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Non Hodgkin's Lymphoma with Right Atrial Intra Cardiac Metastases

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Abstract

Background: Diffuse Large B-Cell Lymphoma (DLBCL) is the most variant of Non-Hodgkin's Lymphoma (NHL) and also the most common variant with secondary intracardiac masses. **Case summary:** 7 years old child presented to emergency with acute decompensated cardiac failure, ascites and tender hepatomegaly. 2D echo evaluation was suggestive of large intracardiac mass in the right atrium almost completely obstructing Tricuspid valve orifice, gross pericardial effusion and dilated Inferior Vena Cava (IVC). Emergency tumor excision surgery was performed which revealed 4 × 4 cm pinkish firm mass arising from anterior Tricuspid annulus which was completely excised. Child was extubated on postoperative day (POD) 0 and was on minimal inotropic support. Ascites reduced significantly on POD1 allowing abdominal palpation which revealed a mass in the epigastric region. This prompted evaluation by pediatrician and oncology workup suggestive of increased 18-Flouro Deoxy Glucose (18-FDG) uptake in the mediastinum, abdomen, bilateral proximal thighs, all mediastinal lymph nodal stations, bilateral lung hilar stations 10R, 10L involving all encasing the heart and great vessels with pleural deposits, Celiac trunk, superior Mesenteric Artery (SMA), Portal vein, IVC and abdominal aorta. Histo pathology Examination (HPE) and Immuno Histo Chemistry (IHC) of intracardiac mass revealed DLBCL which is metastatic in nature. Chemotherapy was started as per (French American British Lymphomes Malins B) FAB LMB-96 protocol with the child currently in the Induction phase having poor prognosis and less survival interval. **Conclusion:** Surgery can be considered a treatment option for metastatic intracardiac masses during emergency scenarios like cardiogenic shock to relieve ob-

struction along the pathway of blood flow in the heart even though we may not be able to completely excise the tumor surgically.

Keywords

Diffuse Large B-Cell Lymphoma (DLBCL), Non-Hodgkin's Lymphoma (NHL), Secondary Intracardiac Metastasis, Cardiogenic Shock, Immuno Histo Chemistry (IHC)

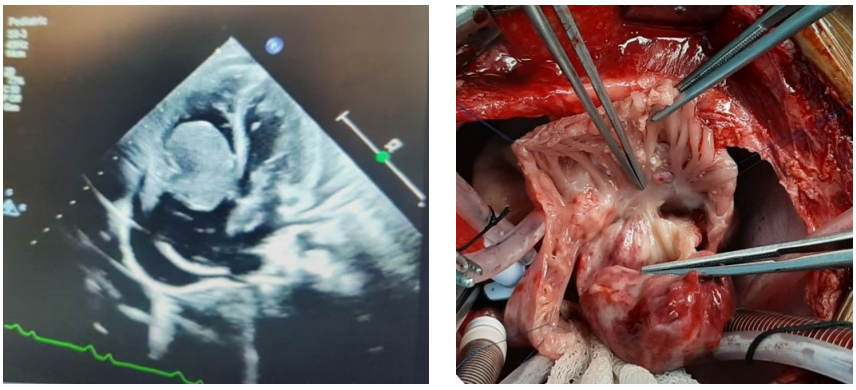
1. Introduction

Secondary cardiac tumours are more common than primary cardiac masses. Lymphoma is one of the common neoplasms that present with intra cardiac metastatic masses accounting to 8.7% - 27.2% of the total Lymphoma cases [1]. Lymphoma also arises as primary cardiac tumour amounting to 1.3% of all the tumours of the heart [2]. Clinical features of cardiac lymphoma comprise of heart failure, cardiac rhythm abnormalities, pericardial effusion and heaviness in the chest [3]. Because of the secondary involvement of the heart in lymphomas, cardiac symptoms are more commonly masked by the symptoms arising from other organ involvement leading to misdiagnosis on several occasions. Diffuse Large B Cell Lymphoma (DLBCL) is the most common subtype of NHL that presents with both primary as well as metastatic cardiac masses among all the Lymphoma variants [4]. In metastatic disease, cardiac involvement occurs as a late presentation and on many occasions, these are diagnosed during autopsy. We report a case of 7 years old child who presented with acute cardiac failure and cardiogenic shock with a right atrial intracardiac mass to the emergency department.

2. Patient Profile

A 7 year old male child presented to the emergency department with hypotension with blood pressure 78/46 mmHg, acute shortness of breath New York Heart Association (NYHA) class IV, tachypnoea with respiratory rate of 33 breaths per minute, pulse oximetry saturation of 72% on room air, cold peripheries, feeble pulse, tachycardia with heart rate of 120 per minute, raised Jugular Venous Pulse (JVP), pedal edema, diastolic murmur at the level of xiphoid, tender hepatomegaly and ascites. Child was started on inotropes Adrenaline and Nor Adrenaline @ 0.1 mcg/kg/min, fluid resuscitation was done, emergency intubation was performed and chest X ray was performed which was suggestive of cardiomegaly. Ultrasonography of the abdomen revealed a large ill-defined mass approximately of 5 × 5 cm near the pancreas and small bowel loops with bowel wall thickening. This mass could not be palpated clinically due to tense ascites. 2D Echocardiography was performed which was suggestive of 3 × 4 cm large echogenic mass in the right atrium which was completely obstructing Tricuspid

valve annulus extending into the right ventricle, gross pericardial effusion and hugely dilated inferior vena cava (**Figure 1(a)**). Child was taken up for emergency surgery with the intention of complete tumour excision via midline sternotomy. Cardiopulmonary bypass was instituted after giving Heparin with Aorto-bicaval cannulation. Superior and Inferior vena cava were looped and systemic cooling to 28 degree Celsius was started. Aortic cross clamp was applied and cardioplegic arrest in diastole was performed with cold del Nido cardioplegia solution. Caval tapes are snared and right atrium was opened which demonstrated 4 × 4 cm pink coloured firm mass attached to the Tricuspid valve anterior annulus extending to right atrium and right ventricle (**Figure 1(b)**). Mass was excised without any grossly visible residual mass and additional biopsy was taken from right ventricular outlet tract. Right atrium was closed and Aortic cross clamp was released. Cardiopulmonary bypass was weaned off after rewarming, haemostasis was assessed and Protamine was given. Pacing wires and drains were placed and sternal wound was closed. Child was shifted to Intensive Care Unit (ICU) with Dobutamine support @ 5 mcg/kg/min and mechanical ventilation. Postoperatively child was extubated after 3 hours and Dobutamine was tapered off on postoperative day 1 followed by drain and pacing wire removal on postoperative day 2 and day 4 respectively. Once ascites was reduced significantly on postoperative day 1, an epigastric mass was palpable on abdominal palpation. All invasive lines, drains and pacing wire was removed on postoperative day 3 and child was shifted to ward. Intracardiac mass biopsy and Immunohistochemistry revealed diffuse proliferation of round cells in sheets which are dis-cohesive with round regular hyperchromatic nuclei with scanty cytoplasm. The cells are positive for markers Cluster of Differentiation antigens CD20, CD10, LCA (Leucocyte Common Antigen), (B-cell Lymphoma) BCL6 and BCL2 suggestive of Diffuse Large B-cell Lymphoma (**Figures 2(a)-(f)**). Paediatric oncologist was consulted who advised a Positron Emission Tomography (PET) combined with Computerised Tomography (CT) scan which revealed



(a) (b)

Figure 1. (a) 2D-Echocardiography image suggestive of well-defined tumour mass across the Tricuspid valve annulus spanning from right atrium to right ventricle. (b) intraoperative image of tumour mass arising from the anterior Tricuspid annulus.

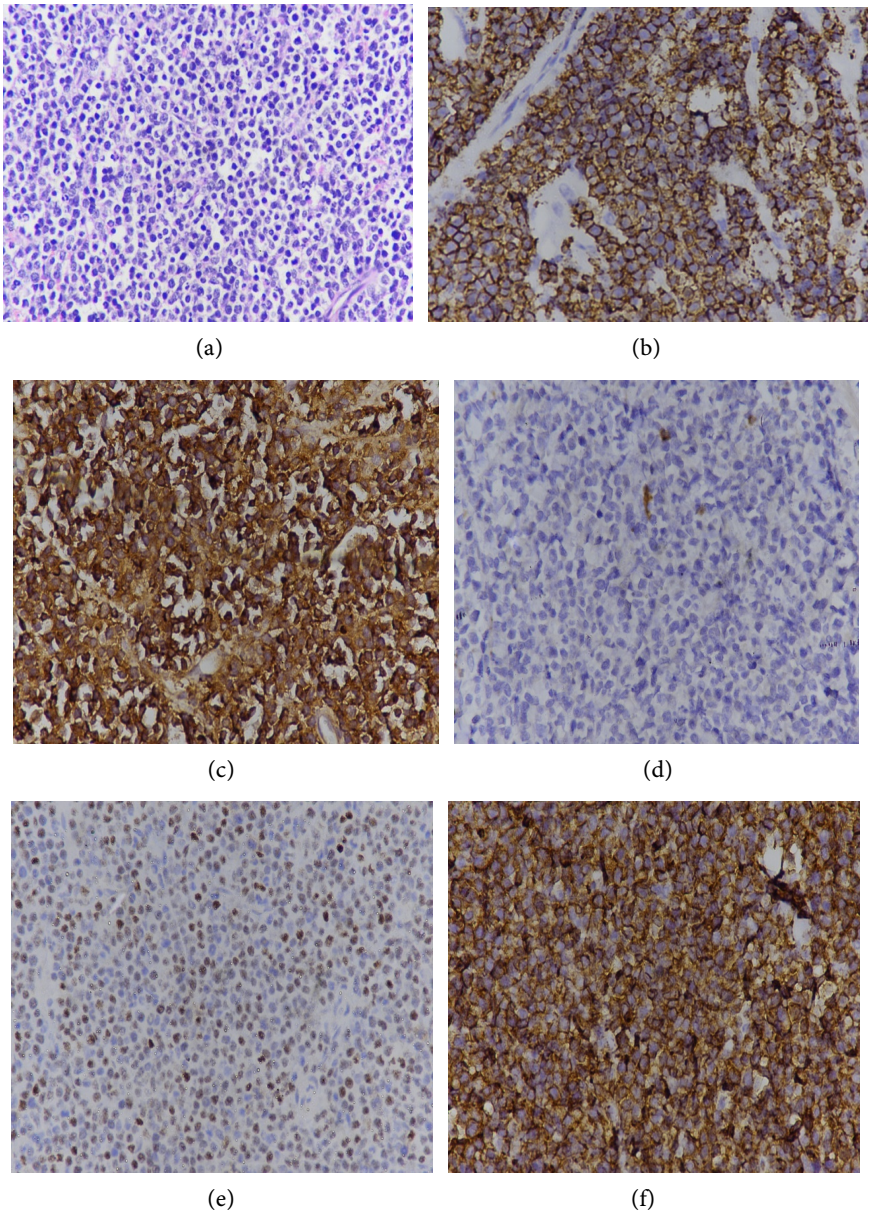


Figure 2. Histopathology and Immuno Histochemistry images. (a) Tumour showing large atypical Lymphoid cells (Haematoxilin & Eosin stain) 400× magnification. (b) Tumour cells positive for CD 20 marker on Immunohistochemistry. (c) Tumour cells positive for CD 10 marker on Immunohistochemistry. (d) Tumour cells positive for marker CD 3 on Immunohistochemistry. (e) Tumour cells positive for BCL-6 marker on Immunohistochemistry. (f) Tumour cells strong diffuse positivity for CD 45 marker on Immunohistochemistry.

increased 18-Flouro Deoxy Glucose (18-FDG) uptake in the mediastinum, abdomen, bilateral proximal thighs, all mediastinal lymph nodal stations, bilateral lung hilar stations 10R, 10L involving all encasing the heart and great vessels with pleural deposits, Celiac trunk, superior Mesenteric Artery (SMA), Portal vein, IVC and abdominal aorta (**Figures 3(a)-(e)**) suggestive of a lymphoproliferative disorder. Bone marrow biopsy was negative for blast cells. With the

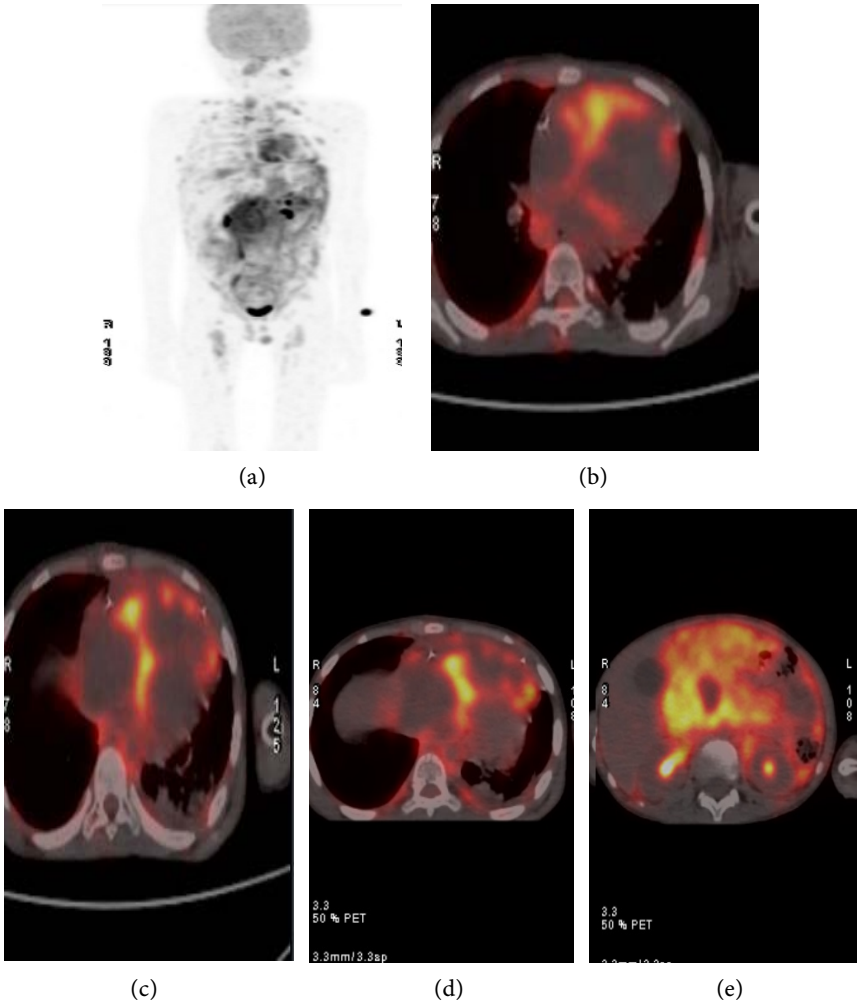


Figure 3. PET CT images. (a) MIP (Maximum intensity projection) images of 18-F FDG PET-CT. Increased FDG uptake is noted in the mediastinum, abdomen and bilateral proximal thighs. Physiological FDG uptake is noted in the brain, myocardium, kidney and urinary bladder. (b) FDG avid sheet like lymph nodal mass involving all the mediastinal lymph nodal stations with pericardial deposits. (c) Faint to non FDG avid fibro-consolidatory changes noted in the basal segments of left lower lobe of lung with multiple pericardial deposits encasing heart and major blood vessels. (d) Diffuse FDG avid pericardial thickening encasing the heart with multiple diffuse pleural deposits. (e) FDG avid ill-defined sheet like lymph nodal mass noted involving bilateral retrocrural, perigastric, gastro hepatic, pre aortic and precaval regions with multiple diffuse FDG avid peritoneal thickening.

above investigations results, the case was diagnosed to be a DLBCL a variant of NHL with intra cardiac metastatic mass. Child was transferred under the care of pediatric oncology team and was started on chemotherapy as per FAB LMB-96 protocol [5].

3. Discussion

It difficult to differentiate a Primary Cardiac Lymphoma (PCL) from Secondary cardiac lymphoma metastasized to heart from nodal primary and there are dif-

ferent opinions regarding this. Zaharia L *et al.* opines that if the tumor involves the pericardial space and myocardium, it can be diagnosed as PCL [6]. On the other hand, presence only cardiac mass without any other lymphomatous malignancy elsewhere in the body is defined as PCL according to Curtsinger CR *et al.* [7]. 2 large case series of PCL comprising of 197 between 1949-2009 and 101 cases between 2009-2019 were published by Petrich *et al.* and Chen *et al.* respectively [8] [9]. Voigt *et al.* reported 558 cases of secondary cardiac involvement due to Lymphoma of which 77% was comprised of DLBCL variant [10]. Gordon *et al.* reported 43 cases of NHL with secondary cardiac metastasis of which 42% was constituted by DLBCL variant [11]. Cardiac involvement as secondary metastasis occurs by means of dissemination via blood, lymphatic spread and through direct invasion from surrounding mediastinal masses.

Clinical presentation of masses in the heart is often nonspecific in the form of pedal edema, shortness of breath, cardiac failure with cardiogenic shock, arrythmias, features of embolization of tumor cells to other organs in the body and that of valvular heart disease. Constitutional symptoms of malignancy such as weight loss, loss of appetite, fever and night sweats are also often quite misleading [12] [13]. In our case constitutional symptoms of malignancy were absent and on initial preliminary noninvasive diagnosis with 2D Echocardiography of a well circumscribed intra cardiac mass after auscultating a diastolic murmur at the level of xiphisternum, we suspected it to be a benign mass rather than a malignant one. Since the presentation is an emergency with acute cardiogenic shock with a mass obstructing the flow of venous return in the heart at the level of Tricuspid valve, the patient was reeled into the operation theatre to relieve the obstruction by excision of the tumor and there was no time and also the patient was not stable for preoperative confirmation of diagnosis by performing tissue biopsy under ultrasonography guidance from the intraabdominal mass detected. Other possible investigation that can be done in emergency scenarios which can give information about the presence of other masses in the body is whole body CT scan.

Chemotherapy is the standard modality of treatment for Lymphoma even in cases of cardiac involvement like that of Superior Vena Cava obstruction to offer symptomatic relief with similar outcomes compared to that of combination therapy with radiation as well [14] a similar clinical scenario of low cardiac output as in our case due to obstruction of pathway of venous blood flow towards lungs. But in our case since it's an emergency scenario with cardiogenic shock, and surgery can be considered as a salvage treatment option to stabilize the patient by improving blood flow to the lungs by excising the tumor in the right ventricular outlet tract though it may not offer complete cure from malignancy [15].

Postoperatively after the cardiac surgical care, the child was transferred under the care of pediatric oncologist and chemotherapy was started as per FAB LMB-96 protocol [5] and child is presently in Induction phase of chemotherapy. The survival rate of untreated DLBCL variant of NHL with cardiac metastasis is

only 10% at the end of 1 year [13] but there is a possibility of cure with intense chemotherapy. Disease not responsive to chemotherapy or with early recurrence is managed by addition of stem cell therapy. Post treatment regular follow up of patients with clinical evaluation and laboratory investigations is needed every 3 - 6 months up to 5 years while imaging follow up is performed every 6 months till 2 years [16].

4. Conclusion

DLBCL is the most common variant of NHL presenting as intra cardiac mass as form of extra nodal metastasis rarely presenting as right heart failure with cardiogenic shock as a result of obstruction of blood flow to lungs which necessitates emergency cardiac surgery during which it may or may not be possible to completely resect the tumour and achieve a R0 resection. If the mass is incidentally detected with primary elsewhere in the body and the patient is hemodynamically stable, chemotherapy is the primary treatment modality after histological confirmation of diagnosis which is effective even for treating malignant masses at both cardiac and non-cardiac sites.

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Conflicts of Interest

The authors do not have any conflict of Interests.

Ethical Approval

The study has been conducted after approval from Institutional Ethics Committee of All India Institute of Medical Sciences, Raipur, India for reviewing the medical records of the patients bearing approval number IECSG-123/06-02-2023. The patient data and other information were used in compliance to the Declaration of Helsinki.

Informed Consent

Informed Consent has been obtained from the patient(s) to publish their information anonymously for academic purpose.

References

[1] Swerdlow, S.H., Campo, E., Pileri, S.A., *et al.* (2016) The 2016 Revision of the World Health Organization Classification of Lymphoid Neoplasms. *Blood*, **127**, 2375-2390. https://doi.org/10.1182/blood-2016-01-643569

[2] Johri, A., Baetz, T., Isotalo, P.A., Nolan, R.L., Sanfilippo, A.J. and Ropchan, G. (2009) Primary Cardiac Diffuse Large B Cell Lymphoma Presenting with Superior Vena Cava Syndrome. *Canadian Journal of Cardiology*, **25**, E210-E212. https://doi.org/10.1016/S0828-282X(09)70110-2

[3] El Sayed, M.J., Gharzuddin, W. and Kharfan-Dabaja, M.A. (2014) T-Cell Acute Lymphoblastic Leukemia Presenting as a Cardiac Tamponade. *Austin Journal of Emergency and Critical Care Medicine*, **1**, Article ID: 1007.

[4] Majano-Lainez, R.A. (1997) Cardiac Tumors: A Current Clinical and Pathological Perspective. *Critical Review in Oncogenesis*, **8**, 293-303. https://doi.org/10.1615/CritRevOncog.v8.i4.10

[5] Patte, C., Auperin, A., Gerrard, M., Michon, J., Pinkerton, R., Sposto, R., Weston, C., Raphael, M., Perkins, S.L., McCarthy, K., Cairo, M.S. and FAB/LMB96 International Study Committee (2007) Results of the Randomized International FAB/LMB96 Trial for Intermediate Risk B-Cell Non-Hodgkin Lymphoma in Children and Adolescents: It Is Possible to Reduce Treatment for the Early Responding Patients. *Blood*, **109**, 2773-2780. https://doi.org/10.1182/blood-2006-07-036673

[6] Zaharia, L. and Gill, P.S. (1991) Primary Cardiac Lymphoma. *American Journal of Clinical Oncology*, **14**, 142-145. https://doi.org/10.1097/00000421-199104000-00009

[7] Curtsinger, C.R., Wilson, M.J. and Yoneda, K. (1989) Primary Cardiac Lymphoma. *Cancer*, **64**, 521-525. https://doi.org/10.1002/1097-0142(19890715)64:2<521::AID-CNCR2820640227>3.0.CO;2-T

[8] Petrich, A., Cho, S.I. and Billett, H. (2011) Primary Cardiac Lymphomaan Analysis of Presentation, Treatment, and Outcome Patterns. *Cancer*, **117**, 581-589. https://doi.org/10.1002/cncr.25444

[9] Chen, H., Qian, S., Shi, P., Liu, L. and Yang, F. (2020) A Presentation, Treatment, and Survival Analysis of Primary Cardiac Lymphoma Cases Reported from 2009 to 2019. *International Journal of Hematology*, **112**, 65-73. https://doi.org/10.1007/s12185-020-02881-2

[10] Voigt, P., Wienbeck, S., Weber, M.A., Oyama-Manabe, N., Beimler, M., Schob, S., *et al.* (2017) Cardiac Hematological Malignancies: Typical Growth Patterns, Imaging Features, and Clinical Outcome. *Angiology*, **69**, 170-176. https://doi.org/10.1177/0003319717713581

[11] Gordon, M.J., Danilova, O., Spurgeon, S. and Danilov, A.V. (2016) Cardiac Non-Hodgkin's Lymphoma: Clinical Characteristics and Trends in Survival. *European Journal of Haematology*, **97**, 445-452. https://doi.org/10.1111/ejh.12751

[12] Aledavood, S.A., Emadi Torghabeh, A., Homaee Shandiz, F. and Memar, B. (2015) Cardiac Involvement in Non-Hodgkin Lymphoma, an Incidental Large Atrial Mass: A Case Report. *International Journal of Cancer Management*, **8**, e3913. https://doi.org/10.17795/ijcp-3913

[13] Habertheuer, A., Ehrlich, M., Wiedemann, D., *et al.* (2014) A Rare Case of Primary Cardiac B Cell Lymphoma. *Journal of Cardiothoracic Surgery*, **9**, Article No. 14. https://doi.org/10.1186/1749-8090-9-14

[14] Talapatra, K., Panda, S., Goyle, S., *et al.* (2016) Superior Vena Cava Syndrome: A Radiation Oncologist's Perspective. *Journal of Cancer Research and Therapeutics*, **12**, 515-519. https://doi.org/10.4103/0973-1482.177503

[15] Jonavicius, K., Salcius, K., Meskauskas, R., *et al.* (2015) Primary Cardiac Lymphoma: Two Cases and a Review of Literature. *Journal of Cardiothoracic Surgery*, **10**, Article No. 138. https://doi.org/10.1186/s13019-015-0348-0

[16] Zelenetz, A.D., Gordon, L.I. and Abramson, J.S. (2019) NCCN Guidelines Insights: B-Cell Lymphomas, Version 3.2019. *Journal of the National Comprehensive Cancer Network*, **17**, 650-661. https://doi.org/10.6004/jnccn.2019.0029