Received: 9 May 2021 | Revised: 26 August 2021 | Accepted: 6 September 2021

DOI: 10.1002/ccr3.4866

CASE REPORT

Clinical Case Reports Open Access WILEY

A case report of interdigitating dendritic cell sarcoma originating from the oropharynx

Shin-ichi Oikawa[1,2] | **Kiyoto Shiga**[1,2] | **Katsunori Katagiri**[1,2] | **Daisuke Saito**[1,2] | **Yu Ohashi**[2,3] | **Kodai Tsuchida**[1,2] | **Jun Miyaguchi**[1,2] | **Takahiro Kusaka**[1,2]

[1]Department of Head and Neck Surgery, Iwate Medical University, Yahaba, Japan

[2]Head and Neck Cancer Center, Iwate Medical University Hospital, Yahaba, Japan

[3]Department of Oral and Maxillofacial Reconstructive Surgery, Division of Oral and Maxillofacial Surgery, Iwate Medical University, Morioka, Japan

Correspondence
Kiyoto Shiga, Department of Head and Neck Surgery, Iwate Medical University School of Medicine, 2-1-1 Idaidori, Yahaba-cho, Shiwa-gun, 028-3695, Japan.
Email: kshiga@iwate-med.ac.jp

Funding information
None

Abstract

Interdigitating dendritic cell sarcoma is an extremely rare tumor and typically originates from lymph nodes. Here, we report a patient with tumor originated from the oropharynx who received successful surgical treatment.

KEYWORDS

head and neck tumor, interdigitating dendritic cell sarcoma, oropharynx

1 | INTRODUCTION

We report an extremely rare case of interdigitating dendritic cell sarcoma that originated from the oropharynx of a patient who underwent surgery and postoperative radiation therapy. She was initially diagnosed with oropharyngeal cancer and neck metastases. Postoperative histopathological examination revealed that the tumor cells were considered dendritic cell origin.

Interdigitating dendritic cells (IDCs) are the dendritic cells that are distributed in the T lymph areas of lymphoid tissues, such as the lymph nodes, spleen, and thymus, and are the major antigen-presenting cells for T lymphocytes in each area. Interdigitating dendritic cell sarcoma is an extremely rare disease that is considered to be a neoplastic lesion derived from IDCs and occurs in the skin, kidney, lung, testis, and gastrointestinal tract, in addition to in the lymphoid tissue.[1,2] Only approximately 100 cases of this disease have been reported so far. Moreover, it is extremely rare that the tumor, as in this case, originates from the oropharynx. Therefore, we report the case of a patient with IDC sarcoma (IDCS) originating from the oropharynx and present a literature review.

2 | CASE HISTORY AND EXAMINATION

A 65-year-old woman became aware of a right cervical mass in mid-January 2020 and was referred to the

hospital near her residence in mid-February. An elastic hard mass was palpable in her upper right neck, and computed tomography (CT) and magnetic resonance imaging revealed a well-defined solid mass. No systemic symptoms were observed. Fine-needle aspiration cytology did not provide a definitive diagnosis. The patient was then referred to our hospital in May for diagnosis and radical therapy against the tumor. A tumor with an irregular mucosa was found at the lower pole of the right palatine tonsil to the right side of the tongue base. The cervical mass was 42 mm in diameter with a smooth surface, elastic hardness, good mobility, and slight tenderness in the right level II region. CT revealed mild mucosal thickening, with a major axis of 16 mm and contrast enhancement on the right wall of the oropharynx. In the right level IIA region, a swollen lymph node that showed irregular contrast enhancement with a major axis of approximately 54 mm was observed (Figure 1A,B), which was larger than when it was seen on the CT from the previous hospital obtained approximately four months before. Fluorodeoxyglucose positron emission tomography (FDG-PET) imaging revealed a 26 × 12 × 33-mm tumor located on the right wall of the oropharynx, accompanied by abnormal accumulation with a maximum standardized uptake value (SUV_{max}) of 24.6. The mass was suspected to be a cancer. Multiple lymphadenopathy was found in the right level IIA region, accompanied by abnormal accumulation with an SUV of up to 24.7, and lymph node metastasis was suspected (Figure 1C,D). In addition, a mass-like shadow was found in the descending colonic intestinal flexion with abnormal accumulation with an SUV_{max} of 6.4. Colon cancer was suspected, and seven or more swollen lymph nodes were found around the mass and in the mesentery. Abnormal accumulation up to an SUV_{max} of 9.2 was observed in these lymph nodes, and metastasis was considered. Gastroscopy and colonoscopy revealed no obvious double cancer, including within the area of accumulation on FDG-PET.

Right palatine tonsil biopsy[3] revealed non-keratinized stratified squamous epithelium and infiltration of inflammatory cells, which were mainly lymphocytes, under the epithelium. No malignant cells were identified. The cervical lymph node fine-needle aspiration cytology revealed atypical cells with a high N/C ratio, and conspicuous nucleoli were found in agglomerates from solitary form against the necrotic and inflammatory cell background. Although the histological type was unclear, carcinoma was suspected (Figure 2A). Its human papillomavirus status was not examined because of the quality of the aspirate obtained. Concurrently, we assumed that the final pathological diagnosis could be determined following the tumor's surgical resection.

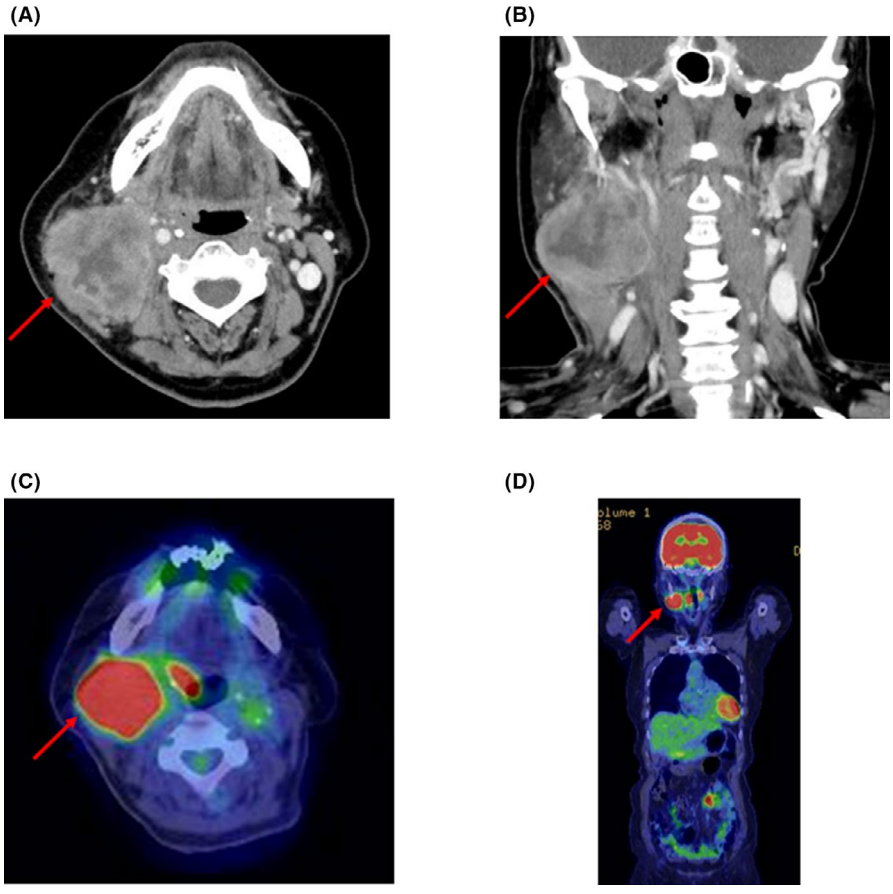

FIGURE 1 A and B Contrast-enhanced computed tomography images of the patient. A: axial image; B: coronal image. In the right level IIA region, a swollen lymph node with a 54-mm diameter that showed irregular contrast enhancement was observed. C and D. Fluorodeoxyglucose positron emission tomography images of the patient. C: axial image; D: coronal image. Multiple lymphadenopathy was seen in the right level IIA region, with abnormal accumulation up to a maximum standardized uptake value of 24.7

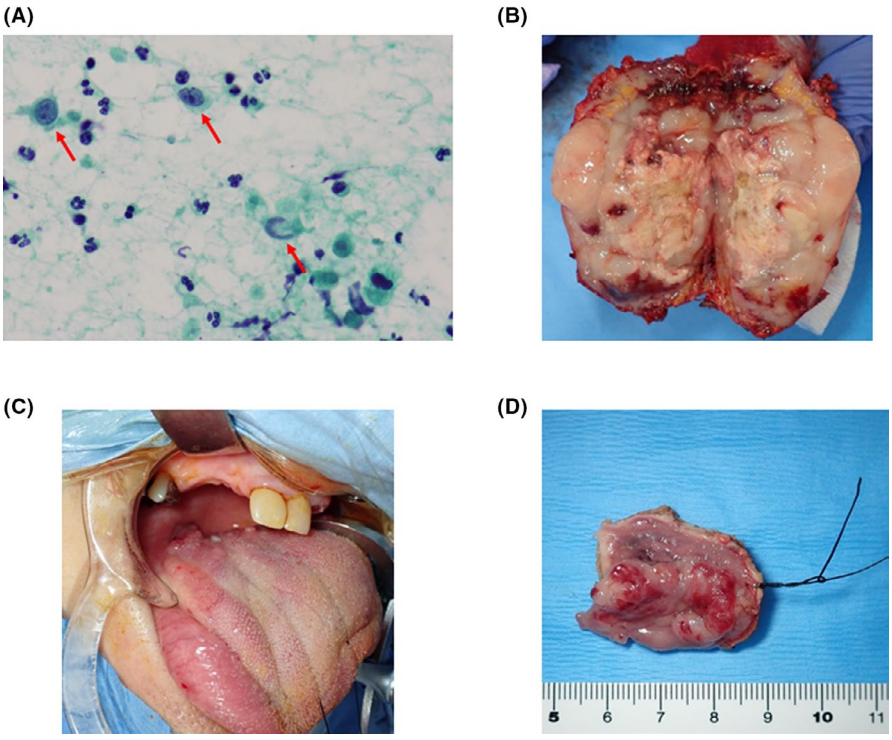

FIGURE 2 Fine-needle aspiration cytology (lymph node) and surgically removed specimens. A, Fine-needle aspiration cytology revealed the presence of atypical cells with prominent nucleoli on a background of necrotic and inflammatory cells. B, Cut surface of specimen. The cervical lymph node removed from the right level II region is shown. C, The operative view of the oropharyngeal tumor. D, The removed oropharyngeal tumor

3 | TREATMENT

As the tentative diagnosis, in this case, was oropharyngeal carcinoma (clinical stage: T1N2bM0), and the pathological diagnosis was only carcinoma of an unidentified type, so we performed pharyngeal tumor resection and neck dissection. Moreover, as there was a possibility that the oropharyngeal lesion was not the primary lesion, and one aim of this surgery was to obtain a definitive pathological diagnosis. Right level II/III lymph nodes had strong adhesion to the surrounding tissues, and a combined resection that included the internal jugular vein, external jugular vein, sternocleidomastoid muscle, great auricular nerve, accessory nerve, and cervical plexus, and posterior belly of the digastric muscle was needed. We used transoral videolaryngoscopic surgery to remove the oropharyngeal lesion.[4] The primary lesion was resected, including the tongue muscular layer and right palatine tonsil, with a margin of approximately 10 mm. The maximum size of the resected lymph nodes was 60 × 50 × 50 mm.

4 | DIFFERENTIAL DIAGNOSIS

Postoperative histopathological examination showed that the oropharyngeal lesion was composed of tumor tissue with ulcers, round nuclei with distinct nucleoli under the epithelium, and mononuclear or multinuclear tumor cells that were proliferating diffusely (Figure 3A). The right level II lymph node showed the same findings as the oropharyngeal lesion and extracapsular spread (ECS) positivity (Figure 3B). Based on these findings, malignant lymphoma and poorly differentiated cancer were included in the differential diagnosis. From the immunohistochemical staining results (Table 1 and Figure 3C–H), the possibility of histiocytic and dendritic cell sarcoma, such as myeloid sarcoma, Langerhans cell sarcoma, and IDCS, was considered. Since the tumor cells were CD4(+), CD1a(focal +), CD45(+), S-100(+), CD68(focal +), CD34(-), MIC2(-), MPO(-), CD21(-), CD23(-), Langerin(-), SMA(-), and D2-40(-), the final diagnosis was IDCS.

5 | OUTCOME AND FOLLOW-UP

Since ECS was observed in the cervical lymph node metastasis, postoperative radiotherapy was performed on the pharynx and whole neck from July to September 2020. No obvious recurrence or metastasis was observed one year and two months after surgery and eleven months after the end of the postoperative radiotherapy.

6 | DISCUSSION

According to the World Health Organization classification (2008), histiocytic and dendritic cell tumors are classified into five types: histiocytic sarcoma, Langerhans cell-derived tumor, IDCS, follicular dendritic cell sarcoma (FDCS), and other rare dendritic cell tumors.[1] Among them, IDCS is an extremely rare tumor, and only slightly over 100 cases have been reported to date. Systemic

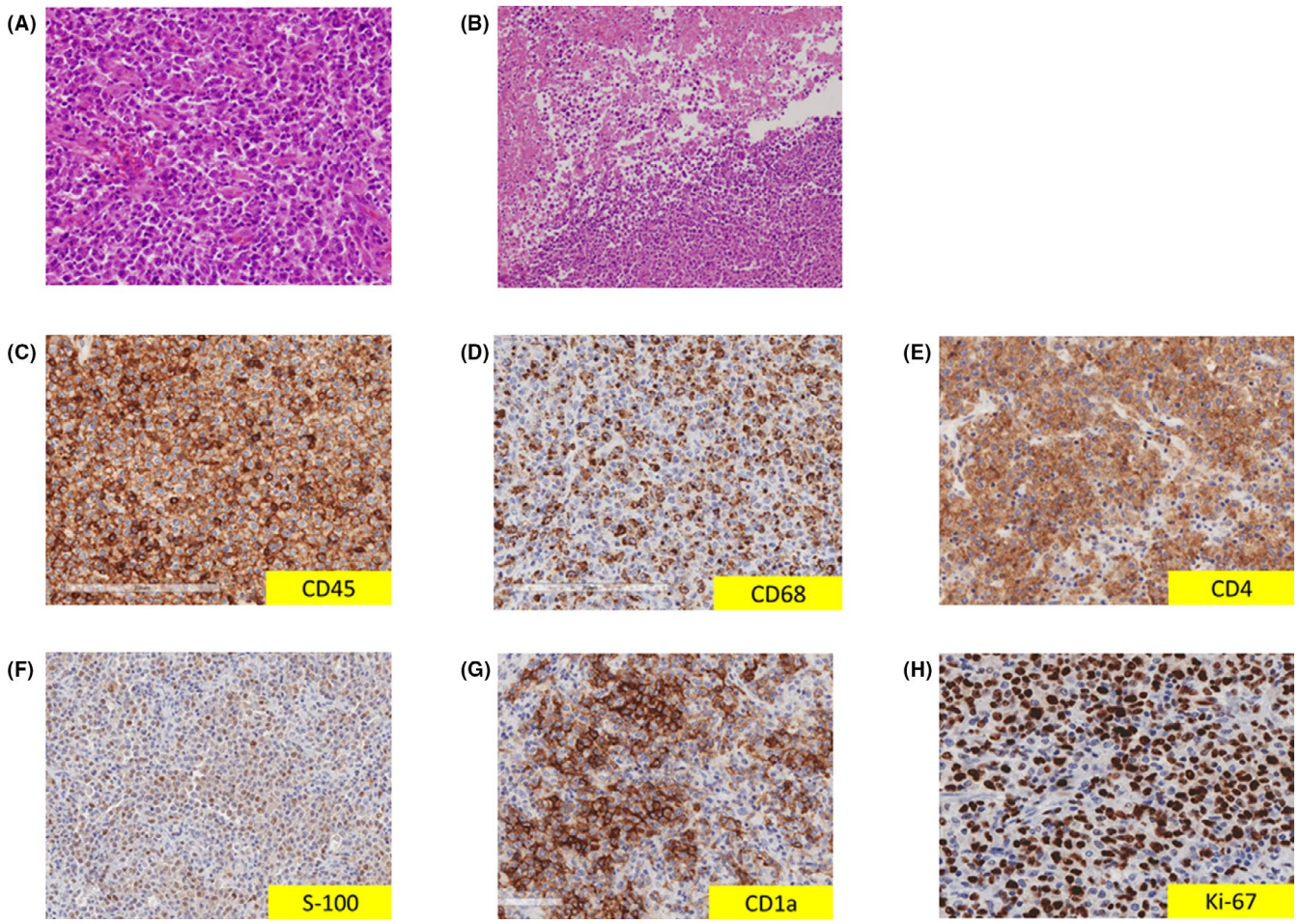

FIGURE 3 Histopathological findings. Hematoxylin-eosin stain of the oropharyngeal tumor (A) and lymph node (B) The two samples were nearly identical. Positive results were observed in the immunohistochemical study for CD45 (C), CD68 (D), CD4 (E), S-100 (F), CD1a (G), and Ki-67 (H) (see also Table 1)

TABLE 1 Immunohistochemistry results

CD3	−	CD34	−
CD10	−	CD45	+
CD20	−	S−100	+
CD30	−	CD68	Focal +
CD56	−	MIC2	−
GranzymeB	−	MPO	−
ALK−1	−	CD4	+
EMA	−	CD21	−
AE1/AE3	−	CD23	−
Ki−67	Positive rate 90%	Langerin	−
EBV-ISH	−	SMA	−
CD1a	Focal +	D2-40	−

symptoms include fever, weight loss, and malaise. Lymph nodes are the most commonly affected site, but in approximately 30% of cases, extranodal lesions in the nasopharynx, small intestine, intestinal membrane, spleen, testicles, skin, tonsils, and bladder, among other regions are observed.[1] Histologically, tumor cells are distributed in the paracortical region of the lymph nodes and show a sheet-like, bundle-like, or flower-like growth pattern. Tumor cells are oval to spindle-shaped and have abundant vesicles, oval nuclei, and distinct nucleoli. Multinucleated cells are also abundant.[5-9] Immunohistologically, tumor cells express macrophage-related antigens, such as S-100 protein, CD68, and CD45. In contrast, CD1a, lysozyme, follicular dendritic cell markers (CD21, CD35), and B cell and T cell markers are negative.[5-9] The desmosome-like focal adhesions normally observed in Birbeck granules, and FDCS, which are specific to Langerhans cell histiocytosis, have not been observed.[7,8,10] Differential diagnoses include histiocyte/dendritic cell tumors, various sarcomas, malignant melanoma, malignant lymphoma, Rosai-Dorfman disease, and undifferentiated cancer. IDCS is characterized by an interdigitation that is complicated and has been found to invade between adjacent cells through numerous cell processes as seen on electron microscopy. Although no electron microscopic images were obtained in this case, the histological findings and

immunohistochemical staining results were generally in agreement with those of IDCS.

According to Jiang et al., the median age of onset of IDCS is 56 years (0.3–86 years), the median's overall survival is 15 months, and disease-free survival and overall 1-year survival are 32% and 51%, respectively.[11] Although a standard treatment for IDCS has not been established, if the lesion is localized, surgical resection, radiation therapy, or a combination of both is performed, and long-term survival has been reported.[2] In addition, CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone), DHAP (dexamethasone, cisplatin, high-dose cytarabine), and EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin) therapy have been used for advanced cases and systemic lesions. ICE (ifosfamide, carboplatin, etoposide) therapy and ABVD (doxorubicin, bleomycin, vinblastine, dacarbazine) therapy for Hodgkin's lymphoma have also been administered with chemotherapy and radiotherapy, similar to treatment for malignant lymphoma.[5] However, it has been reported that CHOP therapy for non-Hodgkin's lymphoma had a poor therapeutic effect, and early recurrence has been observed, even in patients who respond to treatment.[12] However, it has been reported that a therapeutic effect was obtained following the administration of ABVD therapy for Hodgkin's lymphoma,[12,13] and the prognosis remains unclear.

In our case, the lesion was confined to the oropharynx and cervical lymph nodes, and the histological type was unknown before surgery. Thus, resection surgery was selected as the initial treatment. Since ECS was found in the cervical lymph nodes, postoperative radiotherapy was performed. No obvious recurrence or metastasis was observed in one year and two months after surgery, and the obvious increase in lymph node size in the abdominal region that was observed on preoperative FDG-PET was not observed on subsequent contrast-enhanced CT.

Since IDCS often occurs in lymph nodes, as described above, it was difficult to determine whether the primary lesion of this disease was in the oropharynx or cervical lymph nodes. However, since lymph drainage routes from the pharynx to the cervical lymph nodes are present, the oropharyngeal lesions were thought to be the primary pathology.

ACKNOWLEDGEMENTS

None.

CONFLICTS OF INTERESTS

The authors declare that they have no conflicts of interests.

AUTHOR CONTRIBUTIONS

SO and KS contributed to study concept and design and wrote the manuscript. KK, DS, YO, KT, JM, and TK contributed to data acquisition. We also confirm that all authors have read and approved the final version of this manuscript.

ETHICAL APPROVAL

The Iwate Medical University Institutional Review Board exempted ethics approval for case reports.

CONSENT

Full consent for participation and publication was provided by the patient.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ORCID

Kiyoto Shiga https://orcid.org/0000-0002-7461-1352

REFERENCES

1. Pokuri VK, Merzianu M, Gandhi S, et al. Interdigitating dendritic cell sarcoma. *J Natl Compr Canc Netw*. 2015;13:128-132.
2. Saygin C, Uzunaslan D, Ozguroglu M, Senocak M, Tuzuner N. Dendritic cell sarcoma: a pooled analysis including 462 cases with presentation of our case series. *Crit Rev Oncol Hematol*. 2013;88:253-271.
3. Licitra L, Bernier J, Grandi C, Merlano M, Bruzzi P, Lefebvre J-L. Cancer of the oropharynx. *Crit Rev Oncol Hematol*. 2002;41:107-122.
4. Tomifuji M, Araki K, Yamashita T, Shiotani A. Transoral videolaryngoscopic surgery for oropharyngeal, hypopharyngeal, and supraglottic cancer. *Eur Arch Oto-Rhino-Laryngol*. 2014;271:589-597.
5. Seki O, Suzuki C, Sasaki A, et al. A case of interdigitating dendritic cell sarcoma involving skin. *Jpn J Med Technol*. 2015;64:698-704.
6. Miyagi T, Nagasaki A, Shinzato O, Oshima K, Takasu N. Interdigitating dendritic cell sarcoma/tumor-A case report. *Jpn J Cancer Chemother*. 2007;34:469-471.
7. Pileri SA, Grogan TM, Harris NL, et al. Tumours of histocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases. *Histopathology*. 2002;41:1-29.
8. Uluoglu O, Akyürek N, Uner A, et al. Interdigitating dendritic cell tumor with breast and cervical lymphnode involvement: a case report and review of the literature. *Virchows Arch*. 2005;446:546-554.
9. Okazaki S, Maeda K, Saitou H, et al. A case report of dendritic cell tumor (possible interdigitating cell sarcoma). *Jpn J Diagn Pathol*. 2000;17:411-414.
10. Fonseca R, Yamakawa M, Nakamura S, et al. Folicular dendritic cell sarcoma and interdigitating reticulum cell sarcoma: a review. *Am J Hematol*. 1998;59:161-167.
11. Jiang YZ, Dong NZ, Wu DP, Xue SL. Interdigitating dendritic cell sarcoma presenting simultaneously with acute myelomonocytic leukemia: report of a rare case and literature review. *Int J Hematol*. 2013;97:657-666.

12. Olnes MJ, Nicol T, Duncan M, Bohlman M, Erlich R. Interdigitating dendritic cell sarcoma: a rare malignancy responsive to ABVD chemotherapy. *Leuk Lymphoma*. 2002;43:817-821.
13. Lee SY, Lee SR, Chang WJ, et al. Successful treatment of disseminated interdigitating dendritic cell sarcoma with adriamycin, bleomycin, vincristine, and dacarbazine chemotherapy. *Korean J Hematol*. 2012;47:150-153.

How to cite this article: Oikawa S-I, Shiga K, Katagiri K, et al. A case report of interdigitating dendritic cell sarcoma originating from the oropharynx. *Clin Case Rep*. 2021;9:e04866. https://doi.org/10.1002/ccr3.4866