Case Report

# Interdigitating Dendritic Cell Sarcoma Arising in The Cervical Node

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## ABSTRACT

Inter-digitating dendritic cell (IDC) sarcoma is a rare tumor presenting as an asymptomatic slow growing lesion. The median age of occurrence is around 56 years and has a male preponderance. Peripheral lymphadenopathy is the commonest presenting symptom. The clinical course of patients with IDC sarcoma varies from indolent to more aggressive with widespread metastases and death within a year. There is no standardized therapeutic approach for patients with IDC sarcoma. We are presenting a case of IDC sarcoma in a 40 year old male with cervical node and liver involvement. Histologically, there was complete effacement of nodal architecture with tumor cells arranged in sheets composed of spindle cells and epithelioid cells having convoluted nuclei. Immuno- reactivity for S100 and vimentin were observed. Other stains such as CD 1a, CD21, CD3 and CD45 were negative. Histology, Immuno histochemistry and differential diagnosis are discussed in our presentation which would help in arriving at the diagnosis of this rare tumor.

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## Introduction

Inter-digitating dendritic cell (IDC) sarcoma is a rare tumor with a male preponderance. [1, 2] Mean age of occurrence is 56 years. [1, 2] Cervical lymph nodes, nasopharynx, tonsil, salivary glands, skin, chest wall, paraspinal region, liver, spleen, gastrointestinal tract, bladder, and testis and bone marrow are involved with this tumor. Of this peripheral lymphadenopathy[cervical node] is the commonest presenting symptom. [3] Along with the histological findings, Immunohistochemistry with S-100 is essential for the confirmation of diagnosis. We are presenting a case of IDC sarcoma in a forty year old male. He had an indolent course for about $1_{1/2}$ years. The histological picture along with Immuno -histochemistry and differential diagnosis are discussed.

## Case Report

40 year male with the complaints of swelling neck and fever was investigated elsewhere $1_{1/2}$ years back. FNAC was done there and he was diagnosed as having tuberculous lymphadenitis. He had anti tuberculous treatment for three months. As the swelling did not subside, he was referred to our center. Further evaluation was done on him in our center.

On examination the patient had multiple bilateral cervical nodes and supraclavicular nodes, [Fig: 1]. CT scan showed cervical node, supraclavicular nodes, para- aortic nodes and liver involvement. Radiologist reported it as lymphoma. FNAC of the node showed spindle and epithelioid cells in an inflammatory background composed of eosinophils, [Fig: 2]. Provisional diagnosis of spindle cell tumor was made and biopsy was advised. Excision biopsy of the nodes was taken and sent to the Department of Pathology, Madurai Medical College for evaluation. The specimen we received in our department was 1x1cm. All tissues were embedded and examined.

Microscopically, complete effacement of the lymph node architecture with a thick capsule was noted, [Fig 3]. The node was replaced by tumor cells arranged in fascicles, whorls and sheets. Individual cells were spindle to ovoid with abundant eosinophilic cytoplasm. The nucleus shows indentation, dispersed chromatin and prominent nucleoli. There were neither necrosis nor mitosis. Lymphocytes and eosinophilic infiltrates were seen among the tumor tissue, [Fig: 4 and 5]. He was diagnosed as dendritic cell neoplasm and immunohistochemical study was done for confirmation and categorization. The tumor cells showed diffuse S100 and vimentin positivity,[Fig: 6 and 7]. Other markers like CD45, CD1a, and CD21,CD3 were negative. That clinched the diagnosis of Inter digital dendritic cell sarcoma.

## Discussion

Dendritic cells[DCs] are the key antigen-presenting cells. Four types of DCs are seen in the lymph nodes. They are follicular, interdigitating, Langerhans, and fibroblastic.[4] Interdigitating cells are distributed in the paracortex and deep cortex of lymph nodes, periarteriolar lymphoid sheaths in spleen and interfollicular areas of mucosa associated lymphoid tissue.[1] Neoplasms arising from these DCs as such are rare and constitute about 1% of all lymph node tumors.[2] Of that Interdigitating dendritic cell sarcoma is a rare neoplasm with only around 100 cases reported in the literature.[1] Median age at diagnosis is 56years with a male predilection.[1,2,3]. An ethnic predilection among Asians was noted. [1]

Peripheral nodes like cervical and axillary nodes are most commonly involved.[1] Extranodal sites involved by this tumor include nasopharynx, tonsil, salivary glands, skin, chest wall, paraspinal region, liver, spleen, gastrointestinal tract, bladder, and testis [5]. In 20% of the cases, bone marrow involvement is seen. [6]Usually asymptomatic, B-type symptoms like fever, weight loss, and night sweats can occur in about 20% of the cases. [6]

Grossly the size of the tumor varies from 1-6cm. It is lobulated and has a solid cut surface. Foci of necrosis and haemorrhage can also be seen. Microscopically the tumor either partially or completely effaces the node architecture. In partial involvement the tumor occupies the paracortical areas. The neoplastic cells are arranged in sheets, whorled, storiform, or nests and a mixture of patterns. Among them, mixed patterns are more common. Individual cells have indistinct cell borders and are spindle-shaped or epithelioid. [7] They have abundant eosinophilic cytoplasm and elongated or oval nuclei with irregular or folded nuclear contours. Nucleoli can be inconspicuous or prominent. Multinucleated giant cells are common. [7] Atypia is mild

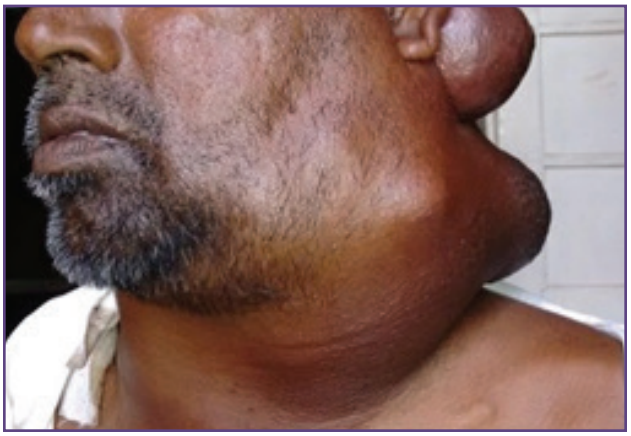


**Fig. 1: 40 years male with multiple cervical and supraclavicular nodes**

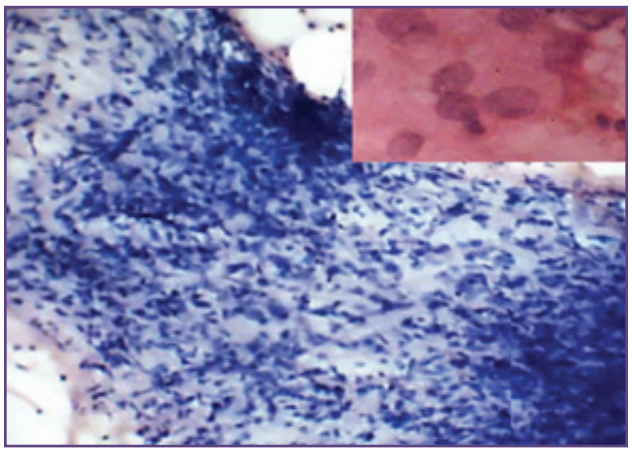


**Fig. 2: FNAC of the lesion show tumor cells arranged in fascicles(H&E 10x) .Inset shows tumor cells with eosinophilic cytoplasm and oval to indented nuclei and eosinophils in the background.(H&E 40x)**

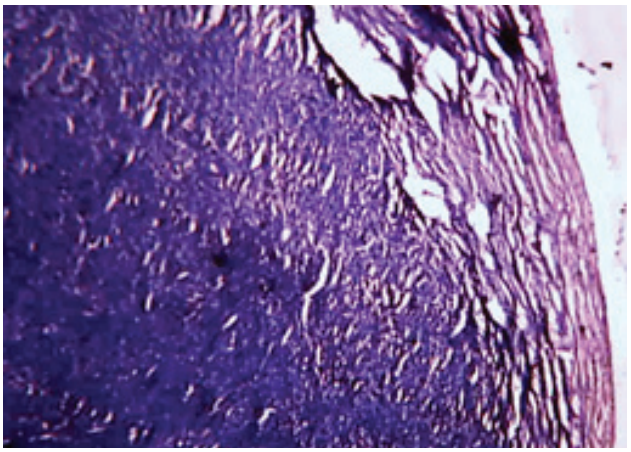
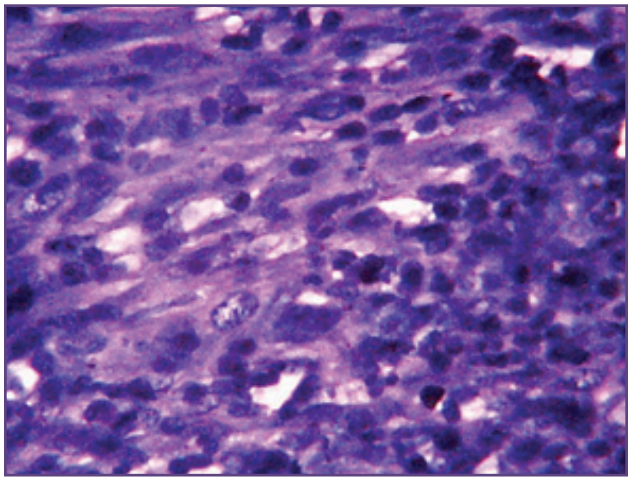


**Fig. 3: Shows complete effacement of lymph node architecture with a thick capsule,[4x H&E]**



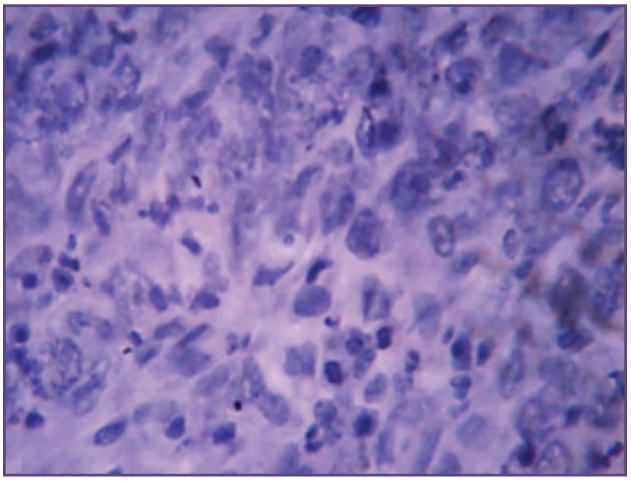


**Fig. 4and5: Show tumor cells arranged in fascicles and whorls. Individual cells show oval and indented nuclei in an inflammatory background composed of eosinophils.(H&E 40x).**

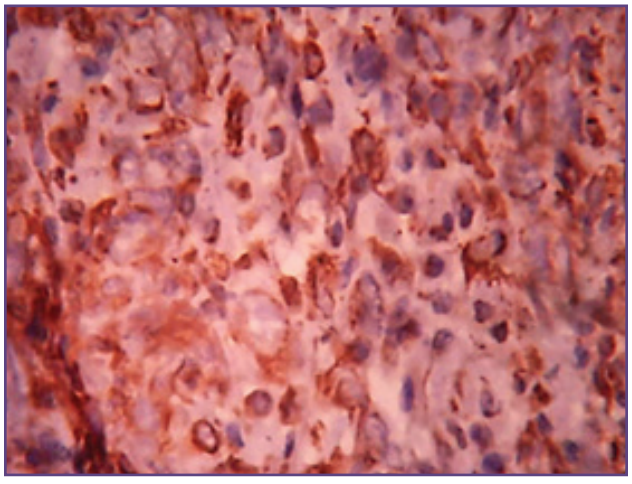


**Fig. 6: Shows diffuse S100 positivity**

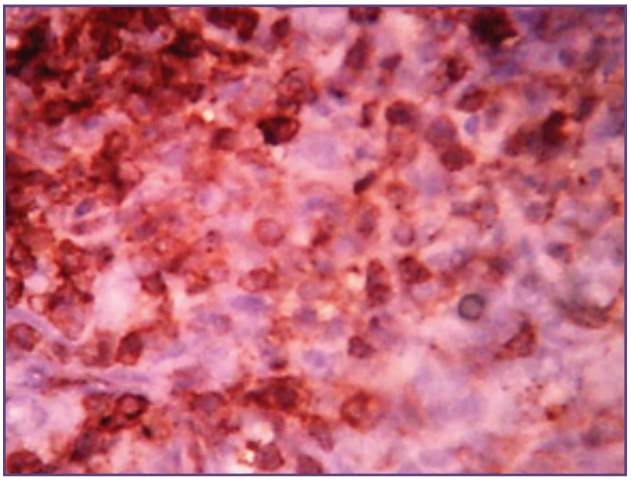


**Fig. 7: Shows Vimentin positivity.**

to marked. The mitotic rate is usually low in this tumor. It is associated with fibrosis and inflammatory cells like small lymphocytes, eosinophils and plasma cells. Usually erythrophagocytosis can be seen focally.

Immunohistologically[IHC], strong S100 and vimentin positivity is characteristic of this lesion. CD11c, HLA-DR, and fascin are the other markers that are positive. Leukocyte common antigen CD45 is focally positive. [8] The histo-morphological differential diagnosis includes Histiocytic sarcoma, Langerhans cell sarcoma, Metastatic melanoma and Follicular dendritic cell sarcoma.

In Histiocytic sarcoma, neoplastic cells have abundant eosinophilic to vacuolated cytoplasm admixed with giant cells having wreath-like nuclei leading on to a xanthomatous appearance. Immunohistochemically, CD11c, CD4, CD14, CD68 and lysozyme are strongly positive in Histiocytic sarcoma. Mixed spindle and epithelioid pattern seen in our case and IHC studies helped to rule out Histiocytic sarcoma.

In Langerhans cell sarcoma [LCS], cells have eosinophilic cytoplasm, twisted and grooved nuclei. CD1a, S100 protein and langerin are positive. Grooved nucleus was not seen in our case and absent CD1a marker helped to rule out LCS.

Metastatic melanoma cells are more epithelioid, have eosinophilic nuclei and show greater pleomorphism than IDC sarcoma. They also express S100 protein and CD68 like IDC. HMB45 and Melan A expression are more specific for melanoma. Both the cytological feature and the marker studies ruled out melanoma in our case.

In Follicular dendritic cell [FDC] sarcoma, oval to spindle-shaped cells are arranged in sheets, fascicles, whorls, storiform, trabecular, or nodular arrangement. In differentiating it from IDC sarcoma Immunohistochemistry is more useful as there are morphological similarities between these two lesions. [9] Strong S100 positivity and vimentin positivity is seen in IDC sarcoma. In FDC sarcoma, clusterin, CD21, CD23, and CD35 are positive. [10]

The immunohistochemical algorithm that was followed by us: As our case was a primary spindle cell neoplasm of the node, marker S100 was tested on the node. S100 was positive, pointing out to four possibilities- IDC sarcoma, Langhans cell sarcoma, Follicular dendritic sarcoma and Melanoma. Negative CD1a ruled out LCS. CD21 and CD 35 were negative and FDC sarcoma was ruled out. Negative Melan A marker ruled out melanoma. Diffuse S100 and vimentin positivity, negative CD1a, CD21 and the histomorphological patterns were in favor of Interdigitating dendritic cell sarcoma, hence the final diagnosis of IDC sarcoma was made.

The clinical course of patients with IDC sarcoma is variable. The course may be benign or aggressive with widespread metastases and death within a year, which is observed in 40% to 50% of patients. Patients with IDC sarcoma develop second neoplasms, like T- or B-cell lymphomas or leukemias or carcinomas. Young age, abdominal involvement, extranodal involvement combined with nodal involvement is associated with a grave prognosis. Our case is a relatively younger male with both nodal and extranodal involvement precluding a graver course.

At present no treatment modality is available for IDC sarcoma. [11] Local disease is treated with surgery. Chemotherapy includes the regimen that is used for lymphoma. More research is needed in this arena.

## Conclusion

In conclusion, Interdigitating dendritic cell sarcoma is a rare tumor of dendritic cells. The disease course can be indolent to aggressive. So far no therapeutic options are available. And there is a 15% risk of developing secondary tumors in these patients. That warrants a careful follow up in these patients. This case is presented to highlight the fact that more research is needed in the treatment aspect of this rare neoplasm.

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## Competing Interests

None

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