Upon physical examination the patient was noted to have a well-demarcated, erythematous patch negative to diascopy with fine wrinkling and mild scale. The patch was located over his right proximal leg measuring 37 by 31 centimeters. The distribution of the rash extended from his inguinal ligament location extending distally halfway to the knee and from the posterior medial to posterior lateral locations on his leg across the anterior surface. The patch was non-tender and did not have any further characteristic change including no ulceration (Figure 1).

The patient was originally given a class I topical steroid to apply twice daily for 2 weeks and was instructed to return for a follow-up appointment in 4 weeks. Upon return for the follow up visit the patient’s lesions were unchanged and the decision to perform punch biopsies was made. Two 4 millimeter punch biopsies were obtained and sent to a Dermatopathology lab for H&E review.

Histopathologic examination of the biopsy specimens revealed a superficial and deep perivascular infiltrate of atypical lymphocytes. In addition there was a lichenoid infiltrate of atypical lymphocytes with associated epidermotropism (Figures 2 and 3). Immunohistochemistry showed positive staining for CD2, CD3, CD4, CD8 and CD44 along with negative CD7 staining. Interestingly the staining pattern showed a majority of cells with dual CD4 and CD8 positive markers (Figures 4 & 5). This yielded a diagnosis of Patch Stage Cutaneous T-Cell Lymphoma, Mycosis Fungoides with aberrant co-expression of CD4 and CD8.

The patient was referred to a Hematology/Oncology specialist to obtain a peripheral smear to check for Sézary Cells, which were not found. Subsequently he was given a final diagnosis of Stage IA Cutaneous T-Cell Lymphoma. He declined recommended topical chemotherapy, and radiation therapy options and opted for phototherapy alone combined with topical steroids, which he is currently completing.

Discussion

Cutaneous T Cell Lymphoma represents a group of variable, heterogeneous tumors of the skin. Mycosis fungoides (MF) is considered an indolent subtype of CTCL with a good prognosis when discovered in early stages. MF represents less than 1% of all Non Hodgkins Lymphomas but has the highest skin infiltration incidence among all lymphomas. The incidence has been reported to be around 0.36 per 100,000 people in the United States. Patch Stage MF is commonly overlooked and mistaken for more common conditions such as eczema, psoriasis, parapsoriasis, pityriasis, and others (such was the case with our patient having been misdiagnosed for over 30 years). Due to the indolent course early patch stage MF is well known to require several biopsies over months to years before definitive diagnosis1.

The first known report of dual CD4 and CD8 positive mycosis fungoides was reported by Knapp et al in 2012 involving a elderly female patient2. A second case was reported by Tournier et al in 2014 involving a 31 year old female patient3. Yang and Shen reported a third case in 2015, the first in a male patient4, Knapp reported that the patient presented with sclerodermoid lesions on her abdomen and thigh, while Tournier reported infiltrated plaques in a generalized photo protected areas of the skin5. In Yang and Shen’s case the patient presented in the plaque stage and quickly progressed to systemic disease which resulted in death four months after presentation6. To our knowledge our case is the fourth reported case of this phenomenon and the second in a male patient7,8,9. It is not known if this unique immunologic type has an impact on prognosis or treatment. As more of these cases are reported, future reports will likely uncover these questions and help guide more individualized treatment.

References

