Dermatology-2014

OMICS Group Conferences
Accelerating Scientific Discovery

4th International Conference on
Clinical & Experimental Dermatology

April 14-16, 2014 San Antonio, USA

Multicentric reticulohistiocytosis and malignancy. a comprehensive review of all reported cases

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Abstract

Multicentric Reticulohistiocytosis (MR) is a rare systemic disease of unknown etiology characterized by mucocutaneous nodules and a progressive destructive arthritis.[1, 2] A Strikingly high percentage of MR cases, 25- 33%, are associated with the presence of an underlying malignancy, leading many authors to believe it represents a paraneoplastic syndrome.[1, 3-5] However, due to the small overall number of cases, wide spectrum of associated malignancies, and observations that MR and the underlying malignancy do not always run parallel courses, this theory has remained debatable.

We have systematically reviewed all reported cases of MR and found 55 cases associated with underlying malignancies. The mean age of patients was 55 years-old with a female-to-male ratio of 1.9. Although a wide variety of neoplasms were found in association with MR, gynecological (26%), breast (19%), respiratory (17%), and GI (11%) neoplasms composed the majority of cases. We examined the clinical, histopathological, and laboratory characteristics in addition to treatments and outcomes in these patients. A very high proportion of malignancies (76%) had metastasized by the time of diagnosis and accounted for a poor outcome in these patients with a 51% mortality rate.

We believe that MR does represent a paraneoplastic phenomenon in a subset of patients; likely arising secondary to release of certain neoplasm-associated cytokines. Evidence also exists supporting osteoclastogenesis (both RANKL-dependent and independent mechanisms) as a potential mechanism for development of MR in these patients. It is possible that immune system-related genetic polymorphisms predispose certain individuals to develop MR in the setting of an underlying malignancy.

Biography

Mazloom has completed his M.D. degree at Chicago Medical School and has done two years of residency training in orthopedic surgery and currently a dermatology research fellow at CCF. He has participated in various basic science and clinical research projects, has published a book, a case report, and a book chapter.