

**Figure 3** (A) In common with the ovarian follicle granulosa cells, the adjacent cell clusters stain strongly positive with  $\alpha$ -inhibin. (B) The cells fail to stain with the epithelial marker CAM 5.2.

and attention to the characteristic location of the granulosa cells.

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## Primary effusion lymphoma (PEL) without effusion: a patient case report of a PEL solid variant

Primary effusion lymphoma (PEL) is a very rare type of lymphoma that is usually confined to the body cavities such as the pleural space, pericardium and peritoneum.

PEL is associated with human herpes virus-8 (HHV-8) infection and commonly observed in HIV-infected patients.

We present a case of PEL confirmed by pathology without effusion in a 38-year-old man at initial HIV diagnosis.

A 38-year-old man presented with a right axillary swelling, restricted arm movement and chest pain for 3 weeks. He reported weight loss of 4 kg and night sweat without fever and/or chills. On physical examination, we found enlarged neck and axillary lymph nodes with a diameter of 3 cm, movable and hard presentation without tenderness. Elevation of the right shoulder was restricted to 20° with intact circulation and sensitivity.

Lab reports showed a haemoglobin value of 7.8 g/dl, albumin 3.4 g/dl, lactate dehydrogenase 255 U/l and creatinine 1.6 mg/dl. Total serum protein was elevated to 10.3 g/dl and the fraction of  $\gamma$  globulins was expanded to 44% without monoclonal globulin. In a 24-hour urine collection test, mild proteinuria (0.52 g/24 h) with  $\kappa$  and  $\lambda$  light chain elevation (278 and 80.2 mg/l, respectively) was present, while immunofixation electrophoresis of the serum was negative.

MR and CT scans of the right shoulder showed enlarged axillary lymph nodes with 3 cm diameter infiltrating medullary space of the caput and corpus humeri without destructing the corticalis (figure 1A). In the CT scan, mild enlarged lymph nodes in cervical, inguinal and iliac sites (1.5–3.3 cm) as well as an enlarged spleen 14×4.5×12 cm were found.

Lymph node extirpation of the right axillary bulk could be performed and gross pathological examination showed a conglomerate with two discernable grey-coloured nodules.

First, the lymph nodes showed the histopathology of acute to subacute HIV lymphadenitis with lymphofollicular hyperplasia with reactive hyperplastic germinal centres and extensive accumulation of plasma cells

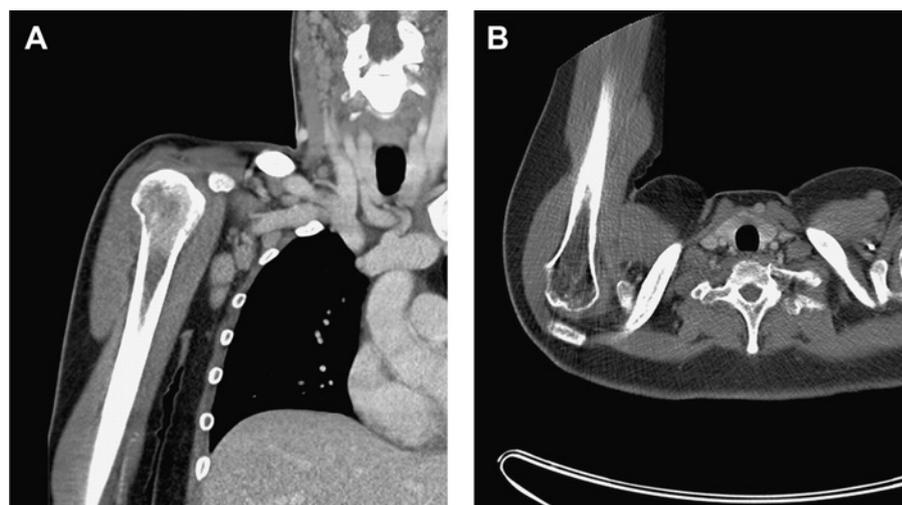
(CD20+, CD79a+, MUM1+, p24-antigen+, EBV+). Second, a predominantly intrasinusoidal (subcapsular and medullary) infiltration was seen, which might represent a very early infiltration of the lymph node, by large blastic cells with large nuclei and prominent nucleoli (figure 2A). In immunohistochemical studies, these blastic cells strongly expressed CD30, CD138, MUM 1 and HHV-8 and were found negative for CD20, CD45, CD79a, PAX 5, CD56 and ALK. Immunoglobulin light chain  $\kappa$  and  $\lambda$  showed polyclonal expression. Additionally, an Epstein–Barr virus-encoded RNA in situ hybridisation was strongly positive (figure 2B–D). After consulting the Reference Center for Lymph Node Pathology (University of Wuerzburg), a PEL was diagnosed.

HIV and hepatitis serology revealed a positive HIV-1 immunoblot and RT-PCR with 19 000 copies/ml. The CD4 cell count was 390/ $\mu$ l.

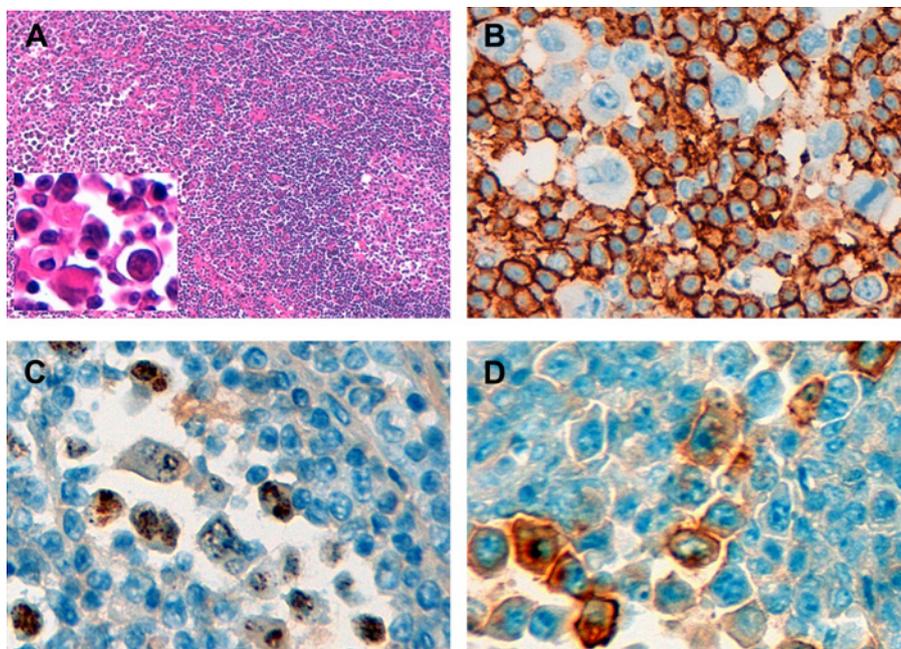
Due to unique clinical presentation of PEL with lymphomatous growth and without tumour cells in effusions, the patient underwent cardiac and abdominal ultrasound imaging; however, the pericardial, pleural and peritoneal spaces were completely free of effusions.

Antiretroviral therapy with nevirapine, lamivudine and abacavir was initiated, but the patient experienced significant progressive disease with tumour growth at all sites and with ongoing destruction of the humerus.

Chemotherapy with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) was administered, and after three cycles, staging CT scans showed partial remission. Moreover, creatinine and lactate dehydrogenase showed normal results (0.9 mg/dl and 196 U/l) and haemoglobin increased to 13.3 g/dl. After another three cycles, the patient achieved complete remission in staging CT scan (figure 1B).



**Figure 1** CT imaging of the right shoulder at initial diagnosis (A) and after six cycles of chemotherapy/HAART (B). (A) Right shoulder with axillary lymph nodes infiltrating caput and corpus humeri. (B) Normal finding of the right shoulder and axillary lymph nodes.



**Figure 2** Histology and immunohistochemistry. (A) Lymph follicle on the right, atypical cells left in between (HE, 100×/400×). (B) Negativity for CD20 (CD20, 400×). (C) Positivity for HHV-8 (HHV-8, 400×). (D) Positivity for CD30 (CD30, 400×).

PEL was first described in 1989. Since then, its unique pathogenesis, the role of HHV-8 and EBV infection have been elucidated.<sup>1</sup> Currently, only very few longitudinal observational series of patients with PEL have defined optimal treatment strategies.<sup>2</sup> PEL solid variants (without effusion) are very rare with only a few cases reported in literature. A retrospective series of four cases with solid variants have been reported by Carbone *et al*. Despite treatment with CHOP chemotherapy and highly active anti-retroviral therapy (HAART), all patients died of disease progression within 15–55 days.<sup>3</sup>

As described by Simonelli and Boulanger *et al*, the absence of HAART before PEL diagnosis is the strongest predictor of poor clinical outcome in the population of HIV-infected patients with PEL.<sup>4,5</sup> This may be related to the reversion of severe immunodeficiency by HAART and therefore enhanced tolerance to chemotherapy

toxicity and reduction of treatment-related mortality. Control of HIV replication and immunological recovery could also slow PEL progression.<sup>5</sup> As reported by Carbone *et al*, Kaposi's sarcoma-associated herpesvirus/HHV-8 (KSHV)-positive solid lymphomas show a PEL-specific profile in the gene expression levels.<sup>3</sup> Patients who develop PEL may be more immunosuppressed than patients with the KSHV-positive solid variant.<sup>6</sup> In a small series of KSHV-positive solid lymphomas, eight patients with solid variant PEL had a slightly better survival rate compared with 21 cases with PEL.<sup>6</sup>

Two major laboratory abnormalities, anaemia and hypoalbuminaemia, have been described in PEL solid variant individuals.<sup>3</sup>

This is a very rare description of a solid variant of PEL without effusion, both at initial diagnosis and in the later course of the disease. The case also accentuates the necessity of molecular testing for HHV-8 even in the absence of the 'classical clinical features' in high-risk patients. CHOP with or without granulocyte colony-stimulating factor support may be a candidate for safe and effective treatment in these patients.

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**Contributors** CG, DDG, BH and BS collected the medical history and carried out clinical investigations of the case. CG and HO wrote the manuscript. AG, HRD, DM, TK, AR, RF, VH and JRB were extensively involved in the clinical and outpatient management of the patient and revised the manuscript. DM and AR provided expert pathological advice and contributed to the pathology aspects of the case report. JRB provided expert infectious diseases advice and contributed to the HIV-associated lymphoma aspects of the case report.

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## Benign phyllodes tumour with intraductal papillary growth of the breast in a young woman

A 24-year-old woman noted a mass in her left breast and visited a local hospital. The mass was ~2.5 cm in size upon imaging and considered to be a benign tumour, so she was monitored. Six months later, the mass enlarged to >3 cm, showed redness in the skin and was painful. She took antibiotics for 1 week, and symptoms improved. She was referred to the Social Insurance Kurume Daiichi Hospital (Kurume, Japan). Ultrasonography revealed an intracystic tumour measuring ~3.3×3.3×3.5 cm consisting of a solid papillary structure in a cystic lesion and with an inhomogeneous interior (figure 1). Dynamic MRI revealed an irregular-shaped solid portion, with enhancement within the

## Key messages

- ▶ Primary effusion lymphoma (PEL) is rarely found in immunocompromised patients.
- ▶ HHV-8 immunohistochemical testing should be applied even in absence of 'classical clinical features'.
- ▶ Highly active anti-retroviral therapy (HAART) in combination with CHOP regimen chemotherapy is a feasible option for patients with PEL solid variant.



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