

# Clinicoepidemiological Profile of Extranodal Lymphoma: The Experience of a Tertiary Care Center in India

## Abstract

**Introduction:** Lymphoma is a neoplastic proliferation of lymphoid cells at various stages of differentiation and affects lymph nodes, with infiltration into the bone marrow, spleen, and thymus, which form the primary lymphatic organs. Extranodal lymphoma, by definition, involves sites other than lymph nodes, spleen, thymus, and the pharyngeal lymphatic ring. Extranodal involvement is less common with Hodgkin lymphoma (HL) than with non-Hodgkin lymphoma (NHL). **Materials and Methods:** A single-center retrospective observational study was conducted over the period of 2017–2019. Age, gender, histologic type, location, type of clinical presentation, histologic diagnosis, and presence of specific symptoms were recorded, as were the specialty of the physician initially consulted and of the physician taking the diagnostic sample. **Results:** Twenty-seven cases of extranodal lymphoma were diagnosed: 12 (44.4%) were male and 15 (56.6%) were female patients. The median age for males was 49.6 years and for females was 45.7 years. In this study, we had varied presentations of the lymphoma with involvement of various structures. We had few patients with very rare site of involvement like an elderly female patient presented with firm swelling over the right forearm which on excision turned out to myeloid sarcoma. Similarly, an elderly woman had presented with pyrexia of unknown origin that was later diagnosed as having primary bone Hodgkin lymphoma (HL) which is a very rare diagnosis and rarely has been described in world literature. The most common site of extranodal lymphoma was gastrointestinal tract (7 out of 27 patients; 25.9%); the other sites reported were testis (14.8%), breast (7.4%) thyroid, and ovary. Few rare sites reported were bone, central nervous system, and mediastinum. We report an extremely rare patient who had presented with anterior chest wall swelling, and on evaluation was detected to have primary mediastinal B cell lymphoma. **Conclusion:** Extranodal lymphoma is the rare presentation of NHL and extremely rare for HL. As it is a very rare disease, there are very limited studies available for its staging and management. We have presented a case series of extranodal lymphoma with few very rare presentations.

**Keywords:** Diffuse large B cell lymphoma, extranodal lymphoma, primary mediastinal B cell lymphoma

## Introduction

The term “lymphoma” is defined as a heterogeneous group of biologically and clinically distinct neoplasms that originate from cells in the lymphoid organs. It involves lymphoid cells at various stages of differentiation and primarily affects primary lymphatic organs, i.e., lymph nodes, bone marrow, spleen, and thymus. It has been historically divided into two distinct categories: non-Hodgkin lymphoma (NHL) and Hodgkin lymphoma (HL).

However, lymphomas have been reported to be arising from nonnodal sites and have

been described as extranodal lymphoma. The definition of primary extranodal lymphomas is still controversial, and this may contribute to explain their varying percentage in comparison with the nodal ones. Extranodal lymphomas are considered to involve sites other than lymph nodes, spleen, thymus, and the pharyngeal lymphatic ring. They consist of a heterogeneous group of malignancies with various histological types and highly varied clinical presentation and are most commonly NHL rather than HL.<sup>[1-3]</sup>

Approximately one-third of NHL arise from sites other than lymph nodes, spleen, or the bone marrow. They may also arise from sites normally devoid of lymphocytes.<sup>[4,5]</sup> These tumors may mimic infection or epithelial cancers based on clinical and radiological

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picture. However, they are different in terms of treatment and prognosis.

The designation of stage III and IV lymphomas as primary extranodal NHLs is indeed questionable because extranodal involvement in the presence of mainly nodal or disseminated disease may represent secondary extranodal disease spread. Currently, it is accepted to operationally define as extranodal those lymphomas with no or only “minor” nodal involvement associated with a clinically dominant extranodal component.<sup>[6]</sup> As for the definition, there is no consensus about the staging of primary extranodal lymphomas: the Ann Arbor staging system is at present widely used for describing the extent of the disease.

As per the National Cancer Institute’s Surveillance, Epidemiology, and End Results Program, referring to the period 1978–1995, approximately 30% of all lymphomas were extranodal and almost half of all extranodal NHL cases reported had diffuse large B-cell lymphoma (DLBCL) histology.<sup>[7]</sup> Extranodal lymphomas can originate in almost every organ. Data from large series reported in the literature have shown gastrointestinal (GI) tract, skin, bone, and brain to be the most common sites of extranodal lymphoma.<sup>[6,7]</sup> While the Ann Arbor staging system considers tonsils and the Waldeyer’s ring as lymphatic localizations, there is controversy about their designation as extranodal sites. Nevertheless, when they are included in the extranodal lymphoma series, head-and-neck localizations are the second most frequent site. Moreover, the incidence of primary extranodal presentation is variable across the different B-cell histologic subtypes, encompassing the majority of Burkitt’s lymphomas (BL), up to 50% of DLBCL and less than 10% of follicular lymphomas (FL).<sup>[7]</sup> The distribution of histologic types may be site specific for some localization such as testis or central nervous system (CNS), where nearly all cases are DLBCL. Conversely, in the GI tract, a wide spectrum of lymphoma types can be found, comprising DLBCL, marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT), BL, mantle cell lymphoma (MCL), and FL.<sup>[6,7]</sup>

Over the last decade, the International Extranodal Lymphoma Study Group (IELSG) has originated several retrospective and prospective trials aimed at clarifying the specific features of primary extranodal NHL (<http://www.ielsg.org>). However, the majority of published reports remains limited to single-institution retrospective reviews, while, in prospective trials, extranodal lymphomas are usually accounted as nodal lymphomas.

This study sought to describe the clinicoepidemiological profile, histological varieties, and survival of the patients with extranodal lymphoma.

## Materials and Methods

A single-center retrospective observational study was conducted over the period 2017–2019 at a tertiary cancer

center in Kolkata, India. Age, gender, histologic type, location, type of clinical presentation, and histological diagnosis and presence of specific symptoms were recorded.

All patients diagnosed as extranodal lymphoma between January 01, 2017, and June 30, 2019, were analyzed retrospectively. Detailed history and clinical examination of all the patients were done. The presenting symptom leading to diagnosis was recorded and counted as the clinical presentation. B symptoms and cervical adenopathies were recorded; B symptoms comprise general symptoms: fever, night sweats and weight loss, associated with both HL and NHL. Age at diagnosis, gender, histological type, location, type of clinical presentation, time interval between symptom onset and histologic diagnosis and known risk factors (HIV-positive status and history of immune disorder) were recorded. The patients were subjected to various hematological and biochemical investigations. They were staged as per the existent guidelines (NCCN version 2.2019). Diagnosis was made on the basis of excision biopsy or tru-cut biopsy. The WHO 2016 classification was used for characterizing histologic type.

## Results

Twenty-seven cases of extranodal lymphoma were diagnosed: 12 (44.4%) were male and 15 (56.6%) were female patients. The median age was for male was 49.6 years and for female 45.7 years [Figures 1 and 2].

In this study, we had varied presentations of the lymphoma with involvement of various structures. We had few patients with very rare site of involvement like an elderly female patient without any previous comorbidity presented with firm swelling over the right forearm which on excision turned out to myeloid sarcoma. Similarly, an elderly woman had presented with pyrexia of unknown origin that was later diagnosed as having primary bone HL, which is a very rare diagnosis and rarely has been described in world literature.

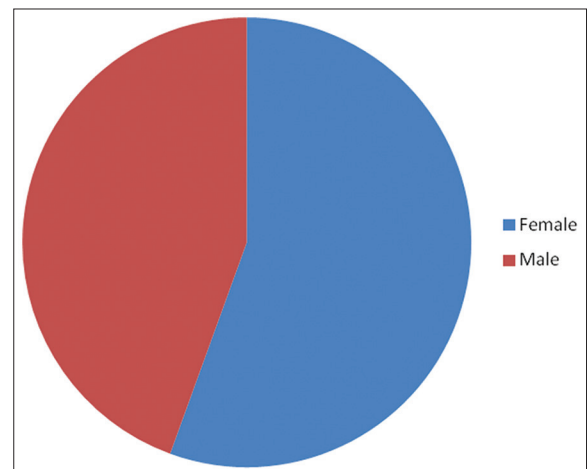


Figure 1: Sex-wise distribution

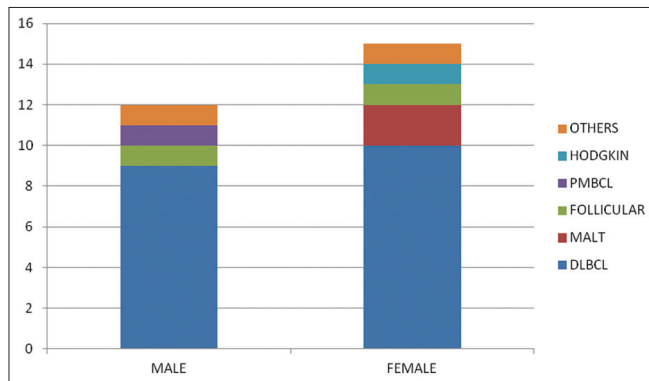
The most common site of extranodal lymphoma was GI tract (7 out of 27 patients; 25.9%), the other sites reported were testis (14.8%), breast [Figures 3 and 4] (7.4%), thyroid [Figure 5] and ovary. Few rare sites reported were bone, CNS, and mediastinum. We report an extremely rare patient who had presented with anterior chest wall swelling, and on evaluation was detected to have primary mediastinal B cell lymphoma [Figure 6].

The most common presentation was with lump or mass of the involved site, almost all the patients presented with mass or the symptoms related to the mass effect. Patients with breast lump had presented and on biopsy were detected to have NHL-DLBCL. Three patients with thyroid swelling were detected to have NHL (one with FL and two had DLBCL) [Table 1].

Two patients with ovarian NHL-DLBCL had presented with complaints of abdominopelvic mass with bloating

**Table 1: Various presentations**

Symptoms	Number of patients	Frequency (%)
Mass	12	44.4
B symptoms	15	55.5
Mass-related symptoms	12	44.4



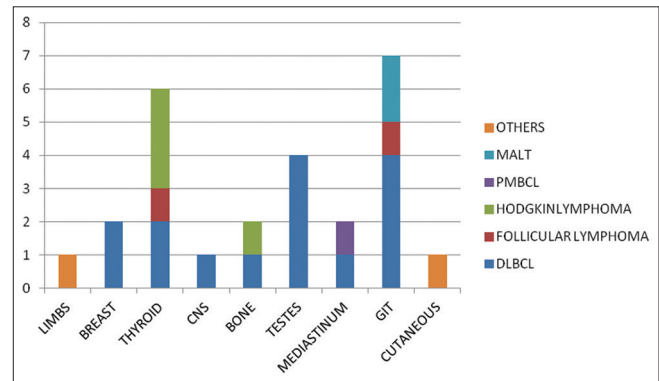
**Figure 2: Sex and various histologies**

sensation with normal CA125 levels. Computed tomography abdomen and pelvis was suggestive of bilateral adnexal mass with no ascites [Figure 7]. Both the women had undergone staging laparotomy with total abdominal hysterectomy with bilateral oophorectomy and on histopath examination, it was confirmed to be NHL DLBCL.

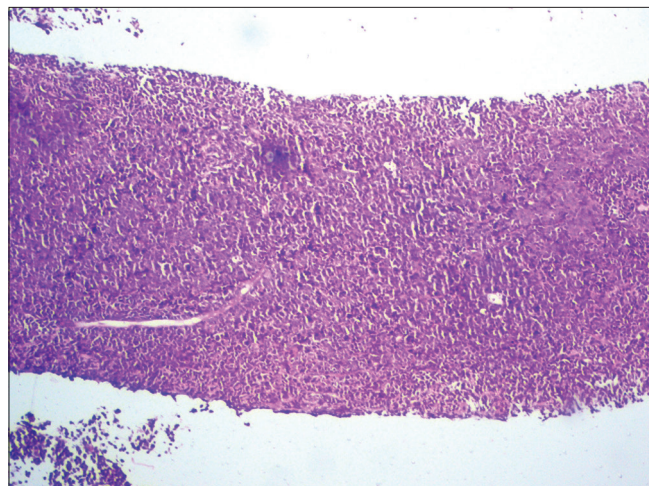
Patients with primary CNS lymphoma had presented with features of raised intracranial pressure and seizure. One patient had left-sided hemiparesis along with features of raised intracranial pressure. On magnetic resonance imaging brain, they had mass lesion for which they had undergone excision of the lesion and was diagnosed as primary CNS lymphoma.

Two patients had presented as pyrexia of unknown origin and on extensive workup diagnosed as primary bone lymphoma (PBL). Both were elderly females and on bone marrow examination detected to have primary bone HL, while the other patient had primary bone DLBCL. Primary bone HL is an extremely rare diagnosis and only very limited cases have been reported in the literature.

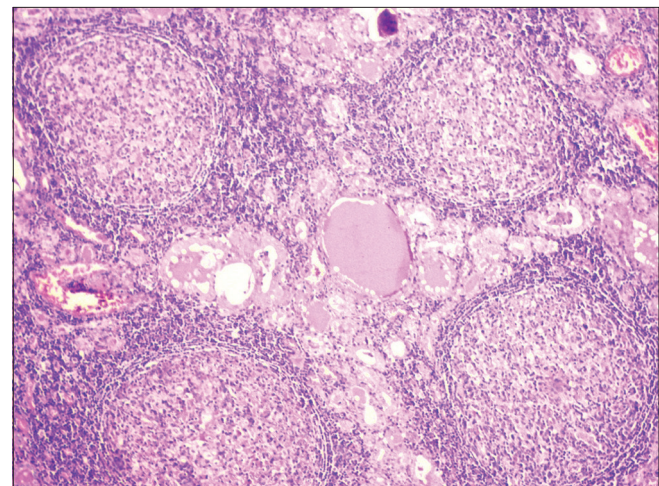
GI lymphoma was the most common cause of extranodal lymphoma, and these patients presented with pain abdomen and anorexia. None of the patients had had abdominal lump and also none of the patients had presented with features



**Figure 3: Site and various histologies**



**Figure 4: Diffuse large B-cell lymphoma breast**



**Figure 5: Follicular lymphoma thyroid**



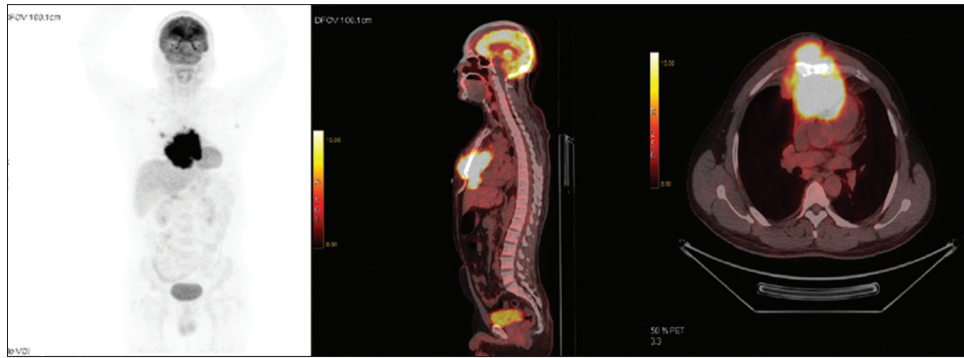


Figure 6: A young male presented with soft-tissue swelling over the chest and diagnosed as primary mediastinal large B cell lymphoma

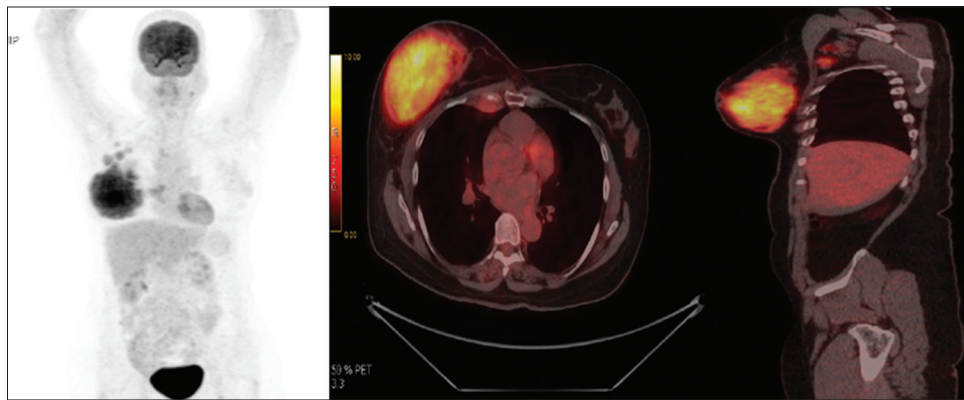


Figure 7: A middle-aged female presented with right breast lump diagnosed as diffuse large B cell lymphoma

of obstruction. Out of seven patients with GI lymphoma, stomach was the most common subsite (5 out of 7 patients followed by duodenum (2 of the 7 patients).

B symptoms have been classically described as fever, drenching night sweats, and weight loss of more than 10% over 3 months. Fourteen out of 27 (51.8%) patients had presented with B symptoms. Most of these patients had NHL DLBCL. The most common B symptom was weight loss followed by fever [Table 1].

In our study, most of the patients had NHL (25 out of 27), while only one had HL and one had myeloid sarcoma. The patient with primary bone HL had been diagnosed as classical HL based on bone marrow biopsy. Rest of the patients was diagnosed with NHL. Twenty-four out of 25 patients were found to have B cell lymphoma and one had T cell lymphoma.

Among B cell lymphoma, DLBCL was the most common histology (19 out of 24; 79.16%). These patients were further subclassified into activated B cell (13 out of 19; 68.4%) and germinal center (06 out of 19; 31.6%) based on Hane's algorithm. Double hit and double expressor lymphoma were not diagnosed in our study.

## Discussion

Almost 25% all NHL and very rarely HL can arise from tissue other than lymph nodes or tissue devoid of any

lymphoid tissue. These forms are broadly referred as primary extranodal lymphomas.<sup>[1,2]</sup> However, in case of the presence of both nodal and extranodal disease, it remains a controversial issue. Classically, lymphomas are considered to be extranodal when, after routine staging procedures or imaging, there is either no or minor nodal involvement along with a predominant extranodal component.<sup>[3]</sup> In our study also, we have considered only those lymphomas which are predominantly extranodal or with minimal lymph nodal involvement.

DLBCL represents 30%–58% of NHL and is the most common variant.<sup>[4]</sup> However, extranodal DLBCL represents only 1%–5% of all NHL. Mostly, it arises from the lymph nodes, but  $\leq 40\%$  initially present in extranodal sites, and they tend to have distinct molecular pathogenesis, clinical presentation, and natural history.<sup>[5]</sup> The most common site of origin is the GI tract; apart from that, many other organs may be involved such as the mediastinum, testis, CNS, breast, and bone. In our study also, the most frequent site noticed was GI tract with other rare sites involvement such as CNS, thyroid, and breast.

The GI tract involvement by NHL accounts 30%–40% of extranodal lymphoma<sup>[6-8]</sup> and from 4% to 20% of all NHL cases.<sup>[9]</sup> Various subtypes can arise from GI tract, with DLBCL being the most common accounting for 60% of gastric and 70% of intestinal cases.

MALT lymphoma represents about 35% of primary gastric lymphoma and less than 10% of the intestinal ones. At least one-third of them are a primary gastric lymphoma, which in approximately two-thirds of the cases is associated with a chronic *Helicobacter pylori* infection. FL is a rare variant, accounting for up to 17% of intestinal cases, while BL and MCL accounts for about 5% of the cases.<sup>[10]</sup>

GI tract lymphoma presents with abdominal pain, dyspepsia, nausea and vomiting, anorexia, obstruction, and hemorrhage. Fever and night sweats are usually absent. Weight loss is common; however, it is mainly due to the localization of the disease.<sup>[11]</sup>

Primary NHL of the testis accounts for about 9% of testicular neoplasms and 1%–2% of all NHL, with an estimated incidence of 0.26/100,000/year overall. NHL is the most common testicular malignancy in adult patients, and the median age of presentation is in the sixth decade. DLBCL accounts for 80%–90% of the cases. Other rare histologies described are Burkitt and Burkitt-like types (10%–20%), mainly in HIV-infected patients. FL of testis is very rare and has been reported mainly in childhood.<sup>[12]</sup>

These patients usually present with unilateral painless scrotal swelling, while B symptoms are usually present only in advanced stage. Rarely, they can present with abdominal pain, ascites, hydrocele, and bilateral testicular involvement. In addition to the contralateral testis, the disease typically spreads to other extranodal sites such as skin, subcutaneous tissue, lungs, bone, and CNS. In the largest series of 373 retrospective patients with testis lymphoma, the IELSG observed a 5- and 10-year incidence of CNS relapse of 20% and 35%, respectively.<sup>[13]</sup>

Primary CNS lymphoma (PCNSL) is a rare lymphoma which is characterized by the primary and exclusive involvement of the brain, spinal cord, leptomeninges, and eyes. It represents 4% of intracranial cancers and 4%–6% of primary extranodal lymphomas. Immunodeficiency is the only established risk factor for the development of PCNSL; patients with HIV infection have a 3600-fold increased risk compared with the general population. Majority of PCNSL in immunocompetent individuals are DLBCL, while indolent subtypes (usually small lymphocytic and lymphoplasmacytic lymphomas) are extremely rare. Patients usually present neurological symptoms depending on the site of lesion: focal deficits, neuropsychiatric symptoms, seizures, and manifestations of increased intracranial pressure.<sup>[14,15]</sup>

PBLs accounts for 5% of all primary extranodal NHLs, and 3%–7% of all bone cancers. Bone lymphoma are described in three: (a) PBL with single bone lesion without regional lymphadenopathies, (b) PBL with single bone lesion with

regional lymphadenopathies, and (c) polyostotic lymphoma characterized by a multifocal disease exclusively involving the skeleton and the disseminated lymphoma with secondary infiltration of the bone.<sup>[16,17]</sup>

The median age of presentation is between 45 and 60 years. Patients present typically bone pain (80%–95%); tumor mass is present in 30%–40% of the cases with pathological fracture in 10%–15% of them.<sup>[16,17]</sup> Spinal cord compression is present in 14% of the patients. Hypercalcemia and osteolysis are described in 10% and 15% of the cases, respectively, mainly in the presence of progressive disease. DLBCL is the most common lymphoma type involving primarily or secondarily the bone, accounting for 70%–80% of bone lymphomas.<sup>[18,19]</sup>

Primary mediastinal large B cell lymphoma is a unique type of NHL, which arises from thymic B-cells with an entirely different clinicopathological and genetic features. It mainly affects young female patients.<sup>[1]</sup> It typically presents as a large, rapidly growing tumor which invades the adjacent thoracic structures such as the chest wall, pleura, lungs, pericardium, and heart, causing pleural/pericardial effusion in approximately 30%–50% of cases. Bone marrow involvement is extremely rare, even in cases of recurrence. The differential diagnoses of PMBCL include other types of lymphomas with mediastinal localization and mediastinal tumors such as thymoma, germ cell tumors, and metastatic carcinomas. Other types of lymphomas with mediastinal localization include gray-zone lymphoma with features intermediate between DLBCL and classical HL, composite lymphoma, DLBCL with anterior mediastinum involvement, nodular sclerosis type of classical HL, and mediastinal sequential lymphomas which is PMBCL relapsing as HL [Figure 6].<sup>[20,21]</sup>

Primary cutaneous lymphoma is defined as the presence of cutaneous localizations alone without any nodal or systemic disease. They represent around 10% of all extranodal lymphomas. Moreover, the skin is a relatively common site of dissemination of many nodal NHLs, especially those of T-cell phenotype. However, the clinical behavior of primary cutaneous lymphomas versus primary nodal lymphomas involving the skin is usually different. Lymphomas of the skin are more often of T-cell type, with mycosis fungoides and Sezary syndrome constituting around 65% of the cases.<sup>[22-26]</sup>

Primary ovarian lymphoma is an uncommon entity, and usually the involvement of the ovary is frequently secondary to disseminated malignant lymphoma, with an incidence of 7%–26%. It accounts for 0.5% of all NHLs and 1.5% of all ovarian neoplasms. Among the various types of lymphoma, DLBCL is the most common and accounts for around 20% of ovarian lymphoma. This may be due to lack of lymphoid tissue within the ovary. It has been suggested that the primary ovarian NHL (PONHL) originates from lymphocytes in the

ovaries, surrounding blood vessels at the hilum and related to the corpus luteum. Most authors consider PONHL as a local involvement of a systemic disease. It should be differentiated from advanced epithelial carcinoma, which usually presents elevated tumor markers and ascites. Chemotherapy, but not radical surgery, is an optimal treatment;<sup>[4]</sup> thus, accurate diagnosis is critical to avoid unnecessary operation.<sup>[27-30]</sup>

## Conclusion

Extranodal lymphoma is the rare presentation of NHL and extremely rare for HL. As it is a very rare disease, very limited studies are available for their staging and management. We have presented a case series of extranodal lymphoma with few very rare presentations.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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