An Unusual Case of IgG4 Associated Marginal Zone B-cell Lymphoma Presenting as Subcutaneous Nodules Mimicking Lipomas

Ryan Mayo MD
Lehigh Valley Health Network, ryan.mayo@lvhn.org

Ranjit R. Nair MD
Lehigh Valley Health Network, Ranjit_R.Nair@lvhn.org

Shereen M F Gheith MD, PhD
Lehigh Valley Health Network, shereen_m.gheith@lvhn.org

Published In/Presented At

Mayo, R. Nair, R. Gheith, S. (2016, Sept). *An Unusual Case of IgG4 Associated Marginal Zone B-cell Lymphoma Presenting as Subcutaneous Nodules Mimicking Lipomas*. Poster Presented at: POSH, Harrisburg, Pa.

An Unusual Case of IgG4 Associated Marginal Zone B-Cell Lymphoma Presenting as Subcutaneous Nodules Mimicking Lipomas

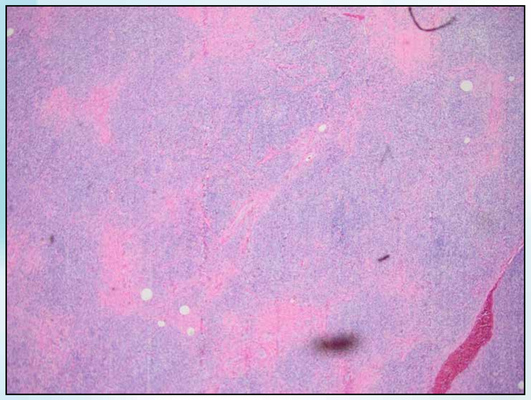
Ryan J. Mayo, MD, Ranjit R. Nair, MD and Shereen Gheith, MD, PhD
Lehigh Valley Health Network, Allentown, Pennsylvania, Health Network Laboratories

INTRODUCTION

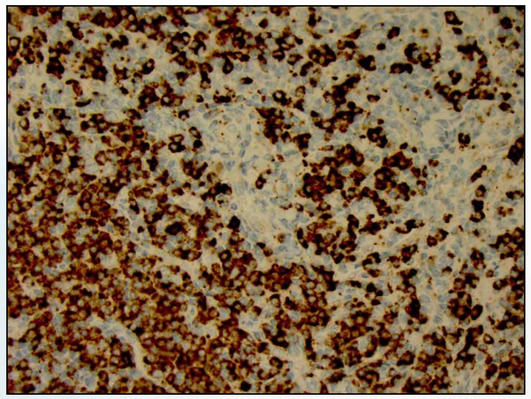
IgG4 related disease (IgG4-RD) are a rare group of immune mediated disorders with heterogeneous clinical presentation but share the same pathologic (lymphoplasmacytic infiltration and fibrosis) and serologic (elevated serum IgG4) features. There are only very few cases of IgG4-RD associated with lymphomas reported in literature.

CASE REPORT

A 55-year-old female presented with increasing pressure and discomfort from long standing, previously asymptomatic, large, bilateral subcutaneous masses involving the upper and lower extremities. She reported multiple excisions in the past with a presumable diagnosis of lipomas. Sonography showed soft hypoechoic masses and given her history and physical examination, a diagnosis of lipomas was favored. An excisional biopsy was performed and the pathology showed a hylanized fibroadipose tissue with extensive lymphoid infiltration with associated prominent internodular plasmacytic component, occasional atrophic germinal centers and frequent scattered eosinophils. CD138 stain showed numerous plasma cells with almost 100% IgG4 coexpression. Additional panel of antibodies demonstrated the plasma cells to be kappa-light chain restricted, consistent with a monoclonal plasma cell population and a diagnosis of low grade marginal zone B-cell lymphoma (MZL) was rendered. The demonstration of IgG4 in neoplastic plasma cells suggested an underlying IgG4-RD with progression to MZL.



This image shows the hylanized fibroadipose tissue with extensive lymphoid infiltration.



The tissue in this image was stained for IgG4 which shows the near 100% co-expression of the immunoglobin.

DIAGNOSTIC STUDIES - TREATMENT - DISCUSSION

DIAGNOSTIC STUDIES:

A serum electrophoresis was performed showing a faint band that the location was indistinct for beta, beta/gamma, or gamma immunoglobin. An immunofixation revealed it as an IgG Kappa monoclonal protein. The serum immunoglobin levels were within normal limits as were the IgG4 subclass level. A PET CT was performed for further staging of the MZL was performed. The results show numerous subcutaneous nodules consistent with the know lymphoma, and FGD avid lymph nodes bilaterally in the axilla, pelvis and inguinal chains.

TREATMENT:

The diagnosis is consistent with an IgG4 associated cutaneous MZL. The prognosis for a cutaneous MZL is excellent, with the disease usually having an indolent course. The 5-year survival is rate has been found to be 98-100%.[1,4] A long term retrospective study showed a median survival of 47 months, with 5 and 10-year survival at 93%.[2] The main stems of treatment for this condition are treatment or excision, with both showing a 99% complete response rate.[4] In this patient after the excision of her lesions, systemic rituximab was added as well. In studies a small number of patients have been treated in this way and have had a good response; 67% complete remission.[3] However, those studies were looking at treatment of MZL with rituximab as initial therapy instead of adjuvant as it is being used in this case.

DISCUSSION:

There are rare reports of MZL arising from IgG4-RD. IgG4-RD presenting as multiple indolent lymphomatous nodules has not been reported in literature. The diagnosis of IgG4-related disease is currently based on certain histologic criteria and is independent of the serologic IgG4 status. The case serves to create awareness about this rare entity and emphasize a low threshold to biopsy, which can lead to successful treatments.

References:

1. Fink-Puches R, Zenahlik P, Bäck B, et al. Primary cutaneous lymphomas: applicability of current classification schemes (European Organization for Research and Treatment of Cancer, World Health Organization) based on clinicopathologic features observed in a large group of patients. *Blood* 2002; 99:800.
2. Hoefnagel JJ, Vermeer MH, Jansen PM, et al. Primary cutaneous marginal zone B-cell lymphoma: clinical and therapeutic features in 50 cases. *Arch Dermatol 2005;* 141:1139.
3. Peñate Y, Hernández-Machín B, Pérez-Méndez LI, et al. Intralesional rituximab in the treatment of indolent primary cutaneous B-cell lymphomas: an epidemiological observational multicentre study. The Spanish Working Group on Cutaneous Lymphoma. *Br J Dermatol* 2012; 167:174.
4. Servitje O, Muniesa C, Benavente Y, et al. Primary cutaneous marginal zone B-cell lymphoma: response to treatment and disease-free survival in a series of 137 patients. *J Am Acad Dermatol* 2013; 69:357.
5. Zinzani PL, Quaglino P, Pimpinelli N, et al. Prognostic factors in primary cutaneous B-cell lymphoma: the Italian Study Group for Cutaneous Lymphomas. *J Clin Oncol* 2006; 24:1376.