



## Extracavitary primary effusion lymphoma (PEL) presenting as bilateral adrenal gland involvement in an HIV-positive patient

Mitja Nabergoj<sup>1</sup> · Olivier Nawej Tshikung<sup>2</sup> · Olga Tsopra<sup>1</sup> · Ana Ghinescu<sup>3</sup> · Kaveh Samii<sup>1</sup> · Alexandra Calmy<sup>2</sup>

Received: 29 August 2018 / Accepted: 2 October 2018 / Published online: 17 October 2018  
© Springer-Verlag GmbH Germany, part of Springer Nature 2018

Dear Editor,

A 44-year-old man with human immunodeficiency virus (stage C2), treated with antiretroviral therapy (tenofovir/emtricitabine/atazanavir), presented to the emergency department with fever, abdominal pain, and a history of weight loss over 2 months. The physical examination was unremarkable. The radiological investigation by CT (Fig. 1a) revealed large bilateral adrenal gland masses with invasion of the left renal vein, highly 18F-fluorodeoxyglucose-avid on PET/CT (Fig. 1b, SUV max 25). Laboratory investigations confirmed adrenal insufficiency and a CD4 count at 525 cells/ $\mu$ l. The HIV-RNA value was 1400 copies/ml.

A CT-guided biopsy showed a massive infiltration of the adrenal gland by an atypical medium to large lymphoid proliferation, with focal plasmacytoid differentiation (Fig. 1c). By immunohistochemistry, the cells were negative for most B cell markers (CD20 (Fig. 1d), CD79a, PAX5, and kappa/lambda light chains), showing positivity for MUM1/IRF4, with more weak and focal expression of VS38C and CD138. A strong

and diffuse aberrant expression of the T cell marker CD3 was observed, whilst other T cell markers were negative. The cells showed strong nuclear positivity for HHV8 (Fig. 1e) and approximately 30% were positive for EBV by in situ hybridisation for EBV-encoded small RNA. The proliferation index (MIB1) was 90%. Subsequent molecular biology analysis revealed rearrangement of the immunoglobulin genes, compatible with a B cell derivation of this lymphoma, with no T cell clonality. The morphologic and immunophenotypic findings led to the diagnosis of a HHV8+, EBV+ primary effusion lymphoma.

Primary effusion lymphoma (PEL) is a rare entity, and it is mostly observed in middle-aged HIV infected men who have sex with men, where it represents barely 1 to 4% of all HIV/AIDS-associated NHLs [1], but also in elderly HIV-negative patients from both sexes from HHV8-endemic areas [2]. The extracavitary PEL could be associated with an effusion or could present as a stand-alone mass and can be localized in nodes, lungs, central nervous system, skin, spleen, bone marrow, liver, and gastrointestinal tract [3].

The illustrated case is, to the best of our knowledge, the first report of an extracavitary PEL with bilateral adrenal gland involvement in the absence of serosal involvement, nor other sites of disease detected at the time of staging. The patient was treated with four cycles of DA-EPOCH with partial response and subsequently with four cycles of DHAP and a mobilization cycle with cyclophosphamide, obtaining a complete response. Despite the patient's refusal to proceed to autologous stem cell transplantation, he is still in complete remission 2 years after his initial treatment, requiring only replacement therapy for residual adrenal insufficiency.

✉ Mitja Nabergoj  
mitja.nabergoj@hcuge.ch

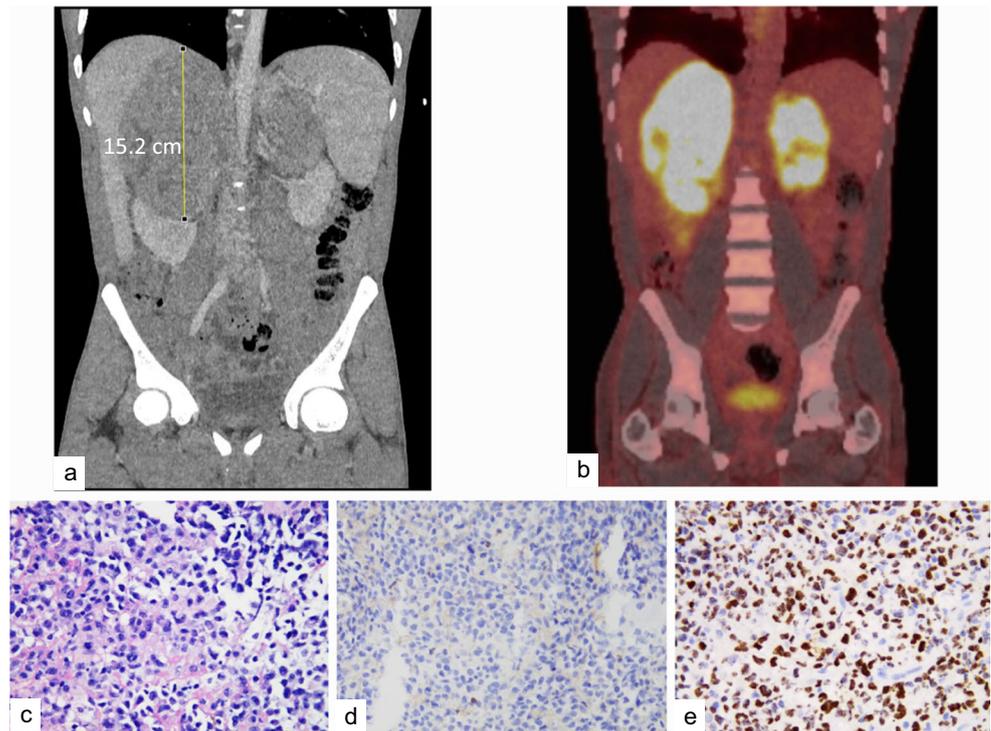
✉ Alexandra Calmy  
alexandra.calmy@hcuge.ch

<sup>1</sup> Division of Hematology, Department of Oncology, Geneva University Hospital, Rue Gabrielle-Perret-Gentil 4, 1205 Geneva, Switzerland

<sup>2</sup> Division of Infectious Diseases, HIV Unit, Department of Internal Medicine Specialties, Geneva University Hospital, Rue Gabrielle-Perret-Gentil 4, 1205 Geneva, Switzerland

<sup>3</sup> Department of Pathology and Immunology, Geneva University Hospital, Geneva, Switzerland

**Fig. 1** **a** CT scan showing bilateral adrenal gland involvement by two large adrenal masses; **b** PET/CT, same adrenal tumors showing a high SUV uptake; **c** Histologic specimen of biopsied tissue stained with hematoxylin and eosin (H&E) showing large and pleomorphic cells with a plasmacytoid appearance; **d** staining for CD20, diffuse negativity of cells for CD20; **e** staining for HHV8 showing a robust nuclear positivity for HHV8



### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no competing interests.

### References

1. Chen YB, Rahemtullah A, Hochberg E (2007) Primary effusion lymphoma. *Oncologist* 12(5):569–576
2. Klepfish A, Sarid R, Shtalrid M, Shvidel L, Berrebi A, Schattner A (2001) Primary effusion lymphoma (PEL) in HIV-negative patients—a distinct clinical entity. *Leuk Lymphoma* 41(3–4):439–443
3. Guillet S, Gerard L, Meignin V, Agbalika F, Cuccini W, Denis B et al (2016) Classic and extracavitary primary effusion lymphoma in 51 HIV-infected patients from a single institution. *Am J Hematol* 91(2): 233–237