Case Report

Peripheral T-cell lymphoma, not otherwise specified

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Abstract

The peripheral T-cell lymphoma, not otherwise specified (PTCL, NOS) belongs to a heterogeneous class of aggressive neoplasms. Although several morphologic subtypes of this tumor have been described, no particular genetic, immunological, or distinct clinical features define this disease. Patients can experience night sweats, fever, lymphadenopathy, weight loss, splenomegaly, and/or skin changes. Common laboratory tests reveal that patients have anemia, thrombocytosis, lymphocytosis, eosinophilia, hypergammaglobulinemia, or increased lactate dehydrogenase. In this case study, a patient presented with massive lymphadenopathy and right lower limb swelling, which he developed over 6 weeks. A tissue biopsy and supporting investigations confirmed the diagnosis of PTCL, NOS.

Keywords: CHOP regimen, lymphoma, peripheral T-cell lymphoma not otherwise specified, pralatrexate

Introduction

Peripheral T-cell lymphoma, not otherwise specified (PTCL, NOS) comprises a common, extranodal, and nodal T-cell lymphomas subgroup. These lymphomas are not characterized by any known clinicopathological criteria. In Western countries, PTCL, NOS is the most widespread subtype of PTCL, affecting about 30% of PTCL patients and 4% of overall non-Hodgkin lymphomas (NHLs).[1-5] The World Health Organization and the European Organization for Research and Treatment of Cancer have grouped PTCL, NOS in a class of primary skin lymphomas having a diverse clinical presentation. Such lymphomas demonstrate a higher frequency in middle-aged males, with a male: female ratio of 2.5:1.[6]

Case Report

An 80-year-old Italian male with a history of hypertension, type II diabetes mellitus, osteoarthritis, and glaucoma presented with a complaint of right groin pain that persisted for 6 weeks. He had a mass in the right groin. He had no fever, anorexia or weight loss, and no other lumps in other parts of the body. His level of energy was good. There was no history of alcohol and smoking. There was no history of similar illness in the family.

We palpated an 11 cm × 6 cm solid mass in the right groin. Distended veins could be seen over the mass. A swelling was also present on the right lower limb, along with pitting edema up to the popliteal fossa. All vital systems were otherwise healthy. He was already taking glipizide 5 mg OD, pravastatin 20 mg OD, glucosamine and chondroitin sulfate BID, and multivitamins.

We ordered a complete blood count (CBC), comprehensive metabolic panel (CMP), and computed tomography (CT) scan for the abdomen and pelvis. His CBC, CMP, and peripheral smear were normal. CT of the abdomen and pelvis showed a right inguinal lymph node mass measuring up to 11.3 cm × 6.3 cm anteroposteriorly and transversely extending up to 13 cm in length [Figure 1]. There was a 2.4 cm cystic or necrotic component in the inferior lateral position of the mass [Figure 2]. Fine-needle aspiration cytology (FNAC) was scheduled. CT-guided FNAC of the mass revealed numerous small to midsized atypical lymphoid

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Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/jfmpc.jfmpc_323_16

How to cite this article: Jha KK, Gupta SK, Saluja H, Subedi N. Peripheral T-cell lymphoma, not otherwise specified. J Family Med Prim Care 2017;6:427-30.

cells, which were consistent with NHL. To confirm NHL, a core biopsy was performed. It showed diffuse effacement of nodular architecture by small to intermediate-sized lymphocytes with moderate cytologic atypia [Figure 3].

The immune cell infiltrates were CD3 [Figure 4] and CD30 being positive. Within the mass, 57% of infiltrates stained strongly for CD2 and CD3. The alpha and beta T-cell receptor stainings were positive while the gamma delta receptor staining was negative. An immunohistochemical stain for ALK1 was negative. Epstein–Barr virus staining was negative. The combination of morphological and immunophenotypical findings was most consistent with PTCL, NOS.

Treatment

After the diagnosis of PTCL, NOS, the patient was sent to an oncologist. Serum protein electrophoresis, erythrocyte sedimentation rate, and positron emission tomography scan were normal. CHOP regimen total of six cycles every 3 weeks was prescribed. After the first cycle, his limb became less swollen, and he reported a significant improvement in his symptoms. He took a total of four cycles of the CHOP regimen and he no longer followed up. Two years later, he returned with the complaint of heart palpitation and breathlessness. On investigations, we found cytopenia. We did a bone marrow biopsy to check for relapse, and we found bone marrow metastasis. After confirmation of relapse, we treated the patient with pralatrexate.

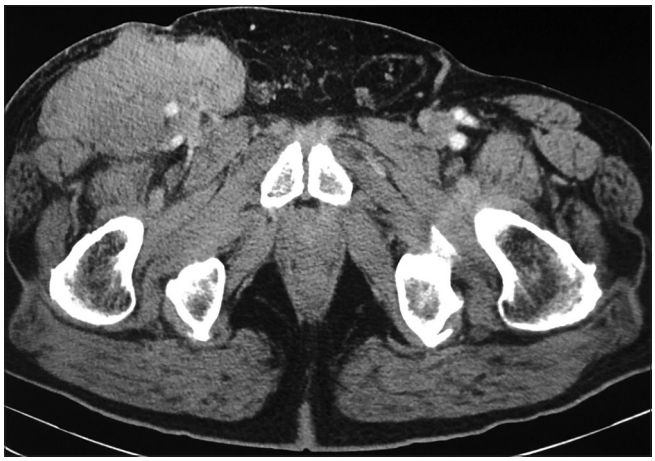

Figure 1: Computed tomography of the abdomen and pelvis showing a right inguinal lymph node mass measuring up to 11.3 cm × 6.3 cm

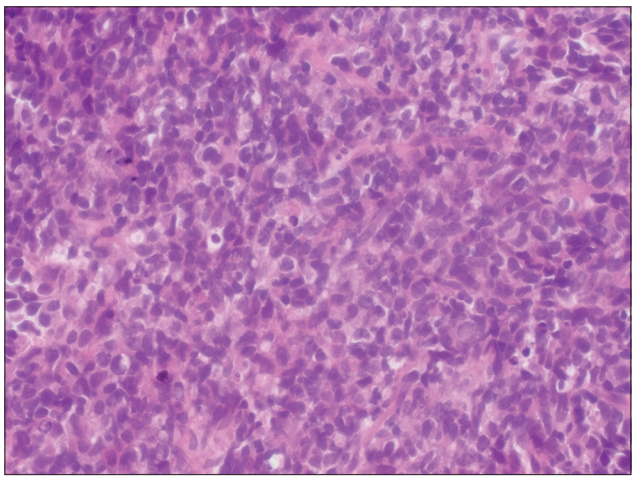

Figure 3: A core biopsy showing diffuse effacement of nodular architecture by small to intermediate-sized lymphocytes with moderate cytologic atypia

Discussion

The PTCLs are aggressive tumors and comprise about 15% of all adult NHL.[1,7] In the United States, from 1992 to 2006, there was an increase in the incidence of PTCL from 0.1 cases per 100,000 patients to about 0.4 cases per 100,000 patients.[8] The disease usually develops in adults and the average age of occurrence is 60 years at the time of diagnosis.[9] The associated risk factors of PTCL, NOS are still not known although immune-suppressing drugs, Epstein–Barr infection, smoking, chemicals like pesticides, and solvents exposure have been linked to the disease.[10,11]

The PTCL, NOS is characterized by symptoms of generalized lymphadenopathy and rare extranodal disease.[9,10,12] Extranodal

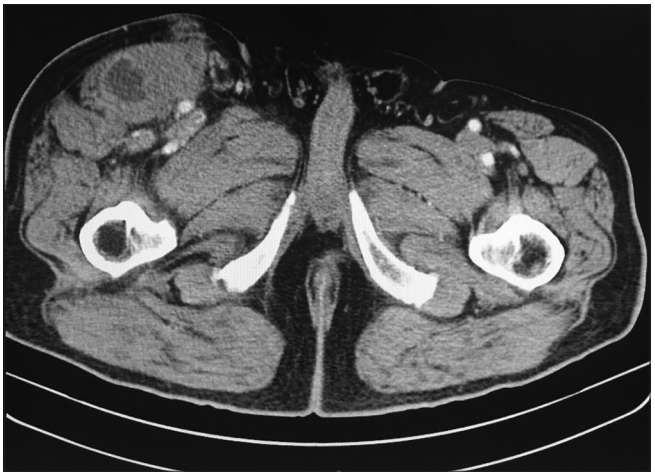

Figure 2: A 2.4 cm cystic or necrotic component in the inferior lateral position of the mass

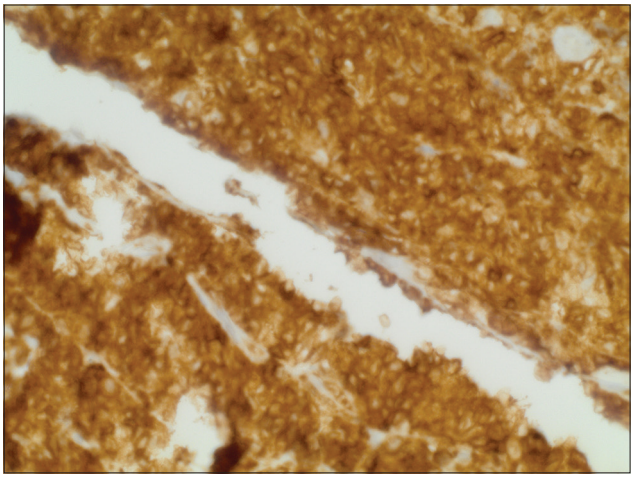

Figure 4: CD3-positive immune cell infiltrates

sites mostly involved are the gastrointestinal tract and skin.[1,13-15] There is no particular morphological aspect of this tumor. PTCL, NOS is usually diagnosed by a biopsy of the tumor tissue or a lymph node. However, to establish the tumor's T-cell origin, assessment of the immunophenotype is needed, and it is done either by flow cytometry or immunohistochemistry. Immunohistochemistry is reliable because large lymphoid cells are available for examination, whereas in flow cytometry, samples are sometimes lost or disrupted during processing.[10,11] PTCL, NOS contains mixtures of small, intermediate, and large atypical cells.[16,17] When genotypically investigated, antigens linked to T-cells are found to be expressed although with no consistent pattern (CD2+/−, CD3+/−, CD7−/+, CD5+/−). PTCL, NOS does not stain positive for B-cell antigens.[17,18] In the majority of cases, a loss of mature T-cell antigens (one or more) occurs, including CD7 or CD5. Immunophenotype of PTCL, NOS is highly variable but always expresses T-cell antigens (such as CD3, CD2, CD7, and CD5).

Clinically, it had been found that PTCL, NOS is an aggressive disease, which often results in relapse.[3,10,19-21] Our patient also suffered a relapse of the symptoms, and he was then given pralatrexate. He responded well to the medication. The most important elements in the treatment of peripheral T-cell lymphoma NOS are early recognition and appropriate management. While PTCL, NOS carries a significant mortality rate, an early diagnosis of PTCL, NOS and compliance with medications may save a patient's life.

Summary

Peripheral T-cell lymphoma can present with massive lymphadenopathy, which can present as limb swelling. The clinical course of PTCL, NOS is aggressive, and if remission is achieved, relapses are frequent. Pralatrexate and CHOP regimen have been used for the treatment of PTCL with variable success.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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