



Lymphoma Involving the Heart and Inferior Vena Cava: Radiological and Pathological Approach

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Abstract

Primary cardiac lymphoma (PCL) is a very rare condition accounts for 2% of all primary cardiac tumours. This is a case report of a 76 year old male patient with shortness of breath, cough and pyrexia of unknown origin for 2 months. In the initial diagnostic workup, echocardiography revealed aortic and mitral regurgitation with no pericardial effusion. With the development of right sided pleural effusion about one month after the initial presentation, ultrasound scan guided pleural fluid aspiration performed. Pleural fluid culture revealed no bacterial growth. Second echocardiography showed no flow in the superior vena cava (SVC) and suspected of a thrombus. Contrast enhanced Computed Tomography scan of the chest demonstrated a mass lesion in the right heart and distal inferior vena cava (IVC). The lesion was compressing the SVC without complete obstruction. The pericardium was intact with no pericardial effusion. No mediastinal lymphadenopathy identified. Initial differential diagnosis was leiomyosarcoma of the distal IVC extending in to the right heart. As there was no pericardial involvement, possibility of lymphoma was made as a remote cause. Subsequently digital subtraction angiography guided biopsy performed from the mass in the distal IVC and right atrium. Diffuse lymphoma was confirmed on histology. As there were no Hodgkin cells in the sample, probable diagnosis of Non-Hodgkin's lymphoma was made. Subsequently patient died due to cardiovascular compromise before starting specific therapy.

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Received Date: 27 Jul 2017

Accepted Date: 31 Oct 2017

Published Date: 12 Nov 2017

Citation:

Duminda WD, Pathirana KG, Lekamge IN, Weerakoon S, De Silva YAP, Ganewatte E, et al. Lymphoma Involving the Heart and Inferior Vena Cava: Radiological and Pathological Approach. *Ann Clin Case Rep*. 2017; 2: 1461.

ISSN: 2474-1655

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Keywords: Cardiac lymphoma; Inferior vena cava; Radiological imaging; Histological diagnosis

Introduction

Primary cardiac lymphoma (PCL) is a very rare condition usually of B-cell non-Hodgkin's type. They account for 2% of all primary cardiac tumours and 1% of extranodal lymphomas [1,2]. They are usually diffuse large cell lymphomas manifest as an ill-defined, infiltrative mass [3]. PCL are more commonly seen in immunocompromised states, either iatrogenic including those associated with solid organ or bone marrow transplantation or HIV related and these are usually associated with EBV infection. Lymphomas in immunocompetent individuals are extremely rare [4]. As there are no specific clinical symptoms, the clinical diagnosis of the condition is difficult. Typically when B symptoms develop, (fever, weight loss, fatigue common in lymphoid malignancies), progressive heart failure will ensue in these patients [5]. However advanced cross sectional imaging modalities allowed early and accurate detection of primary cardiac malignancies [6]. The definitive diagnosis of the condition needs histological evaluation of the tumour. One of the accessible diagnostic methods is transesophageal echocardiography -guided transjugular biopsy, which has a relatively low risk of complications [7]. Endomyocardial biopsy via percutaneous cardiac approach is also another approach for histological diagnosis [8]. We present a case of lymphoma arising in the heart with extension in to the superior and inferior vena cava.

Case Presentation

A 76 year old male was initially investigated for shortness of breath and cough of 2 months duration in a specialized hospital for respiratory disease. On initial presentation the patient was not dyspnoeic but found to have 94% of saturation of oxygen in blood. Bilateral ankle oedema was evident at that time (Figure 1). The blood pressure was normal on admission to the hospital. During hospital stay patient develops mild fever episodes intermittently. Sputum culture and blood culture were negative during the febrile illness. Subsequently patient underwent transoesophageal

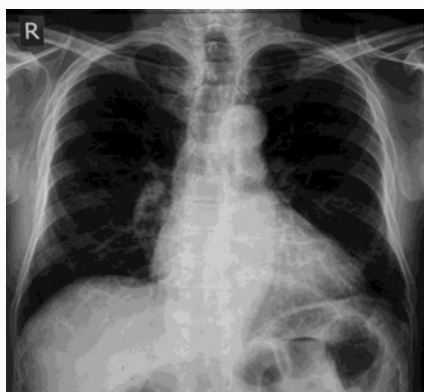


Figure 1: Chest radiograph in the initial stage of the disease.



Figure 2: Longitudinal section through the inferior vena cava on ultrasound scan showing no demonstrable flow within the IVC.

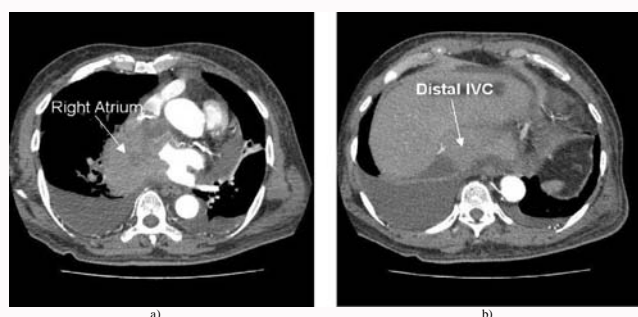


Figure 3: Contrast enhanced Computed Tomography images showing irregular filling of the right side heart chambers and distal IVC. a) Mass lesion in the right heart chambers. b) Mass lesion appears in the distal IVC.

echocardiography suspecting infective endocarditis, and it revealed grade ii aortic regurgitation and mitral regurgitation (Figure 2). There was no evidence of pericardial effusion or features of congestive cardiac failure.

About a month following initial presentation the patient develops right sided pleural effusion and then ultrasound scan guided pleural fluid aspiration was performed (Figure 3 a and b). There were 3.8g/dl of protein and 80% of lymphocytes in the pleural fluid. However there was no evidence of bacterial growth or acid fast bacilli in the pleural fluid.

Again patient was send for cardiology opinion and underwent 2 dimensional Echocardiogram which revealed no flow in the superior vena cava (SVC) with suspicion of a thrombus in the SVC extending in to the right ventricle (Figure 4). Right atrium was dilated at that time. Then the patient was transferred to the National Hospital of Sri Lanka (NHSL) for further evaluation and management.

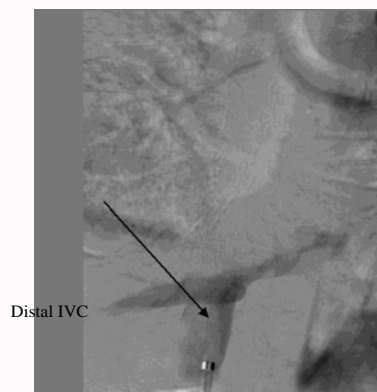


Figure 4: Incomplete filling identified within the distal IVC during digital subtraction inferior venocavography.

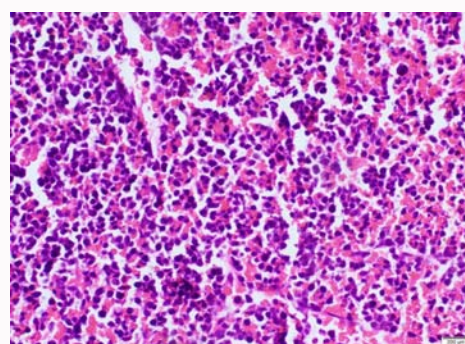


Figure 5: Haematoxylin and Eosin staining showing sheets of discohesive medium to large lymphoid cells with enlarged hyperchromatic nuclei and narrow rim of cytoplasm.

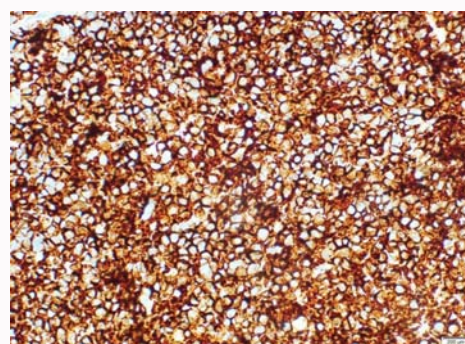


Figure 6: Immunohistochemistry (IHC) with Leukocyte Common Antigen (LCA) revealed strong membranous positivity in all lymphocytes.

A contrast enhanced Computed Tomography (CT) scan of the chest performed in the NHSL and detected a contrast enhancing mass lesion with few hypodense areas within the lesion involving the distal inferior vena cava (IVC), right atrium and right ventricle. The lesion was appearing to compress the distal SVC without complete obstruction of the lumen. Right main pulmonary artery was pushed superiorly by the mass lesion without significant luminal narrowing. The pericardium was not involved by the lesion and there was no pericardial effusion. There was bilateral pleural effusion on CT scan. There was no significant hilar or mediastinal lymphadenopathy. In conclusion differential diagnosis of leiomyosarcoma of the distal IVC extending in to the right atrium and right ventricle was made. As there was no pericardial effusion or pericardial involvement by the mass lesion, the possibility of lymphoma was made as a remote

possibility.

Subsequently the patient referred for digital subtraction angiography guided biopsy of the mass lesion in the distal IVC and right atrium. With right femoral vein puncture the venous access gained and IVC catheterization was performed. Five biopsy samples were obtained using 10 F guiding catheter and an Alligator forcep.

On histological assessment with haematoxylin and eosin staining there were sheets of discohesive medium to large lymphoid cells with enlarged hyperchromatic nuclei and narrow rim of cytoplasm. Mitoses were frequently found. Immunohistochemistry (IHC) with Leukocyte Common Antigen (LCA) revealed strong membranous positivity in all lymphocytes (Figures 5 and 6). Diffuse lymphoma was confirmed by histology. As there were no Hodgkin cells in the biopsy, probable diagnosis of Non-Hodgkin type lymphoma was made. Subsequently the patient was transferred to The National Cancer Institute, Sri Lanka for further management. However the patient died from cardiac arrest before starting specific therapy.

Discussion

Primary cardiac lymphoma (PCL) is a very rare condition. On histology they are usually of B-cell non-Hodgkin's type lymphoma. The most frequent location of PCL is the right atrium, followed by the pericardium. Signs and symptoms are non-specific and depend on the location and extent of the tumor; these include right-sided heart failure precordial pain, arrhythmias, conduction disorders and cardiac tamponade [7]. In literature there is a case of atypical presentation of ischemic hepatitis in a young adult with PCL, which highlights the wide variety of clinical manifestations of this rare tumour [9]. In our patient the initial presentation was nonspecific and the disease progressed rapidly. Radiological imaging played major role in the identification and sampling of the tumour. The diagnosis of diffuse lymphoma was confirmed by histology with probable diagnosis of Non-Hodgkin lymphoma.

The most effective treatment method for cardiac lymphoma is chemotherapy. Palliative surgery may be necessary to correct haemodynamics when venous blood flow to the lungs is disturbed [10]. Guilherme HO et al. [11] concluded in their study that cardiac lymphomas had worse survival compared with extra-cardiac lymphomas.

Unfortunately this patient died due to cardiovascular compromise before starting chemotherapy or palliative surgery.

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