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# Primary Ovarian Non-Hodgkin's Lymphoma with Late Diagnosis Due to Unusual Presentation - A Case Report

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**Abstract** Involvement of NHL in ovary is seen in disseminated Lymphoma but primary ovarian lymphoma still not commonly seen. We presented the case where the patient had suffered for unexplained fever for more than 1 year and lately presented with gross weight loss as well as painless lower abdominal lump. CT scan showed a huge heterogeneous mass of about 15x12.4cm in pelvic cavity with mild ascites. Surgery was performed in view of ovarian neoplasm as CA125 was also raised. Histopathology confirmed the diagnosis of diffuse large B cell lymphoma with Immunohistochemistry confirmation. Treated with R-CHOP protocol for 6 cycles as IHC expressed CD20 positive. Post chemotherapy follow up PET scan showed complete response of treatment, though in spite of all treatment Primary ovarian lymphoma have a poor outcome with a range from 0% to 36% expected to survive for less than three years.

***Keywords:*** *primary ovarian non-hodgkin's lymphoma, late diagnosis*

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## 1. Introduction

Primary Non-Hodgkin's Lymphoma (NHL) in gynecological tract may involve among them ovarian primary is the commonest. It accounts for 0.5% of total NHL and 1.5% of all ovarian tumors [1], where Diffuse Large B-cell type accounts for about 20% of Primary Ovarian NHL (PONHL) [2]. Though involvement of the ovary by malignant lymphoma, particularly Non-Hodgkin's lymphoma (NHL), is a well-known manifestation of disseminated lymphoma with a frequency of 7%to 26% [3]. As ovary is a pelvic organ, most of the time it is very difficult to diagnose in early settings. The case described below reside from remote place and also noticed the painless abdominal mass after long history of constitutional symptoms.

## 2. Case Report

A 48 years Bangladeshi pre-menopausal woman with married for 30 years presented with history of fever for 1-2 years with low grade and on off manner. For her persisting fever she was treated by medicine consultant and failed to resolve. Peripheral Blood Film showed- Microcytic hypochromic anaemia with neutrophilia, Tuberculin test (Mantoux test) - Negative. But last 1 month she had high grade fever associated with loss of appetite with gross weight loss of about 7 kg. She also had complaints of a painless lower abdominal mass moving from right to left side. Per abdominal examination there was large mobile mass involving umbilical-hypogastric, right and left lumber region (20 weeks size) about 20x15cm in size, mild tender soft to hard in consistency. Per vaginal examination the cervix is broad, hypertrophied. She was evaluated and Computed Tomography scan of whole abdomen and pelvis done preoperatively that showed a large heterogeneous mass measuring about 15cm in CC, 9.3cm in AP and 12.4cm in TD s noted in the pelvic cavity extending up to the umbilicus, the lesion causing compression over urinary bladder antero-inferiorly and over sigmoid colon posteriorly with mild ascites also noted, liver is mildly enlarged 17.7cm (Figure 1, 2). Chest X-ray showed normal. CA125 was 193.7. She underwent Total Abdominal Hysterectomy with Right sided Salphingo Oophorectomy. Histopathology revealed- Ovarian Tumor- Nom Hodgkin's Lymphoma, Intermediate grade, Tumor size- 16x12x8.5cm, Fallopian tube- 10.5x1.5cm, Invades the broad ligament and Fallopian tube. Ascitic fluid- Negative for malignancy (Figure 3). Immunohistochemistry (IHC): LCA, CD20, CD79a, PAX-5, Vimentin- Positive; CD3- Positive (Scattered); Inhibin, Calretinin, CD99- Negative, Confirmed as- Diffuse large B-Cell lymphoma (Figure 4). Post-operative period was uneventful. She was diagnosed as Primary Ovarian Lymphoma, Ann Arbor stage- IEXB.

She started adjuvant chemotherapy with R-CHOP (Rituximab, Cyclophosphamide, Hydroxydoxorubicin, and Oncovin schedule and completed 6 cycles R-CHOP without any adverse effects. Post-operative LDH was 216 as well as post treatment PET-CT scan showed no metabolic residual or recurrence of disease.

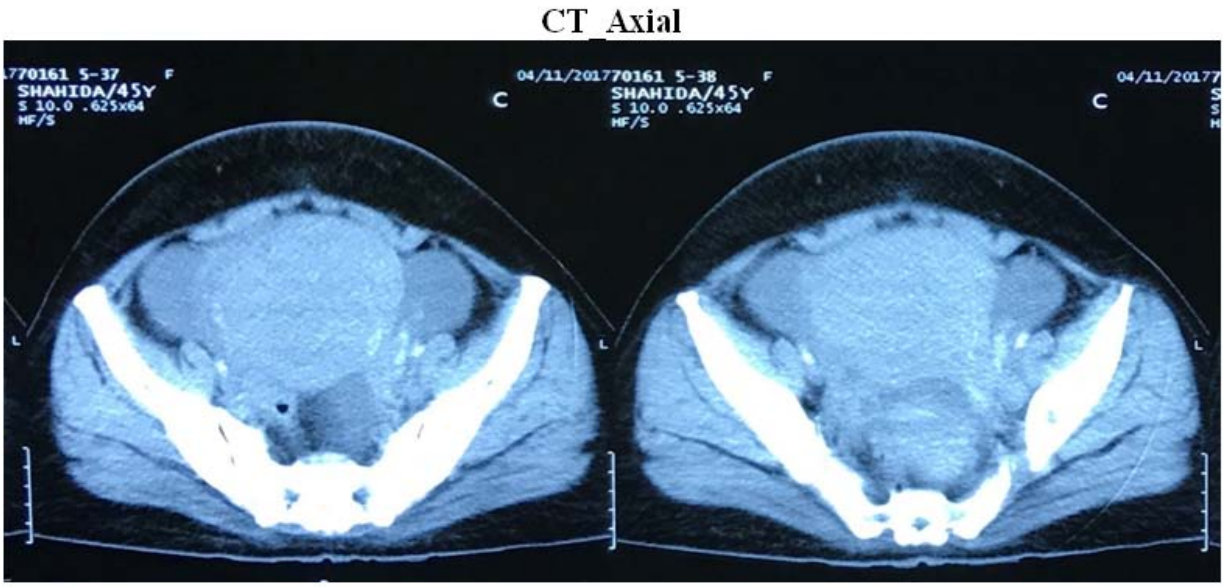


**Figure 1.** CT scan - Axial

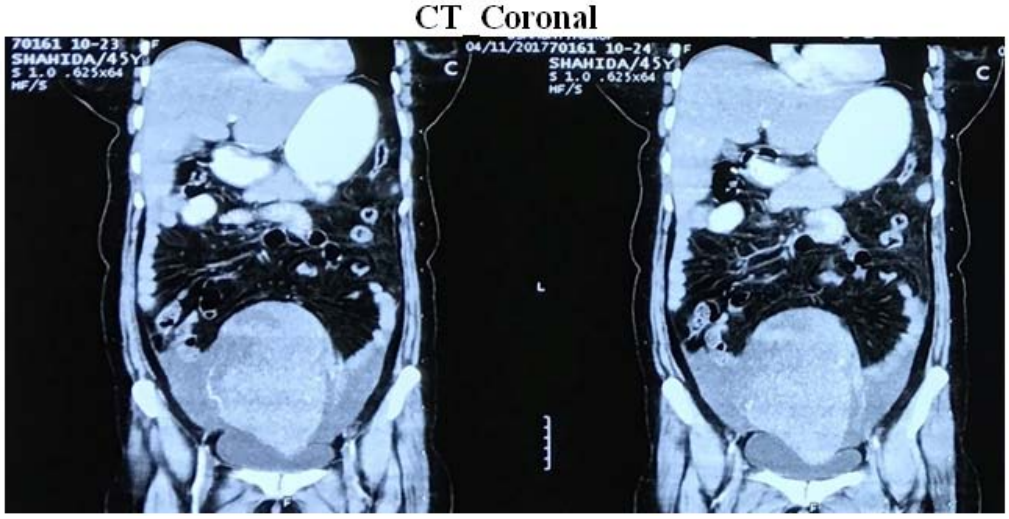


**Figure 2.** CT scan - Coronal

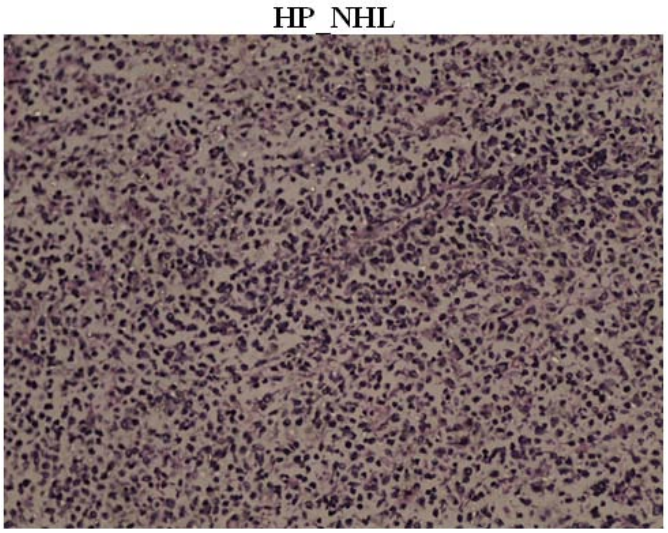


**Figure 3.** Histopathology

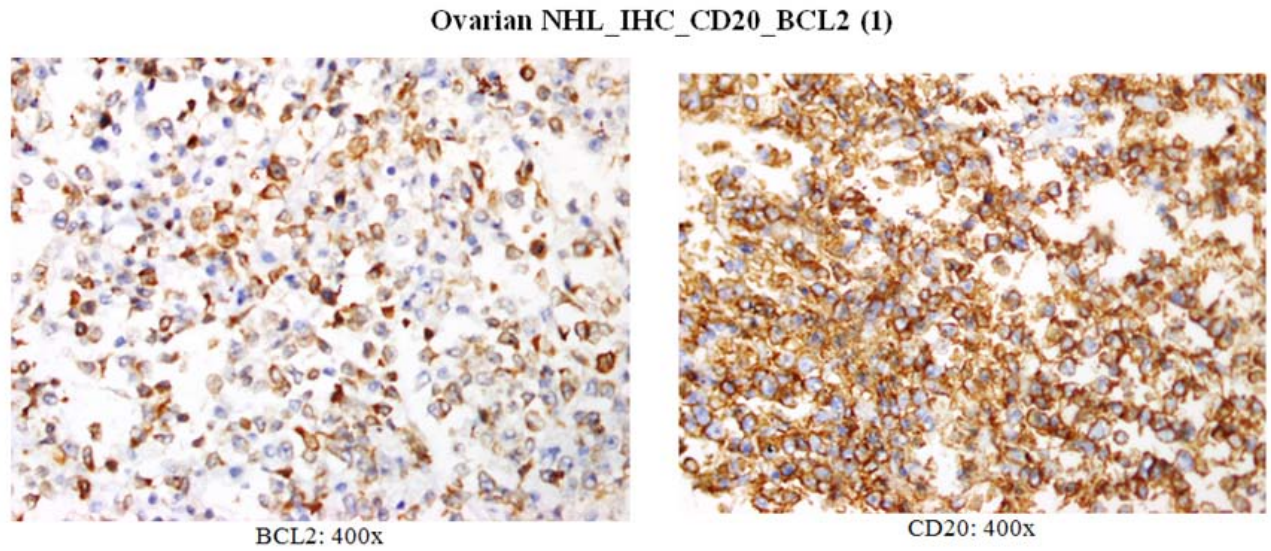


**Figure 4.** IHC (BCL2 & CD20)

## 3. Discussion

Primary ovarian Non-Hodgkin's Lymphoma (PONHL) accounts for 0.5% of total NHL and 1.5% of all ovarian tumors [1], where Diffuse large B-cell type accounts for about 20% [2]. Though involvement of the ovary by malignant lymphoma, particularly Non-Hodgkin's lymphoma (NHL), is a well-known manifestation of disseminated lymphoma with a frequency of 7%to 26% [3]. Primary Lymphomas of the Ovary (POL) can occur at any age, but most often in women over the age of 40 [4]. Lymphomas of the ovary may have varied presentations such as abdominal pain, pelvic mass, and ascites. Fever, night sweats, fatigue, or weight loss (B symptoms) was noted in 10%-33% of the patients [1,2]. In our case presented with fever for 1-2 years followed by high grade fever for 1 month with B symptoms of NHL with noticeable painless movable abdominal mass. Her fever was evaluated extensively but not resolved. But her noticeable pelvic mass correlates with ultrasonogram / CT scan and further gynecological evaluation making in conclusion of ovarian epithelial malignancy as her pre-operative CA125 was also raised (CA125-193.7). After surgery her histopathology showed the Intermediate grade NHL with negative ascitic fluid for malignant cells. Immunohistochemistry(IHC) showed Positive for -LCA, CD20, CD79a, PAX-5 & Vimentin; and expressed negative for- Inhibin, Calretinin, CD99, CD3 showed - Positive Scattered, final diagnosis as per IHC was- Diffuse large B-Cell lymphoma.

It is difficult to determine whether ovarian lymphomas are primary or secondary. There has been much controversy in the histogenesis of Primary Ovarian Lymphomas. It is demonstrated that normal ovaries contain lymphocytes of B-cell and T-cell lineage within cortical granulomas and rare lymphocytes are present throughout the ovarian stroma and within ovarian follicles and corpora lutea, which can be the source of NHL [3]. There is a possibility that chronic inflammation involving the ovaries predisposes to the development of NHL, analogous to low-grade B-cell MALT-lymphoma involving the stomach (4).

Fox et al. proposed the following criteria for the diagnosis of PONHL [5]. (1) At the time of diagnosis, the lymphoma is clinically confined to the ovary and a complete investigation fails to reveal evidence of lymphoma elsewhere. However, an ovarian lymphoma can still be considered as primary if it has spread to immediately adjacent lymph nodes or if it has directly spread to infiltrate immediately adjacent structures. (2) The peripheral blood film (PBF) and bone marrow should not contain any abnormal cells. (3) If further lymphomatous lesions occur at sites remote from the ovary, then at least several months should have elapsed between the appearance of the ovarian and extra-ovarian lesions.

In our case the radiological and PBF did not prove the other primary sites though bone marrow study not performed. It was staged as Ann Arbor stage- IEXB. Less than 10% of all ovarian NHLs reported have been localized, presumably arising in the ovary [[6] (journal-3)].

The protocol for chemotherapy used in diffuse large B-cell histology is the standard R-CHOP regimen. Rituximab plays an essential role in treatment of CD20-positive B-cell lymphoma [7]. Radiotherapy is used in Partial response cases [7]. Overall survival (OS) ranged from 6 months to 41 months with median OS of 23 months [[8] (Journal-13)].

## 4. Conclusion

Primary Ovarian Lymphoma is difficult to diagnose early due to its location, but most of the time with the presence of B symptoms patients' need further evaluation which leads to make the diagnosis. For confirmation the diagnosis with IHC, Surgery has no alternative way. But chemotherapy plays an important role to treat the cases as like as nodal lymphoma. Rituximab combination with CHOP regimen remains standard protocol in case of CD20 Positive cases. In spite of all treatment Primary ovarian lymphoma have a poor outcome with a range from 0% to 36% expected to survive for less than three years [9].

## Acknowledgements

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## References

[1] M. A. Dimopoulos, D. Daliani, W. Pugh, D. Gershenson, F. Cabanillas, and A. H. Sarris, "Primary ovarian non-Hodgkin's lymphoma: outcome after treatment with combination chemotherapy," Gynecologic Oncology, vol. 64, no. 3, pp. 446-450, 1997.

[2] Monterroso, E. S. Jaffe, M. J. Merino, and L. J. Medeiros, "Malignant lymphomas involving the ovary: a clinicopathologic analysis of 39 cases," *American Journal of Surgical Pathology*, vol.17, no. 2, pp. 154-170, 1993.

[3] I. Chorlton, H. J. Norris, and F. M. King, "Malignant reticuloendothelial disease involving the ovary as a primary manifestation. A series of 19 lymphomas and 1 granulocytic sarcoma," *Cancer*, vol. 34, no. 2, pp. 397-407, 1974.

[4] Hussell T, Isaacson PG, Crabtree JE, Spencer J. The response of cells from low-grade B-cell gastric lymphomas of mucosaassociated lymphoid tissue to Helicobacter pylori. Lancet 1993; 4:571-4.

[5] H. Fox, F. A. Langley, A. D. T. Govan, S. A. Hill, and M.H. Bennett, "Malignant lymphoma presenting as an ovarian tumour: a clinicopathological analysis of 34 cases," British Journal of Obstetrics and Gynaecology, vol. 95, no. 4, pp. 386-390, 1988.

[6] Russell Vang, M.D., L. Jeffrey Medeiros, M.D., Roger A. Warnke, M.D., John P. Higgins, M.D., Michael T. Deavers, M.D, "Ovarian Non-Hodgkin's Lymphoma: A Clinicopathologic Study of Eight Primary Cases" *Modern Pathology*, Mod Pathol 2001; 14(11): 1093-1099, VOL. 14, NO. 11, P. 1093, 2001

[7] N. Sakurai, K. Tateoka, J. Taguchi, and T. Terada, "Primary precursor B-cell lymphoblastic lymphoma of the ovary: case report and review of the literature," International Journal of Gynecological Pathology, vol. 27, no. 3, pp. 412-417, 2008.

[8] Suresh Babu[1], Usha Amirtham2, Lokesh K.N1, Govind Babu.K1, Lakshmaiah K.C1, Lokanatha Dasappa1, C.S. Premalata2, Linu Abraham Jacob1, Rudresh AH1, Rajeev LK1, Venugopal Arroju1, "Ovarian Non-Hodgkin's Lymphoma from a Regional Cancer Centre in South India" International Journal of Science, Vol- 5, Issued- 2016(05).

[9] M. A. Dimopoulos, D. Daliani, W. Pugh, D. Gershenson, F.Cabanillas, and A. H. Sarris, "Primary ovarian non-Hodgkin'slymphoma: outcome after treatment with combination chemotherapy," Gynecologic Oncology, vol. 64, no. 3, pp. 446-450, 1997.