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Case Report Open Access

Littoral Cell Angiomatosis: Splenic Lesions in a Patient with Follicular Non Hodgkin Lymphoma

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Case Report

A 67 year old patient was diagnosed with grade I extranodal follicular lymphoma of the parotid gland. She suffered from a painless swelling of the right parotid gland. No other symptoms, especially no B-symptoms were present on admission. The physical examination as well as the laboratory study was all normal. Histological, immunhistochemical and molecular genetic workup of the parotid gland showed an infiltration by a population of small CD20- and CD10-positive clonal B-cells, consistent with an extranodal follicular Non Hodgkin Lymphoma (FL), grade I. The staging examinations (CAT scan, bone marrow trephine) showed no further lymphoma involvement apart from multiple hypoattenuating lesions in the spleen suspicious of a splenic infiltration of the lymphoma (Figure 1, white arrow). Since the treatment approach for a limited stage IEA (FLIPI 1) FL is curative, whereas a more extensive Ann Arbor stage [1] IIIEA (FLIPI 2) with spleen involvement is palliative, splenectomy was performed to define the exact stage of the disease. The histological workup showed multiple anastomosing neo-vascular channels lined by cells which have endothelial and histocytic (CD 68) properties, indicative of a littoral cell angioma (LCA) of the spleen (Figure 2 white arrow) beneath physiological lymphoid follicles (Figure 2, black arrow). The LCA is a rare splenic vascular tumor that derives from littoral cells which cover the splenic sinus channels of the red pulp. It was first described in 1991, normally shows a benign behavior and can be associated with solid and haematological neoplasias. Characteristic, but not specific CAT-findings are multiple defects in the spleen with low contrast. To exclude malignant primary splenic entities such as an angiosarcoma, splenic metastases, or, as suspected here, a lymphoma infiltration, a splenectomy is advisable in most cases [2]. In our patient, a splenic involvement of the spleen could be ruled out, and she underwent an involved field radiation and is still in remission 5 years later.

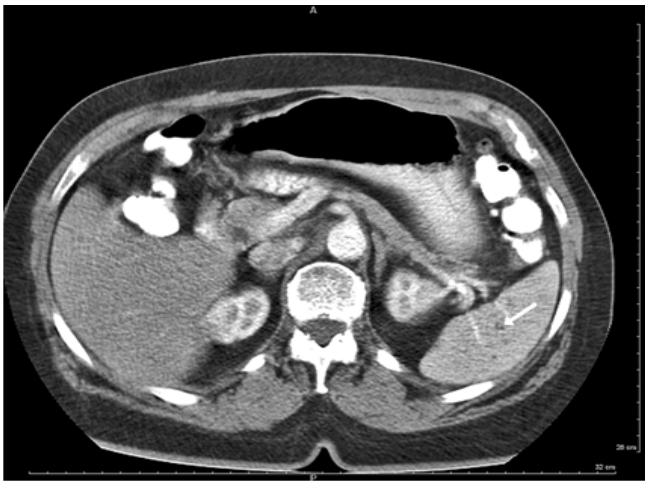

Figure 1: CT scan with multiple hypoattenuating splenic lesions (white arrow).

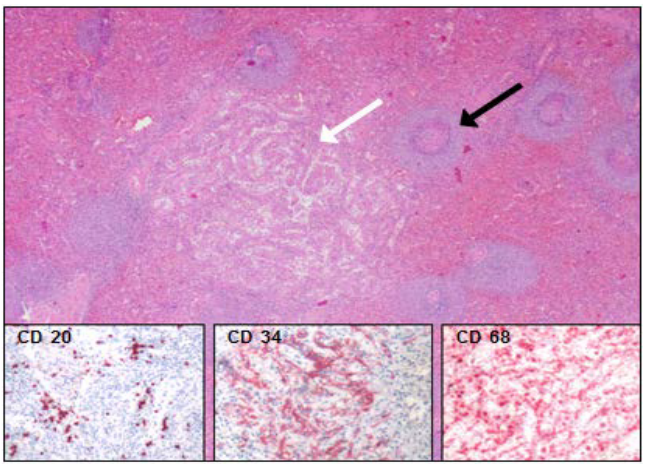

Figure 2: Hematoxylin-eosin stain (x10) with physiological lymph follicles (black arrow) and littoral cell angioma (white arrow) which is CD20 negative and CD34 and CD68 positive.

References

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