Trabajo Presentado en el
V Congreso Venezolano de Dermatología,
Caracas, Noviembre de 1991

LYMPHOMATOID PAPULOSIS

Dr. Lawrence E. Gibson*

Lymphomatoid papulosis is a disorder originally described by McCauley in 1968 as a continuous self-healing eruption, clinically benign; histologically malignant". This disorder is characterized by erythematous papules or nodules which progress to form vesicular crusted or hemorrhagic lesions, and then undergo spontaneous resolution healing with scar. The histology of these papular lesions is similar to lymphoma, resulting in the name lymphomatoid papulosis (LyP). Earlier reports of similar rhythmic eruptions appeared in the literature, but McCauley is given credit for recognizing lymphomatoid papulosis as a unique disorder (McCauley, 1968, Verallo, 1966). Clinically, the lesions of LyP resemble those seen in pityriasis lichenoides acuta (PLEVA, Mucha-Habermann disease). However, the bening-appearing histologic pattern usually serves to distinguish PLEVA from lymphomatoid papulosis (McCauley 1968; Black and Wilson-Jones, 1972).

Willemze in 1982 published a detailed histologic analysis of lymphomatoid papulosis and divided the lesions into two types: type A characterized by large, atypical, histiocytic-appearing cells, strongly resembling Reed-Sternberg (R/S) cells, present mainly in the dermis admixed with varying numbers of inflammatory cells; and a type B infiltrate composed mainly of smaller, cerebriform lymphocytic cells, also admixed with a large number of inflammatory cells often involving the epidermis. (Willemze, et al, 1982: Willemze et al, 1983). It has now been convincingly shown that the infiltrate including the large atypical R/S cells consists mainly of T cells along with inflammatory cells. Characteristic T cell antigens present include CD2, CD3, CD4, with sparse of absent staining with CD7, CD1, and CD8. Most studies also confirm that the large R/S-like cells stain positively with the activation marker ki-1 (CD30). (Kadin, 1990; Burg, 1990; Harrington, 1989).

Studies on the DNA of the cellular infiltrate showed that most of the infiltrate in the LyP lesions are hyperdiploid and tetraploid, aneuploidy being less common (fhomasen and Wantzin, 1987).

T-cell clonal rearrangements in skin lesions from patients with lymphomatoid papulosis have been inconclusive (Weis et al, 1986; Kadin, 1986).

The evidence, thus far, linking lymphomatoid papulosis to lymphoma is still in part circumstantial since 10 to 20% of patients with lymphomatoid papulosis have developed lymphoma (Scheen et al, 1981; Sánchez et al, 1983). Stronger evidence linking lymphomatoid papulosis to lymphoma is based on strong, histomorphologic similarities between the large Reed-Sternberglike cells of lymphomatoid papulosis and those in lymphoma.

This study reports our experience to date with lymphomatoid papulosis, both with clinical follow-up, as well as histopathology and special studies.

We reviewed 53 cases of lymphomatoid papulosis seen at the

* Médico Dermatólogo. Clínica Mayo, Rochester, USA.

Mayo Clinic from 1965 throgh 1990. Thirty-one of these cases had been previously reported (Sanchez et al, 1983). Twenty-nine cases seen from 1983 through 1990 were studied in more detail. There were 37 male and 16 female patients with a male to female ratio of 2.3:1. The man age of onset was 40 years. The duration of individual lesions was to weeks to six months with an average of six weeks.

There were ten patients with onset of lymphomatoid papulosis prior to the age of 20. None of these patients had any significant associated medical problems, and none have developed lymphoma as of this time.

Eight patients have developed lymphoma. Two of these eight patients have died and the others are living. The onset of lymphoma followed lymphomatoid papulosis in all cases.

Follow-up shows that only three patients are currently clear of all lesions. The duration of the disease in the entire group has a mean of 11,9 years, with a mean follow-up period of 12,2 years. Histologically, the dermis showed either a wedge or band-like infiltrate of two types: type A with predominantly large, atypical, lymphocytes with Reed-Sternberg features and type B consisting of small lymphocytes with hyperchromatic cerebriform nuclei. Leukocyte monoclonal antibody studies demonstrated a CD30 staining of the large cells in all type A lymphomatoid papulosis.

Six of the eight patients who developed lymphoma had lymphomatoid papulosis lesions with type A histology.

Patients who have onset of lymphomatoid papulosis prior to the age of 20 tend to have a chronic protacted but benign course. Approximately 12% of all patients and 22% with type A histology who presented with lymphomatoid papulosis after the age of 20 may eventually develop lymphoma.

In conclusion, lymphomatoid papulosis appears to be a reactive skin condition with bizarre atypical cells on histopathology. These cells stain with T-cell markers and with the activation marker CD30. To date neither DNA flow cytometry nor Tcell gene rearrangement studies have been helpful in predicting those 1522% of the patients with Lymphomatoid papulosis who will develop a malingnant lymphoma. Whether lymphomatoid papulosis is considered a reactive process or an immunologically controlled localized malignancy is still not clear. Continued longitudinal follow-up of patients is essential and needs to be correlated with histopathology, lymphocyte, and genetic studies to better characterize this most challenging problem.

REFERENCES

1.- Black MM, Wilson-Jones E: Lymphomatoid pityriasis lichenoides: A variant with histological features simulating a lymphoma. Brit J Dermatol 86: 329-347, 1972.

2.- Burg G, Ziffer S, Lanz U: Light and electron microscopy findings in lymphomatoid papulosis. Proc of first international parapsoriasis symposium, pp 78-81, Mayo Foundation, Rochester, Minnesota.

3.- Harrington DS, Braddock SW, Blocher KS, Weisenberger DD, Savger W, Armitage JO: Lymphomatoid papulosis and progression to T cell lymphoma: An immunophenotypic and genotypic analysis. J Am Acad Dermatol 21: 951-952, 1989.

4.- Kadin ME: Characteristic immunologic profile of large atypical cells in lymphomatoid papulosis. (Editorial) Arch Dermatol 122: 1388-1390, 1986.

5.- Kadin Marshal E: Immunohisthochemistry of lymphomatoid papulosis. Proc of first international parapsoriasis symposium, pp: 56-60, Mayo Foundation, Rochester MN, 1990.

6.- McCauley WL: Lymphomatoid papulosis: A continuing self-healing eruption, clinically bening-histopathologically malignant. Arch Dermatol 97: 23-30, 1968.

7.- Sanchez NP, Pittelkow MR, Muller SA, Banks PM, Winkelmann RK: The clinicopathologic spectrum of lymphomatoid papulosis: Study of 31 cases. J Am Acad Dermatol 8: 8194, 1983.

8.- Scheen Sr III, Doyle JA, Winkelmann RK: Lymphoma-associated papulosis: Lymphomatoid papulosis associated with lymphoma. J Am Acad Dermatol 4: 451-457, 1981.

9.- Verallo VM, Haserick JR: Mucha-Habermann's disease simulating lymphoma cutis. Arch Dermatol 94: 295-299, 1966.

10.- Weiss LM, Wood GS, Trela M, Warnke RA, Sklar J: Clonal T-cell populations in lymphomatoid papulosis. N Eng J Med 315: 475-479, 1986.

11.- Willemze R, Meijer CJLM, Van Vloten WA, Scheffer E: The clinical and histological spectrum of lymphomatoid papulosis. Brit J Dermatol 107: 131-144, 1982.