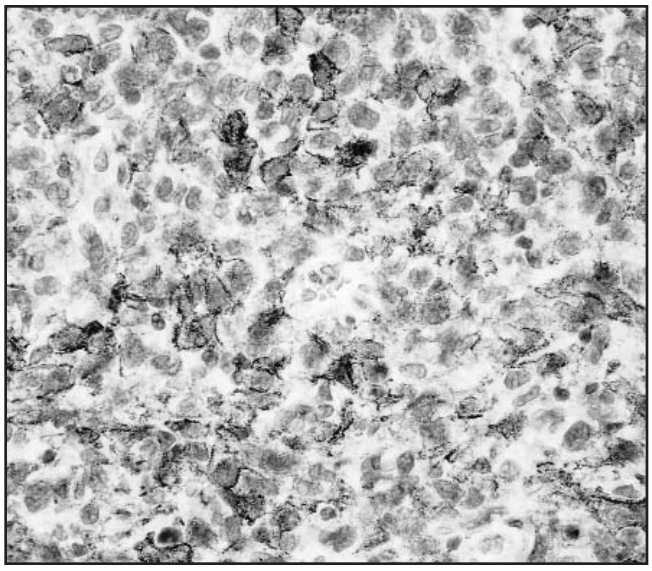

Image 8 (Case 3) Variable immunoreactivity for CD1a (×348).

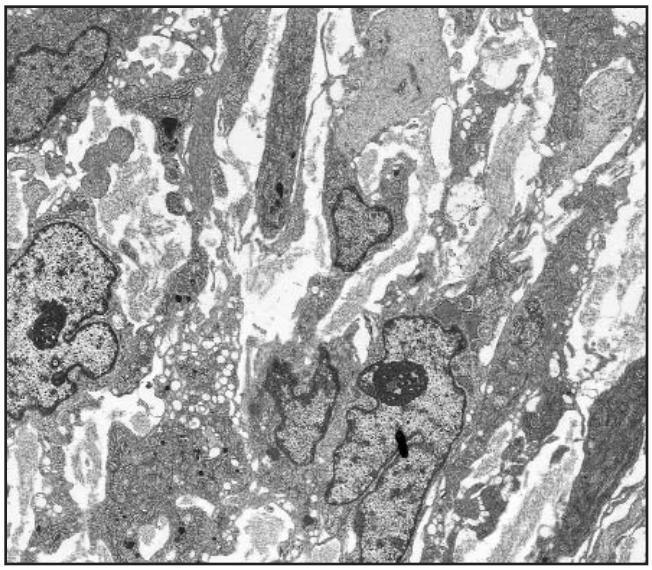

Image 9 The tumor cells exhibit long, slender processes and indented nuclei. The cytoplasm contains well-developed endoplasmic reticulum and, in some cells, a fair number of lysosomes (×2,610).

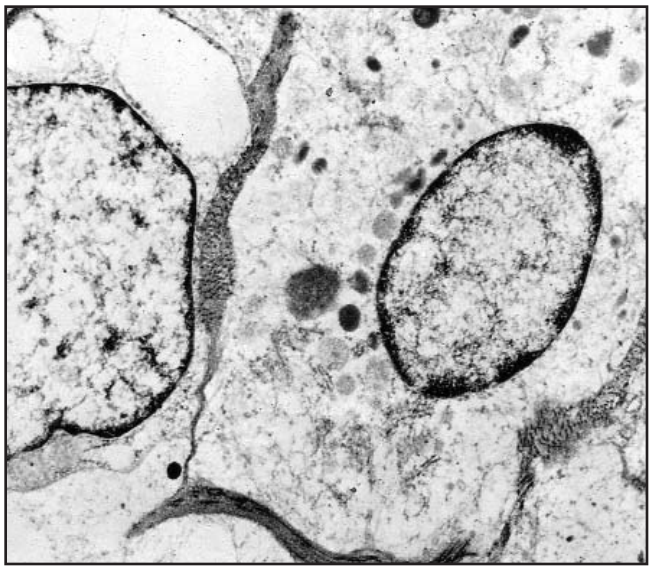

Image 10 Higher magnification of a tumor cell shows cytoplasmic endoplasmic reticulum and multiple lysosomes (×17,186).

IDCS demonstrates a spectrum of morphologic features, as described in the preceding text. Architecturally and cytologically, the neoplastic proliferation can range from dense, whorled spindle cells to haphazardly arranged, plump, histiocytoid cells. These patterns can be admixed within a single tumor. The described cases illustrate this gamut of morphologic features, with cases 1, 2, and 4 showing primarily spindled morphologic features and case 3 demonstrating randomly arrayed plump, histiocytoid cells. Although the number of reported cases is limited, histologic features do not seem to predict survival accurately. For example, necrosis, a prominent feature in case 4, also has been described in 4 other cases of IDCS, generally as small foci or individual cells.[6,7,16,17] Of these cases, 1 patient was alive with disease and 4 had died of disease.

There is no specific marker for IDCS, and the tumor cells demonstrate a heterogeneous immunophenotype. Similar to normal interdigitating dendritic cells, malignant interdigitating dendritic cell proliferations are positive for S-100, vimentin, HLA-DR, and CD68. Variable staining has been reported for CD1a, CD11c, CD45RB, CD45RO, CD4, and CD14.[7] Two reported cases demonstrated positivity for epithelial membrane antigen,[16] whereas other epithelial markers have been negative. IDCS is negative for CD21, CD35, CD3, and CD20. Fascin, an actin-bundling protein expressed in follicular and interdigitating dendritic cells, was positive in 3 cases and may represent a useful immunohistochemical adjunct in the diagnosis of IDCS.[28,29] Strong, diffuse nuclear staining with p53 was present in the 3 tested cases. Similarly, p53 overexpression has been reported in a high percentage of hematologic malignant neoplasms, with increased staining positively correlated with grade.[30,31]

The variable immunophenotype may reflect the multiple pathways that have been postulated in normal dendritic cell development. The first involves epidermal Langerhans cells, which arise from CD34+ bone marrow progenitors following cytokine stimulation and migrate to the T-cell areas of lymph nodes. The second pathway produces myeloid dendritic cells, which also are derived from the bone marrow. They transiently express CD14, as well as various myeloid and macrophage antigens, and have the potential to develop into macrophages or dendritic cells. The third pathway involves lymphoid dendritic cells, which are present in the thymus and share a common progenitor with T cells. They are CD4+, CD11– and lack myelomonocytic antigens. With cytokine stimulation, they also can develop into typical dendritic cells.[32-34]

Ultrastructurally, IDCS shows indented nuclei with dispersed, peripherally condensed chromatin. The cytoplasm shows numerous elongated interdigitating cytoplasmic processes.[1,6,10,16,17,19-21] Generally, organelles are sparse, with limited mitochondria, endoplasmic reticulum, and occasional lysosomes. Golgi complexes were prominent in several reports,[10,17] as were focal elaborate complexes of tubules and vesicles.[10,19] The presence of junctional complexes and desmosomes has been reported variably, but they are generally absent.[6,7,10,12,15,16,19,21,22,25] Unlike Langerhans cell histiocytosis, IDCS lacks Birbeck granules.[1,35] Interestingly, occasional cells in case 1 demonstrated rare cytoplasmic filaments and basal lamina, a finding associated

Table 3
Present and 25 Reported Cases of Interdigitating Dendritic Cell Sarcoma

Authors	Patient Sex/Age (y)	Sites of Involvement	Treatment	Follow-up
Lennert and Mohri[13]	F/69	Lymph nodes	Not reported	Not reported
van Heerde et al[8] and Feltkamp et al[9]	M/37	Mediastinum; cervical and axillary lymph nodes; skin	Chemotherapy (C, V, P); radiation	DOD, 4 mo
Turner et al[12]	M/17	Lymph nodes; spleen; liver	Chemotherapy (D, B, V)	DOD, 4 mo
	M/30	Nasopharynx	Radiation	AWD, 20 mo
Daum et al[14]	M/43	Jejunum; mesentery	Chemotherapy (P, V, MX, D, C, ET, MC, PH); surgery	FOD, 5 mo (died of myocardial infarction)
Salisbury et al[15]	F/41	Diffuse lymphadenopathy	Chemotherapy (M, B, DX, C, V, CI); autologous bone marrow transplant	AWD, 6 mo
Chan and Zaatari[21]	M/67	Diffuse lymphadenopathy; spleen; liver; lungs	Chemotherapy (B, D, C, V)	DOD, 1 wk
van den Oord et al[10]	F/74	Axillary lymph node; mediastinum	Radiation	FOD (follow-up time not reported)
Nakamura et al[11]	M/58	Cervical, mesenteric lymph nodes; jejunum	Chemotherapy (C, ME, PR, D, V, MX, P); radiation; surgery	Alive, 5 y
Rabkin et al[16]	M/17	Diffuse lymphadenopathy; spleen; bone marrow	Chemotherapy (MX, D, C, V, B); surgery	DOD, 9 mo
	F/13	Diffuse lymphadenopathy; spleen; mediastinum; ovaries	Chemotherapy (C, V, corticosteroids); radiation	DOD, 7 wk
Weiss et al[6]	M/60	Supraclavicular lymph node	Surgery; radiation	FOD, 1.5 y
	M/34	Inguinal, thoracic, and abdominal lymph nodes; bone marrow; skin; liver; kidney; lung; heart	Chemotherapy; surgery; autologous bone marrow transplant	DOD, 10 mo
Yamakawa et al[19]	M/54	Diffuse lymphadenopathy; pleural effusion	Chemotherapy (C, V, D, P)	AWD, 1 y
Hammar et al[20]	M/67	Cervical lymph node	Surgery	FOD, 4 y
Horschowski et al[18]	M/8	Inguinal, submandibular lymph nodes; colomesenteric mass	Chemotherapy (VIN, CYT, DA, AS, CO)	DOD, 8 mo
Miettinen et al[17]	F/52	Small intestine and abdominal masses; retroperitoneal, mediastinal lymph nodes	Chemotherapy (MX, B, D, C, V, CI, CY, ET); surgery	DOD, 16 mo
	M/58	Ileum; mesentery; liver	Chemotherapy (C, D, V, P); surgery	FOD, 12 mo
Rousselet et al[7]	F/20	Cervical, supraclavicular, and mediastinal lymph nodes	Chemotherapy (MX, V, B, C, P, MI, ET, IF, CA, CYH, ML); radiation	AWD, 1 y
Vasef et al[22]	F/56	Cervical and axillary lymph nodes	P	Not reported
Andriko et al[1]	M/23	Cervical and axillary lymph nodes	Surgery; radiation	AWD, 4 y
	F/32	Cervical lymph node	Surgery	Not reported
Banner et al[24]	F/68	Cecum; regional lymph nodes	Chemotherapy	Not reported
Luk et al[25]	M/74	Right testicle	Surgery	FOD, 9 mo
Hui et al[23]	M/67	Skin	Not reported	AWD, 6 y
Present cases				
1	M/61	Cervical and mediastinal lymph nodes; chest wall; lung; spleen	Chemotherapy (D, V, P, C)	AWD, 1.5 y
2	M/70	Axillary lymph node	Surgery	FOD, 9 mo
3	F/77	Tonsil	Radiation	FOD, 5 mo
4	F/73	Inguinal lymph node; widespread metastases	Surgery	DOD, 2 mo

AS, asparaginase; AWD, alive with disease; B, bleomycin; C, cyclophosphamide; CA, carmustine; CI, cisplatin; CO, cortisone; CY, cytosine arabinoside; CYH, cytarabine hydrochloride; CYT, cytarabine; D, doxorubicin; DA, daunorubicin; DOD, died of disease; DX, dexamethasone; ET, etoposide; FOD, free of disease; IF, ifosfamide; M, methotrexate; MC, mechlorethamine hydrochloride; ME, mercaptopurine; MI, mitoxantrone; ML, melphalan; MX, methotrexate; P, prednisone; PH, procarbazine hydrochloride; PR, predonine; V, vincristine; VIN, vindesine.

with fibroblastic reticular cells,[1] suggesting the possibility of a minor subpopulation of these cells within the sarcoma.

The differential diagnosis of IDCS has been discussed extensively in previous reports.[1,6-26,36] Of the nonhematopoietic spindle cell neoplasms, IDCS may be difficult to distinguish from metastatic melanoma, which can have a similar immunohistochemical profile (S-100 positive, CD68+/–).[37] In the reported cases, positive staining for S-100 and CD68 (4/4), fascin (3/4), and CD45RB (3/4) and the ultrastructural observation of long, intertwining cell processes, coupled with the absence of melanosomes, were helpful in excluding metastatic melanoma. Of the hematopoietic neoplasms, IDCS shares morphologic and immunohistochemical features with other tumors of histiocytic or dendritic origin. Follicular dendritic cell tumors, in particular, have similar morphologic features and can be reliably distinguished only by their consistent immunoreactivity with the complement-related antibodies CD21 and CD35.[35] Langerhans cell sarcoma or malignant histiocytosis X is a rare entity, characterized by atypical and malignant-appearing Langerhans

cells and an aggressive clinical course.[38] Although case 3 in this series was immunoreactive for CD1a, the malignant cells did not show the characteristic cytologic features of Langerhans cells. Furthermore, inflammatory infiltrates, which often accompany Langerhans cell proliferations, were not observed.[39] Finally, rare cases of true histiocytic lymphoma have been described that express S-100 and multiple histiocytic antigens and show morphologic similarity with IDCS.[12]

Based on review of the present and previous cases, the minimal diagnostic criteria for the diagnosis of IDCS are compatible histologic features as described and positive staining for CD45RB, S-100, and CD68. In the absence of CD45RB positivity, ultrastructural examination is essential to confirm the diagnosis of IDCS.

IDCS is an aggressive neoplasm, generally unresponsive to conventional therapy and leading to widespread disease. Of the 24 cases with follow-up, 9 patients died of disease and 7 were alive with persistent or progressive disease after therapy. Eight of the nine patients died within one year of diagnosis. One died at 16 months after diagnosis.[17] Therapy generally consisted of surgical excision of the mass lesion with multiagent, systemic chemotherapy. Radiation therapy, alone or with multiagent chemotherapy, was used in several cases. Compared with follicular dendritic neoplasms, IDCS seems to behave more aggressively.[1,40]

The reported cases document the histologic, immunophenotypic, and ultrastructural features of IDCS and expand the known sites of occurrence. Because IDCSs are rare and can show morphologic and immunohistochemical heterogeneity, correct diagnosis requires a high index of suspicion and complete pathologic study.

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