CASE REPORT

Primary skin B-cell lymphoma manifested as a sole large nodular lesion

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Abstract

Background: **Primary cutaneous B-cell lymphoma (PCBCL) has only recently been recognized as a distinct clinicopathological entity which is characterized by an expansion of hematopoietic cells in the special microenvironment of the skin with no evidence of extracutaneous disease within the first six months after staging. PCBCLs represent heterogeneous lymphoproliferative conditions comprising 20–25 % of all cutaneous lymphomas.**

A case report: **A Caucasian, sixty five years old female patient with a 4-year history of asymptomatic nodular lesion in the lumbosacral region, firm-elastic to touch and painless, red-purple with smooth surface covered with multiple papules, was referred for the evaluation. Histopathological examination disclosed perivascular lymphocytic infiltrates in the upper part of the dermis, composed of medium-sized lymphocytes with light cytoplasm, especially in deeper dermis margining with fat tissue. Immunohistochemically, the tumor cells were CD20 positive, with a low admixture of CD45RO, CD3 and CD43-positive cells mostly at the periphery of the lesion. The tumor cells exhibited membrane or cytoplasmic expression of immunoglobulins with kappa light chains restriction.**

Diagnosis: **Primary cutaneous marginal-zone B-cell lymphoma (PCMZL). The surgical excision was suggested. The patient is in a complete remission at the 3-year follow-up.**

Conclusion: **The primary B lymphoma is more frequently present then considered. The numerous lesions previously thought to be pseudolymphomas are genuine B lymphomas with a low grade malignancy. It is necessary to consider this condition in the differential diagnosis *(Fig. 5, Ref. 12)*. Full Text (Free, PDF) *www.bmj.sk*.**

Key words: primary cutaneous marginal-zone B-cell lymphoma, immunohistopathology, treatment.

Primary cutaneous B-cell lymphoma (PCBCL) has only recently been recognized as a distinct clinicopathological entity which is characterized by expansion of hematopoietic cells in the special microenvironment of the skin with no evidence of extracutaneous disease within the first six months after staging. PCBCLs represents heterogeneous lymphoproliferative conditions comprising 20–25 % of all cutaneous lymphomas (1, 2, 3). The EORTC classification distinguishes four principal types of PCBCLs: the primary cutaneous marginal-zone B-cell lymphoma (PCMZL) which belongs to the spectrum of mucosa-associated lymphoid tissue lymphomas, follicle-center lymphoma (FCL), diffuse large BCL, leg type and other types, and intravascular large BCL (4, 5, 6). PCMZLs are rare, low-grade lymphomas with good prognosis, with the 5-year survival of 100 % (7). It may occur at any age, but elderly people are more often affected. It usually presents as solitary or multiple asymptomatic papules, plaques or nodules localized mainly on the trunk and extremities. The management of PCBCL requires a staging evaluation to confirm the diagnosis of primary cutaneous lymphoma and to determine the type and the cutaneous extension of cutaneous BCLs. The evaluation includes diagnostic biopsies for histopa-

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Acknowledgement: The work was partially supported by the APVV grant No. 20 025204.

We thank to Mrs. Ivica Uhnavá for an excellent technical help. The work was partially supported by the APVV grant No. 20 025204.

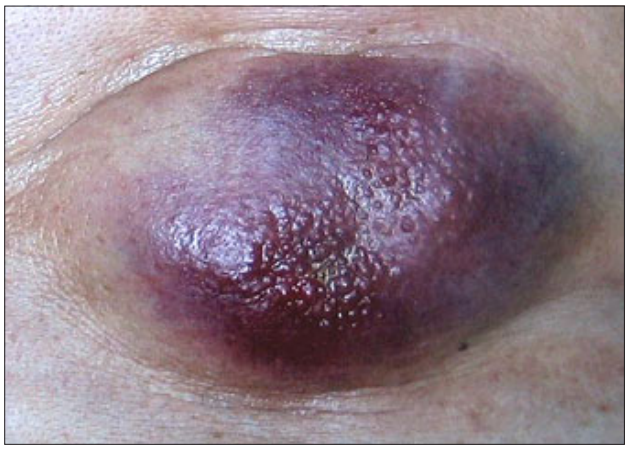

Fig. 1. Nodular lesion 10x8x3 cm large, situated on lumbosacral region is of red-violaceous color with sharp margins. The surface is partly covered with multiple papules 0.3 cm in size.

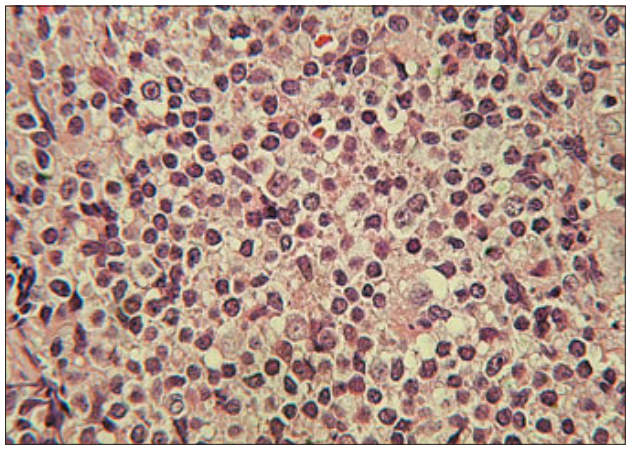

Fig. 3. Compact lymphoid infiltrate nearby the margin of fat tissue composed predominantly of medium-sized lymphocytes with pale cytoplasm (hematoxylin and eosin, HE, original magnification x400).

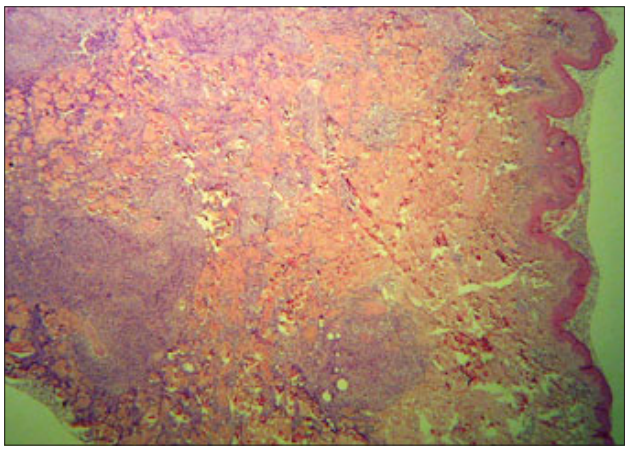

Fig. 2. Lymphoid infiltrate in the upper dermis surrounding the blood vessels, in the deep dermis composed predominantly of medium-sized lymphocytes with pale cytoplasm (hematoxylin and eosin, HE, original magnification x25).

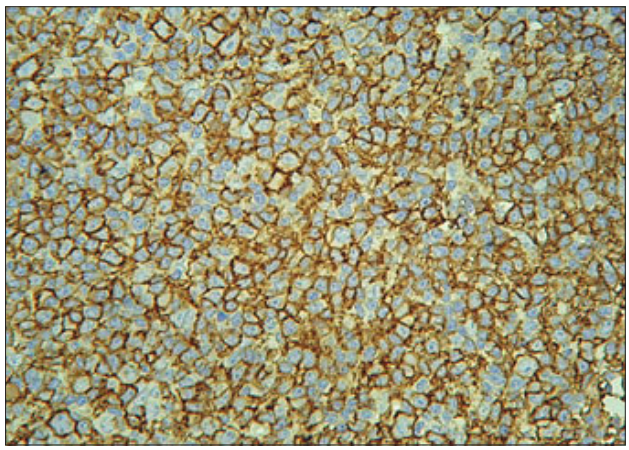

Fig. 4. The tumor lymphocytes showed strong membrane positivity of CD20 antigen (avidin-biotin-horseradish peroxidase complex, ABC-Px, original magnification x400).

thology, immuno-phenotypes, and preferably molecular studies. Currently accepted PCBCL therapies are excision and/or radiotherapy, a intralesional or systemic interferon (INF)-gamma, chemotherapy, and more recently biological agents as monoclonal antibody anti-CD-20 (rituximab).

Case report

In September 2004, Caucasian 65-year-old female patient with a 4-year history of asymptomatic solitary nodular lesion of red-purple color in the lumbosacral region was referred for evaluation. In addition, she presented with multiple fibrolipomas on the back, with a typical clinical presentation confirmed by histopathological examination from skin biopsy. The tumor examination revealed a nodular lesion in the lumbosacral region of red-purple color with sharp borderlines. The smooth surface was partly covered with multiple papules 3 mm in size. The asymptomatic nodular lesion was firm-elastic to touch and painless. The lesion was 10x8 cm at the base exceeding 3 cm of height above the surrounding skin level (Fig. 1). Lymphadenopathy was not evident and other physical findings were unremarkable.

Laboratory findings. Additional investigations were performed to exclude systemic involvement, including routine blood tests, urine analysis, biochemistry, electrophoresis, chest X-ray, with no alterations found except an elevated triglycerides level (3.07 mmol/L, normal range up to 2 mmol/L). No atypical lymphocytes were found in the peripheral blood and bone marrow biopsy showed no invasion of neoplastic lymphocytes. Staging-computed tomographic scans of the chest, abdomen and pelvis and bone marrow proved negative except the hepatopathy with steatosis. Oncological markers CA125, CA 19-9 and CEA were within normal ranges. Metastatic disease of the skin was excluded. Borrelia burgdorferi antibodies titer proved negative by ELISA.

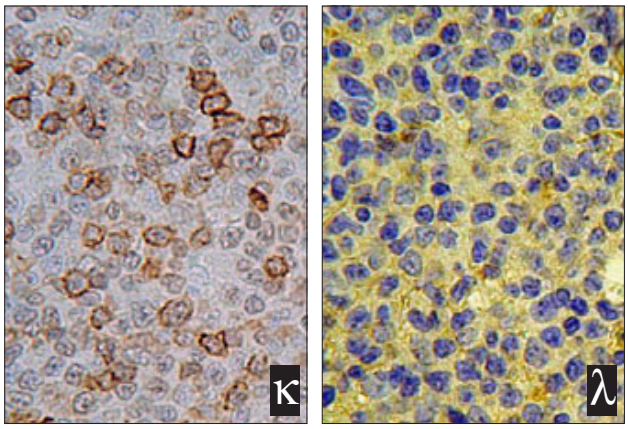

Fig. 5. The lymphocytes exhibit cytoplasmic and membrane monoclonal κ light chains (ABC-Px, original magnification x400).

Histopathological findings. The skin biopsy from the superficial part of the dermis discovered patchy infiltrates with small lymphocytes, at periphery differentiating to medium large lymphoid cells with clear cytoplasm. The second biopsy specimen from the nodular skin lesion in the lumbosacral region revealed dense lymphocytic infiltrate, within the upper dermis surrounding the vessels, composed predominantly of medium-sized lymphocytes with bulky clear cytoplasm propagating to the border with fat tissue, mitotic rate was low (1–2 mitosis/10 fields 40x) (Figs 2 and 3). Immunohistochemically, these atypical neoplastic cells showed CD20 membrane protein expression (Fig. 4). The cells exhibited membrane and/or cytoplasmic expression of immunoglobulins with kappa light chain restriction (Fig. 5). There was a low admixture of CD45RO, CD3 and CD43-positive T-cells more concentrated at the borders of the lesion and in the upper dermis. The lesion was diagnosed as primary cutaneous marginal-zone B-cell lymphoma (PCMZL) of low-grade malignancy.

Treatment. Surgical excision was proposed on the isolated nodule. The site of surgical probationary biopsy developed demarcation in a short time and a deep defect was created at the site of excision. The administration of hydrocolloid paste resulted in complete remission of the defect, which was healed with the atrophic cicatricial skin. The patient is in complete remission at the 3-year follow-up.

Discussion

The diagnosis of lymphoproliferative disorders involving the skin remains one of the most challenging areas in dermatology. Malignant lymphoid neoplasms may involve the skin as the primary process or they present as secondary cutaneous involvement due to the spread of lymphomas or leukemias. In addition, there is a variety of cutaneous pseudolymphomatous reactions. The conditions may have many histological, immuno-phenotypic and genetic features in common. The diagnosis and establishment of the biologic nature of the process is therefore crucial. PCBCL is defined by the absence of an extracutaneous disease both in the initial staging procedure and within the period of 6 months (8). In the presented case report the diagnosis of PCMZL was based on the histological and immuno-histochemical examinations. The prognosis is excellent with the 100 % survival at 5 years. Thus, radiotherapy or surgical excision remain the treatment of choice either alone or combined in solitary lesions. Monochemotherapy with chlorambucil in patients with diffuse cutaneous lesions may obtain approximately 50 % rate of an complete remission (9). Intralesional injections of IFN- may be an alternative therapy. A new approach in the treatment of PCBCLs is the use of a monoclonal antibody that targets the CD20 antigen (rituximab) as intralesional therapy in isolated or small number of lesions (10). Retrospectively evaluated study of 66 patients with PCBCL of the Sheffield lymphoma group revealed the surgical excision as a modality of treatment in 33 % (11). Surgical excision was used also in the patient, who was the subject of this case report. A deep defect after the surgical excision was covered after 8-week administration of hydrocolloid paste with a cicatrial scar. Hydrocolloid wound dressing is an interactive therapy, which provides a moist wound environment that supports the natural healing process. On absorbing wound exudate, the dressing forms gel, which fills the wound cavity and protects granulation tissue and nerve endings. It could be used not only in ulcer of common origin but also in cutaneous ulcer of unusual causes due to malignancy, hematologic disorders, vasculitis, sarcoidosis, calciphylaxis, Buerger's disease, and pyoderma gangrenosum (12).

Conclusion

The diagnosis of a lymphoproliferative process in the skin is often a challenge for clinicians and dermatologists. Histological assessment is only one step in the diagnosis in a patient with lymphomatous condition. The diagnosis results from a close clinicopathological correlation achieved by the co-operation of all specialists involved in patient's management. The primary B lymphoma may be present more frequently then it is thought. Numerous lesions previously considered to be pseudolymphomas are determined as B lymphomas of low-grade malignancy. It is important to consider this eventuality in the differential diagnostics.

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Received August 21, 2007.
Accepted September 21, 2007.