Case Report

Aggressive Disseminated Non-Hodgkin's Lymphoma with Involvement of Rare Extranodal Sites- A Case Report

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A B S T R A C T

Introduction: Lymphoma is a type of lymphoproliferative disorder characterized by malignant proliferation of lymphomatous cells and can be broadly categorized into Hodgkin's and Non-Hodgkin's lymphomas. They can involve both nodal and extranodal sites in the body. Although, most common form of lymphoma is lymphnodal type, the involvement of extranodal structures can be seen in 24-39% of cases.

Case report: We report such a rare case of young male with disseminated Non-Hodgkin's lymphoma with extranodal infiltration into the subcutaneous tissues, vertebrae and both testes.

Conclusion: Extranodal infiltration is more commonly seen in Non-Hodgkin's lymphoma and can involve almost any organ in body. The commonly involved extranodal sites include GIT, waldeyer's ring, liver, and lungs. The extranodal involvement of skin and testis may also occur, although very rare.

Keywords: Lymphoma; Non-Hodgkin's; Testis; Extranodal; Skin

INTRODUCTION

Lymphoproliferative disorders are type of hematologic malignancies that include Hodgkin's disease (HD), Non-Hodgkin's lymphomas (NHL), plasma cell myeloma & lymphocytic leukemias.[1] Lymphoma is a malignant proliferation of lymphomatous cells and primarily affects lymphnodes with further infiltration into other primary lymphatic organs which include spleen, thymus and bone marrow cavity. Lymphoma is seventh most commonly reported malignancy in both genders. Hodgkin's & Non-Hodgkin's lymphomas can involve both nodal and extranodal sites in the body, although nodal form is the most commonly reported.[2,3]

Infiltration of malignant lymphomatous cells in the organs other than lymph nodes and primary lymphatic structures is known as extranodal lymphoma. Extranodal involvement is more frequently seen in Non-Hodgkin's lymphomas than in Hodgkin's disease. Almost 24 to 39% of lymphomas may show extranodal infiltration which can affect any organ in the body. Extranodal involvement is characterized by distinct radiologic manifestations and it is always challenging to differentiate primary extranodal disease from the disseminated nodal diseases with extranodal infiltration. The most commonly affected extranodal sites are GI tract followed by Waldeyer's ring, lung, liver, bone. Very rarely lymphoma may spread to skin.[1-3]

The exact etiopathogenesis of lymphoma is still elusive and majority of lymphomas are thus idiopathic. Few lymphomatogenic factors such as bacteria (H. pylori) and viruses (EBV, HCV, HTLV-1, HIV, Herpes virus) have been implicated, although only in certain types of lymphoma. Other possible risk factors include chronic immunosuppression (especially due to drugs after organ transplantation), certain substance exposure like dioxin and agricultural pesticides or a history of chemotherapy (alkylating agents).[2,4]

In this report, we present a case of abdominal lymphoma in a young male patient with spread of lymphoma into chest and abdominal wall and also in bilateral testis, which are reported as very rare sites of extra nodal involvement.

CASE REPORT

A 27 year old male patient presented to routine OPD with

complaints of multiple painless swellings all over the body since 4 months which were gradually increasing in size. He also complained of dull abdominal pain and fullness with associated loss of appetite and significant weight loss. From last one-month, patient also developed low backache which was severe and progressive in nature. There was no significant past history. On clinical examination, multiple firm painless nodular swellings were noted in neck, chest, abdomen, and thighs mainly in the superficial planes. All Laboratory investigations were unremarkable.

Based on clinical findings, patient was referred to Radiology department for further evaluation on Ultrasonography (USG) and contrast enhanced CT (CECT). USG neck revealed multiple discrete and enlarged bilateral cervical lymph nodes (Fig 1). USG and CECT abdomen revealed mild hepato-splenomegaly with multiple discrete to conglomerated enlarged mesenteric and retroperitoneal lymph nodes with some of them showing intranodal necrosis. The largest size node measured upto 5.3x 4.3 cm in right iliac fossa region. Multiple discrete solid nodules (suggesting metastatic deposits) with some showing internal necrosis were noted in the subcutaneous tissues of chest and abdominal wall with associated subcutaneous oedema (Fig 2 & 3). CT, in addition, also revealed multiple lytic lesions in the vertebral bodies (suggesting bony metastasis). USG of inguino-scrotal region revealed sharply demarcated highly

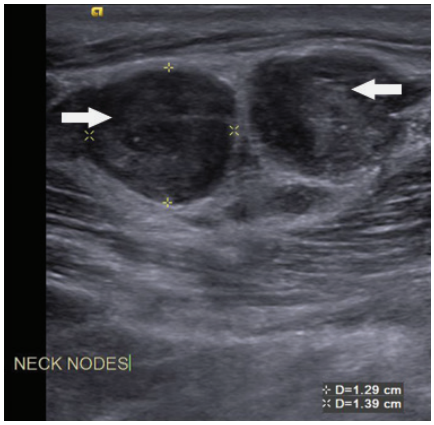

Figure-1: USG of neck shows multiple discrete round to oval non necrotic enlarged (short axis diameter upto 1.3cm) lymph nodes (white arrows)

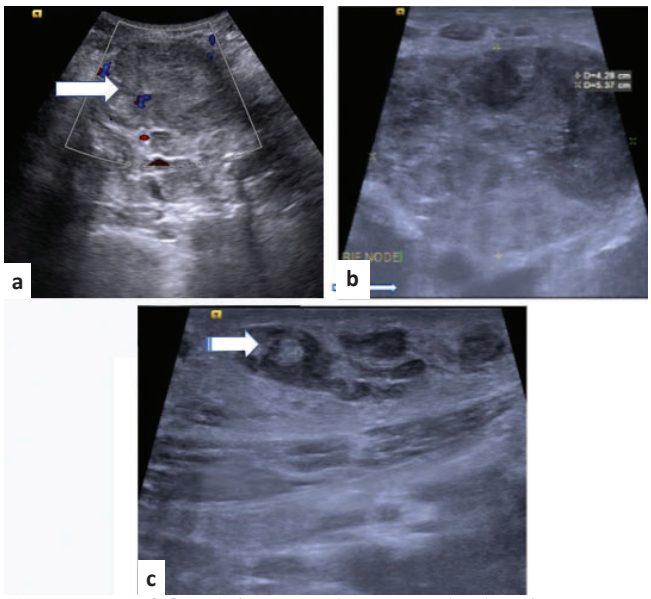

Figure-2: USG abdomen shows multiple discrete to conglomerated, round to oval enlarged mesenteric and retroperitoneal lymph nodes (thick arrow in a) with some of them showing intranodal necrosis. The largest size node measured upto 5.3x 4.3 cm in right iliac fossa region (thin arrow in b). Multiple similar nodes/deposits are also visible in subcutaneous tissue plane of anterior abdominal wall (striped arrow in c)

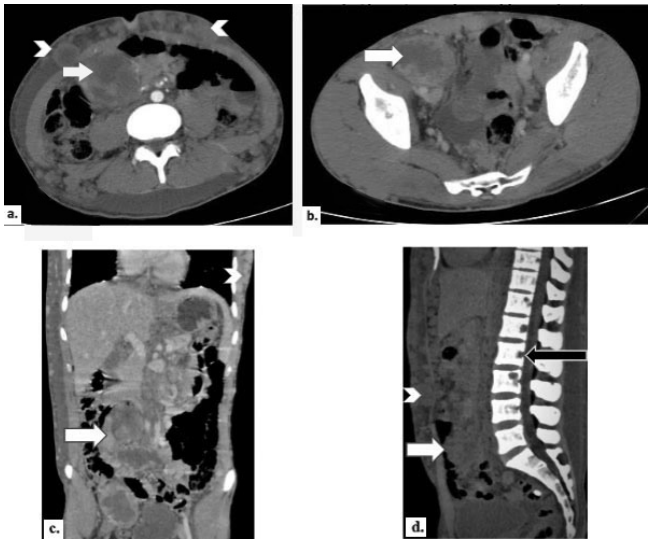

Figure-3: CECT axial (a,b), coronal (c) and sagittal (d) sections of abdomen shows multiple enlarged heterogeneously enhancing necrotic mesenteric and retroperitoneal lymph nodes within the abdomino-pelvic cavity (thick white arrows) with mild pelvic ascites. Multiple solid metastatic deposits with some showing internal necrosis are also noted in the subcutaneous tissue plane of abdominal wall and visualized lower chest wall (white arrowhead) with associated subcutaneous oedema. Visualized sections of spine show multiple focal lytic lesions in the vertebral bodies suggestive of bony metastasis (long black arrow in d).

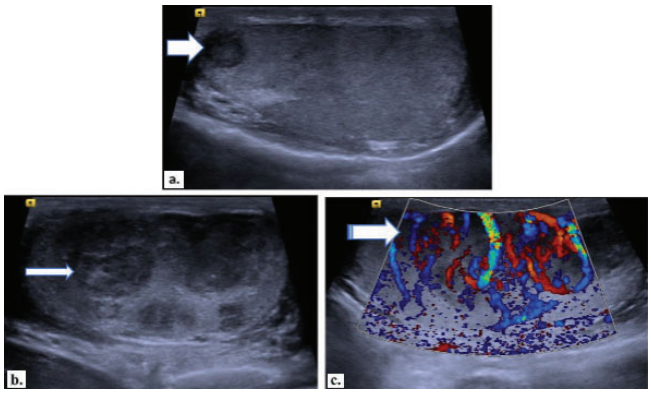

Figure-4: USG scrotum shows a solid well-defined hypoechoic lesion in the right testis (thick arrow in a) and similar multiple lesions in left testis (thin arrow in b) with markedly increased central and peripheral flow on color doppler (striped arrow in c).

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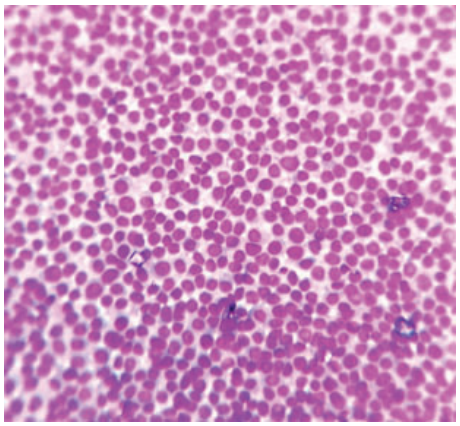

Figure-5: Photomicrograph showing cellular smear exhibiting singly dispersed monomorphic population of malignant cells having moderate cytoplasm with vacuolation, hyperchromatic nuclei about five times the size of RBC and noticeable nucleoli.

vascular hypoechoic lesions in both testis with mild reactive hydrocele and multiple enlarged bilateral inguinal nodes (Fig 4).

FNAC done from the cervical lymph node, chest wall and abdominal wall swellings showed cellular smears exhibiting singly dispersed monomorphic population of malignant cells having moderate amount of cytoplasm showing vacuolations, nuclei about five times the size of RBC, coarse chromatin and noticeable nucleoli. Few binucleate cells along with mitotic figures were also present. The cytomorphology suggested Non-Hodgkin's lymphoma (Fig 5).

DISCUSSION

Approximately 5% to 6% of all malignancies are comprised of Hodgkin disease's (HD), Non-Hodgkin's lymphoma (NHL) and malignant lymphomas.[2] Non-Hodgkin's lymphomas are a group of heterogenous malignancies which involve lymph nodes and lymphoid organs, along with extra-nodal organs and tissues.[3] Lymphoma strictly confined to lymph nodes and primary lymphatic organs is considered nodal disease while those infiltrating into other organs are termed extranodal.[4] Liver, soft tissue, dura, bone, stomach, intestine and bone marrow are common primary extra nodal sites.[5] Involvement of spleen is regarded as nodal disease in cases of Hodgkin's lymphoma but in NHL it is considered as extranodal involvement.[3] Apart from the lymph nodes adjacent to the primary organ involved, if there is involvement of any other lymph nodes or more than one extranodal site is involved then it is classified as secondary extranodal lymphoma.[1]

Lymphomatous involvement can be unifocal (restricted to single site), multifocal (involving >1 site) or can be disseminated (diffuse involvement). It can affect isolated lymph nodes or any organ system with wide ranging imaging manifestations.[6] In abdomen, NHL has a broad spectrum of dissemination involving all sites of abdominal lymph nodes with frequent involvement of the mesentery and retroperitoneal regions.[7] In our case also, both mesenteric and retroperitoneal lymph nodal sites were involved.

Normal lymph nodes appear elongated in shape with fatty hilum. Lymphomatous infiltration into lymph nodes are visualized incidentally on cross sectional imaging or during assessment at Ultrasonography (USG). The involved lymph nodes typically appear homogenously hypoechoic, rounded and enlarged in size (>1cm in short axis diameter) on USG.[6] Computed tomography (CT) is the preferred imaging modality to assess shape, size and relation of the lymphomatous lesions to adjoining organs in NHL and HD. It also allows imaging evaluation of almost all anatomical regions of the body. Currently, CT is modality of choice for diagnosis and staging of abdominal lymphomas with the sensitivity of 88%, and specificity of 86%.[8]

On CT, involved lymph nodes are generally discrete, enlarged and show homogeneous attenuation.[6] However, in NHL, the enlarged lymph nodes commonly fuse to form a conglomerated lymph nodal mass which may encase adjacent vessels or bowel loops resulting in 'Vessel imbedded signs' and 'Intestinal imbedded signs.[7] Our patient also showed multiple discrete to conglomerated enlarged lymph nodes with some of them showing internal necrosis.

Cutaneous lymphomas may be primary or secondary to systemic lymphomas. Primary cutaneous lymphomas are classified as tumors that are confined to the layers of skin with no signs of dissemination and remains so for at least 6 months.[9] Cutaneous lymphoma can be seen as focal mass/nodule and diffuse thickening of skin. On USG, they appear as hypo-echoic nodule with well-defined margins, which may fuse, resulting in a polypoidal lesion in the focal type and homogeneous dermal thickening with increased echogenicity or heterogeneous dermal and subcutaneous thickening in the diffuse type. On CT, they present as diffuse dermal thickening or focal cutaneous mass/nodule. In our case, multiple discrete nodules with subcutaneous oedema were seen in cutaneous/subcutaneous regions of thoraco-abdominal wall.[10]

Primary testicular lymphoma constitutes only 5% of all testicular malignancies and nearly 1-2% of all cases of NHL. The involvement of testis as extra nodal site in Non-Hodgkin's lymphoma is very uncommon and is mainly secondary to disseminated diseases. It affects younger patients and appears as well defined unifocal or multifocal hypoechoic lesions on USG.[11] In our case, we found multiple hypoechoic lesions in the both testis with markedly increased flow on color doppler.

Non-Hodgkin's lymphomas primarily originate in extra skeletal sites with skeletal involvement commonly occurring after hematogenous spread or direct spread from adjacent involved lymph nodes or soft tissues. Primary lymphoma of skeletal tissue is relatively rare as compared to secondary.[12] Multiple lytic lesions found in vertebrae of our patient most likely represent secondary skeletal involvement.

Diagnosis of lymphoma can be confirmed by cyto-pathological examination of bone marrow, lymph node, or lymph nodal mass, & peripheral blood analysis along with other laboratory tests.[1]

CONCLUSION

The wide-ranging imaging manifestations of disseminated Non-Hodgkin's lymphoma along with broad spectrum of extra nodal involvement pose a diagnostic challenge

for clinician and radiologist. CT scan plays a vital role in assessing the extent of the disease and staging but diagnosis is such cases can be confirmed only by biopsy.

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