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► **To cite this version:**

Sonja Reitter, Peter Neumeister, Christine Beham-Schmid, Werner Emberger, Dirk Strunk, et al.. A case of generalized MALT lymphoma with IgM paraproteinemia and peripheral blood involvement. *Annals of Hematology*, 2009, 89 (2), pp.213-214. 10.1007/s00277-009-0787-6 . hal-00535076

HAL Id: hal-00535076

<https://hal.science/hal-00535076>

Submitted on 11 Nov 2010

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A case of generalized MALT lymphoma with IgM paraproteinemia and peripheral blood involvement

Sonja Reitter · Peter Neumeister · Christine Beham-Schmid · Werner Emberger · Dirk Strunk · Ruth Brezinschek · Werner Linkesch · Helmut H. Schmidt

Received: 26 May 2009 / Accepted: 24 June 2009 / Published online: 7 July 2009
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Dear Editor,

Extranodal marginal zone lymphomas of mucosa-associated lymphoid tissue (MALT lymphoma) comprise a well-defined entity of non-Hodgkin lymphomas (NHL) but may also share clinical and histopathologic findings with other low-grade lymphomas, thereby causing diagnostic problems. A 35-year-old woman was diagnosed with *Helicobacter pylori* (HP)-positive gastritis and MALT lymphoma in July 2007. Without further staging, HP eradication therapy was initiated; however, control endoscopy after 6 months showed persisting lymphoma, and the patient was referred to our hospital for further evaluation and therapy. She presented with B symptoms, and computed tomography scans revealed a diffuse gastric infiltration of antrum and corpus, enlargement of epigastric lymph nodes, as well as an 8 × 7 cm hypodense liver lesion and multiple opacities in both lungs. All of these lesions showed increased FDG uptake by positron emission tomography scan. Histologic examination of the stomach, liver, and lung lesions confirmed the diagnosis of generalized MALT lymphoma exhibiting patchy infiltrates of small centrocyte-like monoclonal B cells with

interspersed reactive and colonized follicles in the involved organs, forming typical lymphoepithelial lesions in the gastric mucosa with accompanying HP infection. Bone marrow biopsy revealed minimal infiltration with lymphoma cells. Peripheral blood parameters showed a normoquantitative white blood cell count with 50% well-differentiated lymphocytes and 5% lymphoplasmacytic cells and a hypochromic anemia with a hemoglobin level of 9.3 g/dl due to iron deficiency. Flow cytometry (FACS) of the peripheral blood revealed 18% CD19+/20+/sIgM+/5-/10-leukemic lambda-clonal B cells. Serum immunoglobulin analysis showed a monoclonal paraprotein of immunoglobulin M (IgM) lambda type (IgM 11.2 g/l; range 0.4–2.3 g/l). Cytogenetic analysis of the peripheral blood cells revealed normal metaphases; however, interphase fluorescence in situ hybridization (I-FISH) revealed 20% cells showing three signals with probes for the MALT and BCL-2 locus on chromosomal region 18q21 and three signals with a probe for the EVI-1 locus on region 3q26 consistent with duplications or (partial) trisomies of these chromosomal regions. I-FISH with MALT/API2 break apart probes showed no pathologic fusion signals, and consistent with this finding, reverse transcriptase polymerase chain reaction performed on tumor specimens failed to detect MALT-API2 specific RNA fusions, thereby excluding the MALT lymphoma specific t(11;18)(q21;q21). Normal I-FISH signals on the peripheral blood cells with an immunoglobulin heavy chain (IgH) break apart probe excluded the other MALT specific IgH related translocations t(1;14)/BCL10-IgH, t(14;18)/IgH-MALT1 and t(3;14)/FOXP1-IgH. Due to widespread dissemination, chemotherapy according to the R-CHOP protocol was initiated, and after two cycles, the differential blood count and FACS analysis of the peripheral blood showed no pathologic findings; after a total eight cycles, a complete remission was achieved, indicated by histologic examination of bone marrow and stomach biopsy

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samples. During therapy serum concentration of IgM reached normal ranges; however, electrophoresis and immunofixation showed persisting paraproteinemia after therapy. Radiologic investigation revealed decreasing size but still a pathologic FDG uptake of the lesion in the liver. The patient refused rebiopsy and since November 2008, rituximab maintenance therapy is given every second month.

Leukemic dissemination has only sporadically been described in MALT lymphoma and thereby complicated the distinction from other small cell B-cell lymphomas in this case [1, 2]. Similarly, serum paraproteinemia is a well-known phenomenon in various types of NHL, but has so far been regarded as a rare event in MALT lymphoma [3]. However, recent studies indicate a higher incidence of monoclonal gammopathy in series of MALT lymphoma (up to 50% of cases) [4]. Thus, paraprotein analysis should be performed routinely at staging of MALT lymphoma and may also be valuable for monitoring therapy response. The described genetic aberrations (trisomies 3 and 18) are observed in MALT lymphoma as well as in other types of low-grade NHL [3]; therefore, they did not contribute to the differential diagnosis in our case. Whether these aberrations

are associated with leukemic dissemination as well as widespread dissemination of MALT lymphoma remains to be investigated.

References

1. Griesser H, Kaiser U, Augener W, Tiemann M, Lennert K (1990) B-cell lymphoma of the mucosa-associated lymphatic tissue (MALT) presenting with bone marrow and peripheral blood involvement. *Leuk Res* 14:617–622
2. Sahli R, Canioni D, Couturier J, Soler G, Mathiot C, Galatoire O, Lumbroso-Le Rouic L, Puttermann M, Hermine O, Validire P, Morax S, Brousse N, Decaudin D (2008) Bilateral MALT-type ocular adnexal lymphoma with marginal zone lymphoma leukaemic cells and ophthalmological diffuse large B cell lymphoma. *Br J Ophthalmol* 92:579–580
3. Isaacson P, Müller-Hermelink HK, Piris MA et al (2001) Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). In: Jaffe ES, Harris NL, Stein H, Vardimann JW (eds) *Tumours of haematopoietic and lymphoid tissues*. IARC, Lyon, pp 157–160
4. Wöhrer S, Troch M, Streubel B, Hoffmann M, Müllauer L, Chott A, Raderer M (2007) Pathology and clinical course of MALT lymphoma with plasmacytic differentiation. *Ann Onc* 18:2020–2024