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LETTER TO THE EDITOR

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Fibrin-associated diffuse large B-cell lymphoma in a hemorrhagic cranial arachnoid cyst

Daniel Kirschenbaum¹ , Peter Prömmel², Flavio Vasella¹, Eugenia Haralambieva³, Ewerton Marques Maggio³, Robert Reisch², Marc Beer³, Ulrike Camenisch³ and Elisabeth J. Rushing^{1*}

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Arachnoid cysts are a common incidental finding on magnetic resonance imaging (MRI) performed for other clinical reasons. They can be found in the brain or spine and are mostly of congenital origin due to splitting of the arachnoid membrane. The vast majority are asymptomatic, with signs and symptoms varying according to size and location. Intracystic hemorrhage is a rare complication. In symptomatic cases, treatment predominantly consists of endoscopic fenestration [4, 5, 7].

In recent years, the features of diffuse large cell B-cell lymphoma (DLBCL), referred to as fibrin-associated DLBCL, have been reported in cases of chronic blood effusions [1]. Fibrin-associated DLBL, which has a favorable clinical outcome, should be distinguished from chronic inflammation-associated DLBCL, which is an aggressive tumor. Fibrin-associated DLBCL has been described in cases throughout the body; however, only isolated intracranial cases have been described, which were found in the subdural space. Here we present the case of an elderly man with an unsuspected fibrin-associated DLBCL in an arachnoid cyst. We would like to draw attention to this entity, which has likely been underestimated in the routine evaluation of subdural hematoma (SDH) or subarachnoid cysts.

An 81-year-old man presented with intermittent tremor and gait ataxia. The patient was diagnosed with classical parkinsonism and L-Dopa treatment was started. Later, the patient developed short-term memory disturbances and the gait ataxia progressed. Based on the MRI findings, a right frontotemporal arachnoid cyst with focal bleeding was suspected. Intraoperatively, the

lesion presented as an arachnoid cyst filled with thick, whitish fluid reminiscent of empyema. The cyst was washed out and the lining of the cyst was resected (Fig. 1). The postoperative course of the patient was uneventful with good recovery of the neurological status. After the pathological diagnosis was rendered, a whole-body PET-CT showed no other lesions. Due to the relatively advanced age of the patient, therapy with rituximab and lenalidomide was initiated.

Hematoxylin-eosin-stained sections (Fig. 2a) revealed small, discohesive islands of large atypical cells against a background of abundant fibrin, without evidence of a large mass-forming lesion. On immunohistochemistry, the atypical cells were strongly CD20 (Fig. 2b) positive. The cells showed high proliferative activity with multiple mitotic figures and a Mib-1 proliferation index of over 80% (Fig. 2c). Immunohistochemical preparations were strongly positive for CD30, Bcl2 (Fig. 2d) and IRF-4 (nuclear, Fig. 2e), with only focal positivity for BCL6. In addition, CD5 immunolabeling was detected in scattered non-atypical cells, which were small reactive T-cells in contrast to the large atypical cells. CD10, TdT, pancytokeratin and melanocytic markers were negative. In situ hybridization showed the presence of non-coding Epstein-Barr virus (EBV)-associated RNA in the majority of the atypical cells (Fig. 2f). C-MYC immunohistochemistry showed expression in more than 50% of the cells. No C-MYC-rearrangement was detected by fluorescent in situ hybridization. A PCR based analysis revealed a monoclonal rearrangement in the *IgH* Gene.

Taken together the diagnosis of a fibrin associated EBV-positive large-cell B-cell lymphoma was rendered.

Diffuse large-cell B-cell lymphomas comprise a group of relatively common hematological malignancies.

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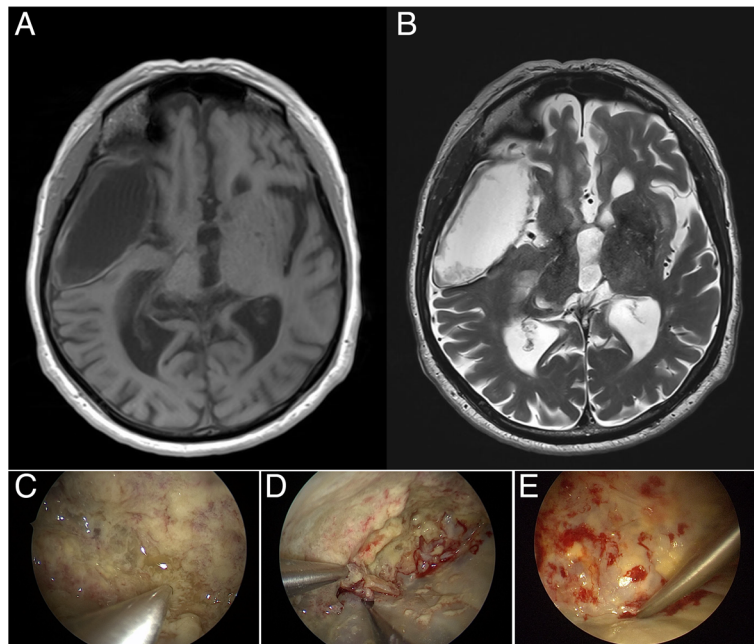


Fig. 1 T2-weighted MRI with right temporal arachnoid cyst with signs of intracystic hemorrhage (a). Postoperative control-MRI shows total resection of the tumoral tissue within the cyst (b). Initial intraoperative endoscopy showed a thick, yellowish lining of the inner wall of the cyst, (c) which was removed under endoscopy (d) with eventual total resection (e)

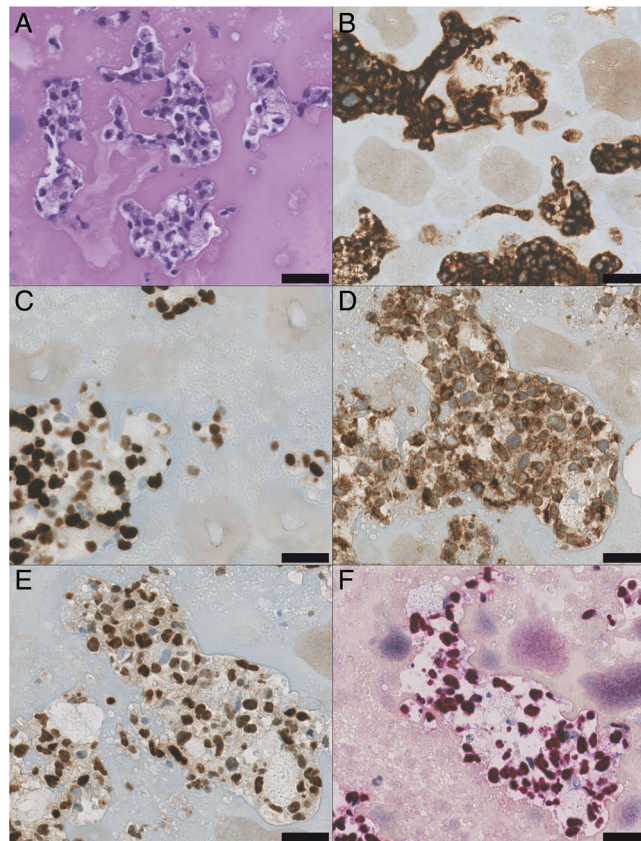


Fig. 2 Microscopic pathology. Islands of atypical blastic lymphoid cells are embedded in abundant fibrin (a). The cells are diffusely positive for CD20 (b), highly proliferative in the Mib-1 (c) stain and show strong positivity for BCL2 (d), IRF-4 (e) and EBV-associated RNA (f). Scale bars represent 30 μ m

DLBCL may be associated with chronic inflammation, now considered a rare EBV-associated subtype in immunocompetent individuals. For the most part, DLBCL with chronic inflammation has an unfavorable prognosis. EBV-positive DLBCL have also been reported as an incidental finding in the setting of chronic hematomas, atrial myxomas and pseudocysts [1]. The median age is 55.5 years with a male: female ratio of 3:1. All cases have been incidental findings, were EBV associated and had an invariably favorable prognosis. Although these lesions have been described in distinct locations, the histology is remarkably similar, with atypical B-cells embedded in a fibrinous background. As reviewed by Boyer et al., three cases of fibrin-associated DLBCL have been documented in SDH found in patients with a median age of 66.5 years and a male: female ratio of 4:0. An additional new case was reported by Boyer et al. All except one of these cases showed similar histology, prognosis and an EBV-association [1, 2, 6]. One of the cases was considered a primary lymphoma presenting as a chronic subdural hematoma [2]. We report for the first time a fibrin-associated DLBCL in an arachnoid cyst with hemorrhage. In rare cases, arachnoid cysts may harbor either primary or metastatic tumors [3]. In the present case, the diagnosis of an arachnoid cyst was based on radiological and intraoperative assessment. Microscopically, there was no evidence of an arachnoid membrane, which could represent a sampling error. Although the radiographic and intraoperative appearance was virtually pathognomonic of an arachnoid cyst, the microscopic appearance could also be interpreted as a resorbed SDH with recurrent bleeding. In most cases, surgery appears to be curative. In the current case, however, rituximab and lenalidomide therapy was administered. Greater awareness is important in order to more accurately assess the natural history of this entity.

Abbreviations

DLBCL: Diffuse large-cell B-cell lymphoma; EBV: Epstein-Barr virus; MRI: Magnetic resonance imaging; SDH: Subdural hematoma

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Not applicable.

Availability of data and materials

The datasets generated and/or analysed during the current study are not publicly available due being protected patient information but are available from the corresponding author on reasonable request.

Authors' contributions

DK and EJ.R analyzed the pathology, made the microscopic photographs and wrote the manuscript. FV, UC, EH, MB, EMM analyzed the pathology. PP and RR were the operating surgeons. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The patient provided consent for publication. No applicable ethical concerns.

Consent for publication

The patient provided consent for publication.

Competing interests

The authors declare that they have no competing interests.

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