Case Report

F18 FDG positron emission tomography revelation of primary testicular lymphoma with concurrent multiple extra nodal involvement

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ABSTRACT

Primary testicular lymphoma (PTL) a relatively rare disease of non-Hodgkin's lymphomas occurring with a lesser incidence of 1-2% has a propensity to occur at later ages above 50 years. PTL spreads to extra nodal sites due to deficiency of extra cellular adhesion molecules. We present detection of multiple sites of extra nodal involvement of PTL by F-18 positron emission tomography/computed tomography study aiding early detection of the dissemination thus aiding in staging and management.

Keywords: Extra nodal sites, F-18 FDG, non-Hodgkin's lymphoma, positron emission tomography/computed tomography, primary testicular lymphoma

INTRODUCTION

Amongst non-Hodgkin's lymphomas (NHL) occurrence of primary testicular lymphoma (PTL) is fairly uncommon, the incidence is a meager 1-2% of all the NHLs.[1,2] Non lymphomatous testicular tumors occur at a relatively earlier age and primary lymphoma testis has a propensity for occurrence at a later age usually above 50 years Usual clinical presentation is one of a localized disease with unilateral testicular involvement and the disease localized to the testis with no extra testicular spread.[2,3] Occasionally, PTL tends to spread to extra-nodal sites such as contra lateral testes, central nervous system, skin, lung, pleura, Waldeyer's ring and soft-tissues.[4,5] Involvement of these sites may occur either concurrently on presentation or subsequently at relapse. Clinical demonstration of extra nodal sites is very often difficult due to paucity of signs and symptoms. Signs and symptoms tend to be very subtle and non-specific.[5]

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CASE REPORT

A 48-year-old male presented with progressive enlargement of right testis of 3 weeks duration. There was associated dragging sensation in the right testicular region and in the lower abdomen. No associated urinary symptoms, fever or weight loss. No medical history of diabetes or hypertension and no family history of any solid or hematological cancers. Physical examination revealed a large right testicular swelling of 10 cm × 8 cm size, firm in consistency with mild pain on palpation, testicular sensation was preserved. There was no accompanying palpable inguinal adenopathy. Contra lateral testis was normal in size and unremarkable on physical examination. Systemic examination was unremarkable. Initial laboratory data revealed mild normocytic normochromic anemia with hemoglobin of 10.2 g/dL, platelet count of 2.9 lakhs/cu mm, lactate dehydrogenase of 369 U/L. Chest radiograph was normal. Patient underwent a computed tomography (CT) scan of the abdomen and pelvis, which revealed an enlarged right testis of uniform density with no retroperitoneal lymphadenopathy and right adrenal was mildly enlarged. FNAC of right testis showed provisional possibility of poorly differentiated carcinoma. Positron emission tomography/CT (PET/CT) was performed for initial assessment and staging, which revealed a markedly enlarged right testis showing intense metabolic activity with standardized uptake value (SUV) max

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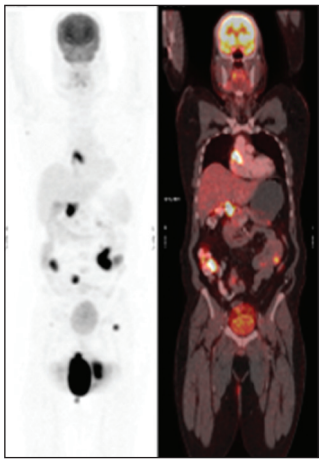

Figure 1: Positron emission tomography/computed tomography showing enlarged right testis with intense metabolic activity, normal sized left testis, loco regional bilateral small volume inguinal nodes also showing intense FDG uptake. Hyper metabolic additional areas of hyper metabolic activity in contra lateral testis, adrenal, small bowel loops, and superior vena cava

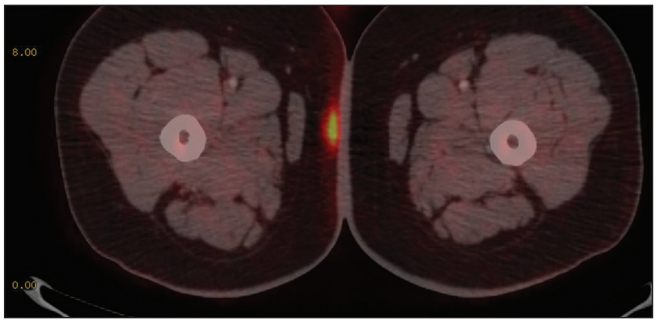

Figure 3: Positron emission tomography/computed tomography revealing hyper metabolic nodule in the sub cutaneous planes of right thigh

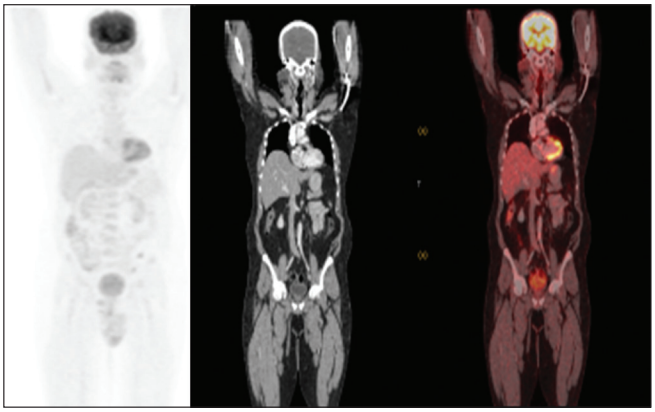

Figure 5: Interim positron emission tomography/computed tomography post-surgery and four cycles of chemotherapy showing complete morphological and metabolic regression of all the lesions

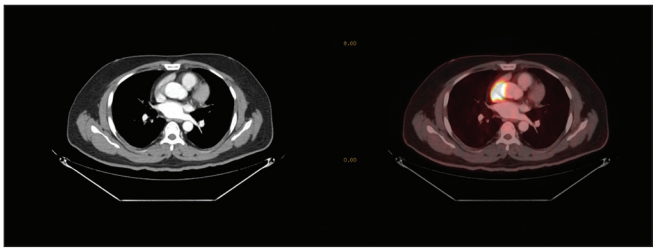

Figure 2: Positron emission tomography/computed tomography image showing metabolically active hypo dense area encircling the superior vena cava right atrial junction

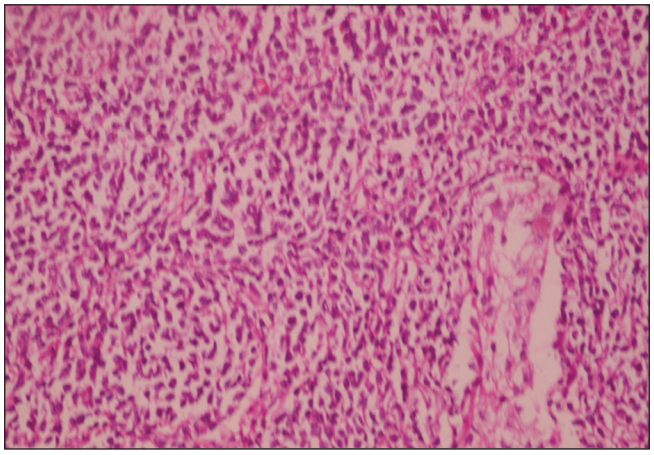

Figure 4: Histopathology of right testis showing microscopically shaved entrapped somniferous tubules in sheets of large monomorphic dyscohesive cells showing vesicular pleomorphic nuclei, clumped chromatin, small nucleoli, and minimal amount of cytoplasm suggesting primary testicular diffuse large B cell lymphoma

of 35.5. Normal sized left testis also showed intense Fluoro deoxy glucose uptake with SUV max of 15.9. Hyper metabolic activity was also noted in loco regional small volume inguinal nodes bilaterally with SUV max of 15.5. There were additional areas of hyper metabolic activity noted in extra testicular extra nodal sites such as, contra lateral testis, adrenals, and small bowel loops [Figure 1]. Metabolically active area of a CT based hypo dense area encircling the superior vena cava right atrial junction [Figure 2]. Hyper metabolic nodule was noted in the subcutaneous planes of right thigh [Figure 3]. Patient underwent a right inguinal orchidectomy. Histopathology showed enlarged testis measuring 9.5 cm × 6 cm × 6 cm. Cut section was fleshy and grey white. Normal testicular tissue was not evident. Microscopy showed entrapped seminiferous tubules in sheets of monomorphic dyscohesive cells. These cells are large and show vesicular pleomorphic nuclei, clumped chromatin, small nucleoli, and a minimal amount of cytoplasm. Mitosis is increased. Surgical margin of the spermatic cord was free of tumor. Tunica was not infiltrated with tumor [Figure 4]. Immuno histo chemistry was carried out for further typing which showed diffuse membranous CD-20 positivity with increased Ki67 index of 85-90%. CD10 was negative. Findings were consistent with primary testicular diffuse large B cell lymphoma, Post surgery chemotherapy with Cyclophosphamide, Hydroxydaunorubicin, Oncovin (Vincristine), Prednisolone regimen was followed by an interim PET after four cycles of chemotherapy, which showed complete morphological and metabolic regression of all the lesions [Figure 5].

DISCUSSION

PTL is a rare disease representing 1-2% of all non-Hodgkin's lymphomas and less than 9% of all testicular cancers[1,2] Unlike other testicular cancers, PTL occurs mainly in patients aged over 50 and in fact 85% of all PTLs are diagnosed in patients over the age of 60.[3,4] Most patients present with localized disease either stage I or II.[2,5] Testis seems to provide a sanctuary for lymphoma cells because of the blood test is barrier; thus, escaping the cell mediated immune surveillance and inhibiting the penetration of the chemotherapeutic agents. Rarely PTL may show relapse at other extra nodal sites such as contra lateral testes, central nervous system, skin, lung, pleura, Waldeyer's ring, soft issues, bone, bone marrow, adrenal glands, liver, gastrointestinal tract, and spleen.[4,6] Waldeyer's ring involvement is attributable to the common embryonic origin of testis, nasopharynx and oropharynx from the endoderm.[7] Usual clinical presentation is with isolated swelling of testis, but synchronous involvement of multiple sites, which may occur either concurrently at presentation or subsequently at relapse is unusual. This propensity for extra nodal dissemination is attributed to aberrant expression of extra cellular adhesion molecules (ECAM) on neoplastic cells. The resultant poor adhesion of lymphoma cells to extracellular matrix of primary neoplasm and alteration of cell mediated host responses may also contribute to the phenomenon of extra nodal dissemination. This selective deficiency of ECAM is peculiar to testicular lymphomas.[8] Extra nodal sites involvement is mostly unearthed at post mortem, with an autopsy incidence of 10-25% of lymphoma cases.[9,10] Unusual and rare site involvement of heart has been documented in only a single publication.[11] Ante-mortem diagnosis of extra nodal sites is often challenging, as signs and symptoms pertaining of the involved organs are totally lacking, subtle or non-specific.[6] Complete initial staging work-up is the same like other non-Hodgkin's lymphomas. It includes a complete physical examination, complete hematological and biochemical exams, total-body computerized tomography, bone marrow aspiration, and biopsy. Incorporation of PET/CT in the initial lymphoma staging has immensely augmented the staging potential by its unique ability to detect and simultaneously identify the primary and localizes all the extra nodal sites in one study making the overall staging more specific and also aiding the therapeutic approach. Lymphomatous involvement of heart itself is a rare phenomenon and is commonly observed in immunocompromised patients such as HIV/AIDS or following bone marrow or solid organ transplantation.[2,12] Our case has not only cardiac spread, but four others extra nodal sites simultaneously involved and unearthed by PET/CT.

CONCLUSIONS

Our case highlights the importance of F-18 FDG PET/CT imaging in this rare case of PTL with concurrent multiple extra nodal involvements. To the best of our knowledge, this is the first reported diagnosis of PTL with simultaneous involvement of cardiac, cutaneous, intestinal, and adrenal and contra lateral testis in an immune competent patient at presentation.

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