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Abstract

Primary cutaneous marginal zone lymphoma (CMZL) is one of the major primary B-cell lymphomas of skin. Two types are recognized: a more common class-chain switched CMZL, and a much less common IgM+ CMZL. The extremely indolent course, together with other features distinct from most other MALT lymphomas, has led some to question whether at least the class-switched cases should be considered an overt lymphoma.

Key words

primary cutaneous marginal zone lymphoma, B-cell lymphoma, MALT lymphoma, IgG4, skin

Primary cutaneous marginal zone lymphoma (CMZL) is a very indolent lymphoma currently defined as being of "marginal zone" origin and with the neoplastic cells including varying proportions of largely small lymphocytes, plasma cells and intermediate forms. By definition, primary CMZL lacks extracutaneous involvement at diagnosis and does not fulfill the criteria for any of the other small B-cell lymphomas. It is one of the very limited number of distinct primary cutaneous B-cell lymphomas

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