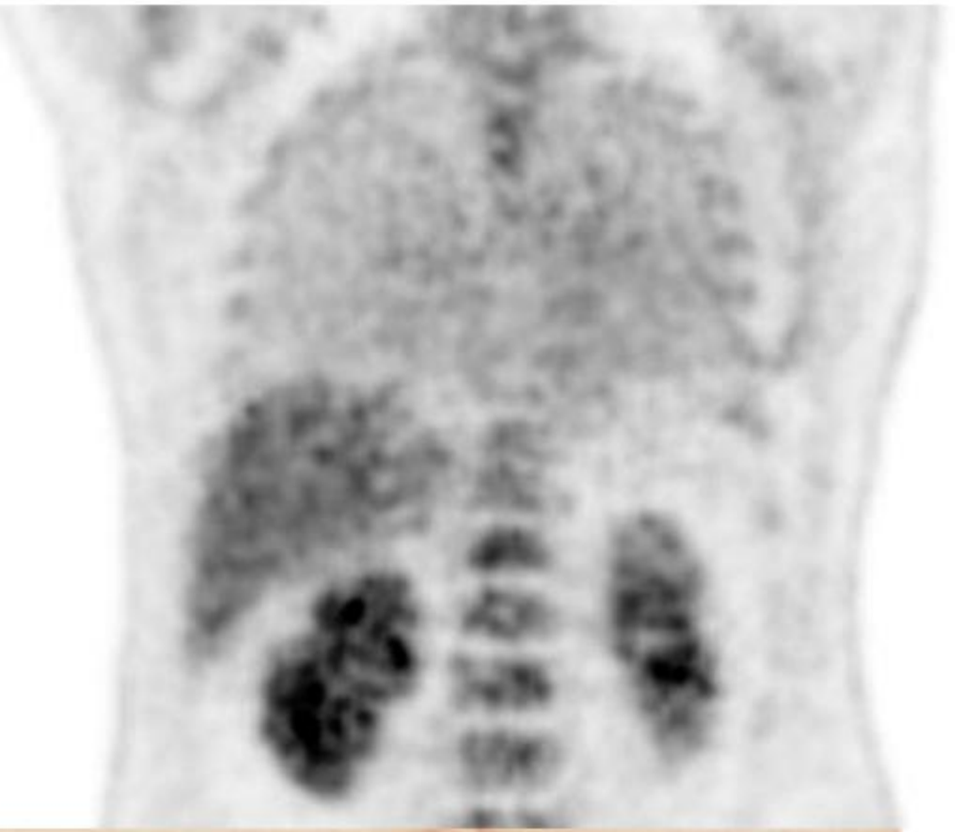
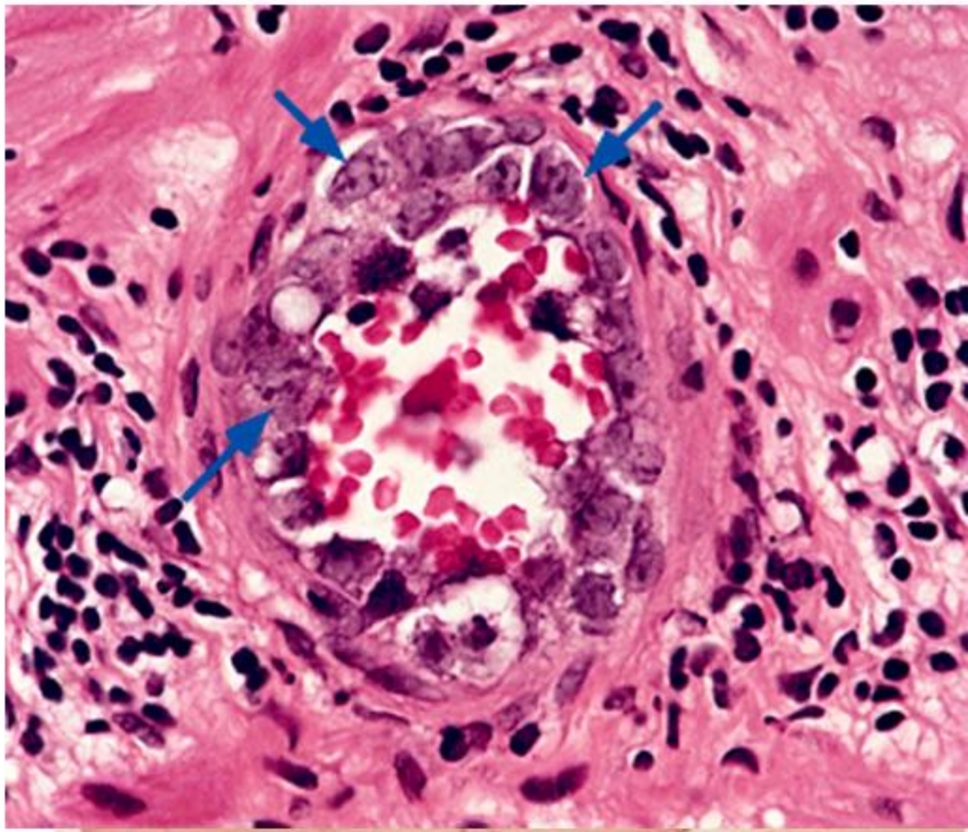
Intravascular large B-cell lymphoma (IVL)





58-year-old man.

Symptoms of stroke, weakness in the left hand and epileptic seizure (grand mal).
Also, 3-month of recurrent fever.

Normal tests:

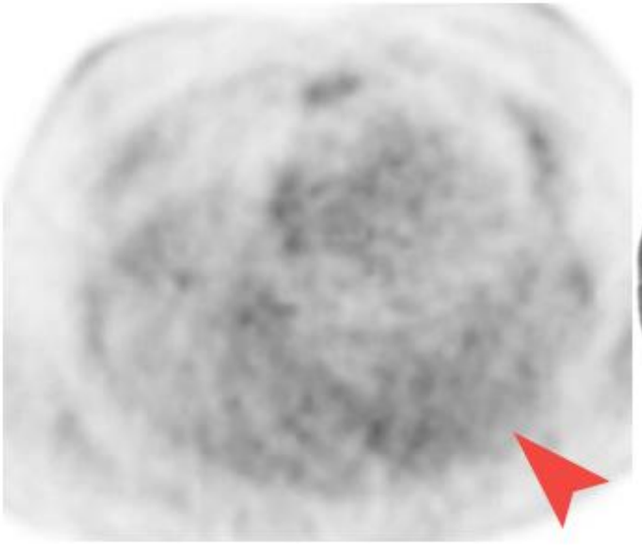
Cerebral CT and MR.
EEG and LP.
Physical examination.
Laboratory studies.
Blood cell counts.
Chest radiography.
Tests for viruses, bacteria and autoimmune diseases.
Bone marrow biopsy.

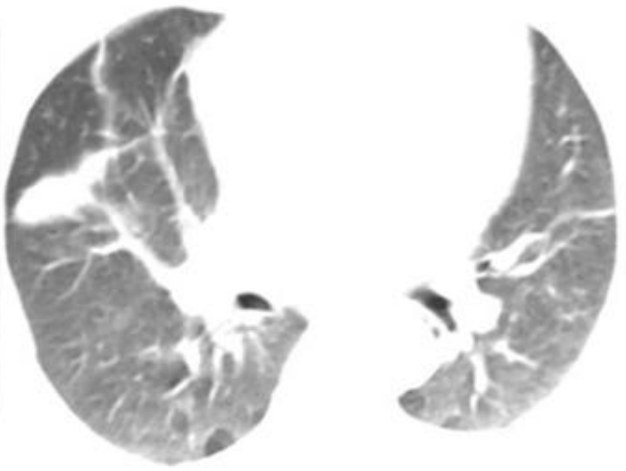
Abnormal tests:

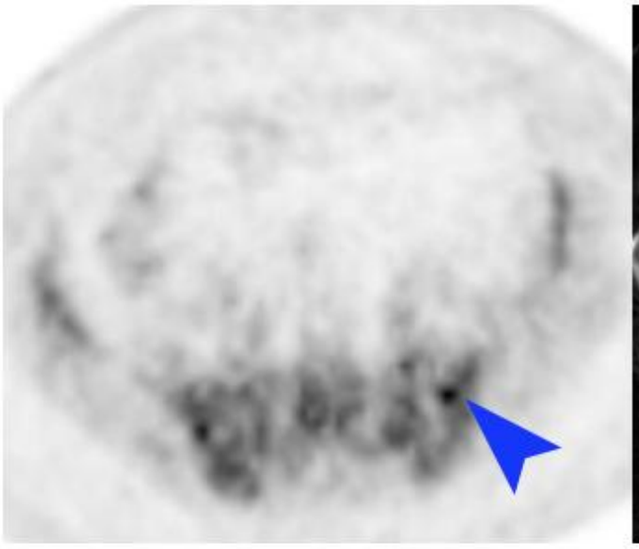
Elevate LDH at 4000 U/L
Hb level of 7.0 g/dL.
FDG PET/CT:
- diffuse uptake in the bilateral lung,
- slightly heterogeneous uptake in the bone marrow,
- slightly elevated uptake in the renal cortex.

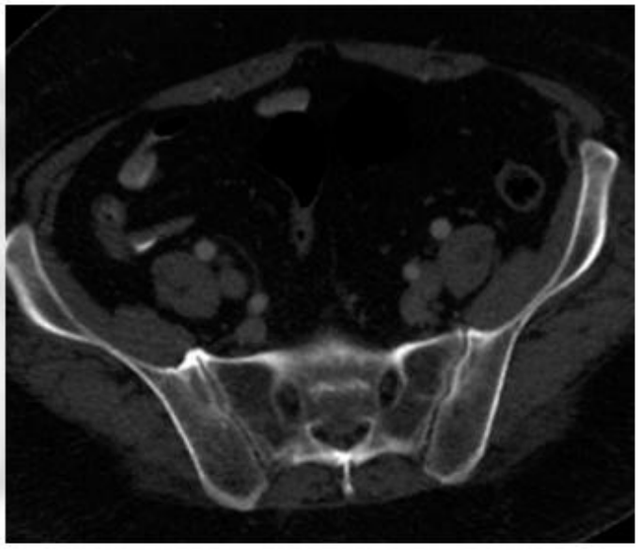
Skin biopsy:

CD20+ large lymphoid cells filling the small vessels in the subcutaneous tissues.
IVL was diagnosed





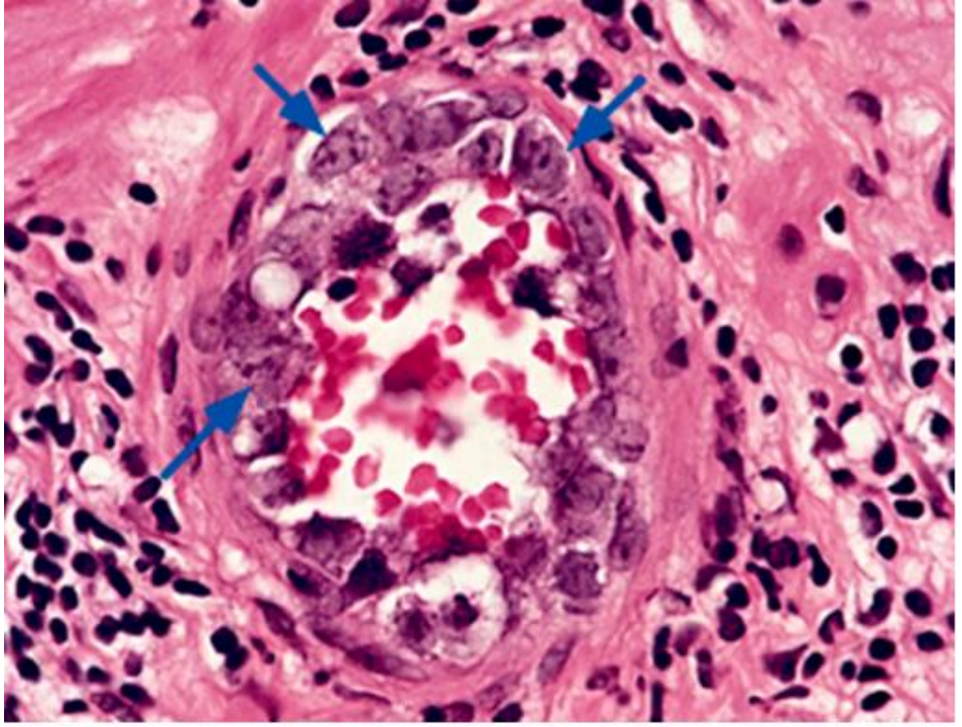




FDG PET

CT

1) Diffuse FDG uptake in the bilateral lung
2) Slightly heterogeneous uptake in the bone marrow with small focal accumulation sites
3) Elevated uptake in the cortex of the kidneys



The blood vessel shown in the center of this section is lined with lymphoma cells (arrows), with red blood cells in the lumen. .

IVL

- rare subtype of large B cell lymphoma.

- proliferation of lymphoma cells within the lumina of small blood vessels, particularly capillaries and post-capillary venules, without an obvious extravascular tumor mass or readily observable circulating lymphoma cells in the peripheral blood.

Freedman AS, UpToDate

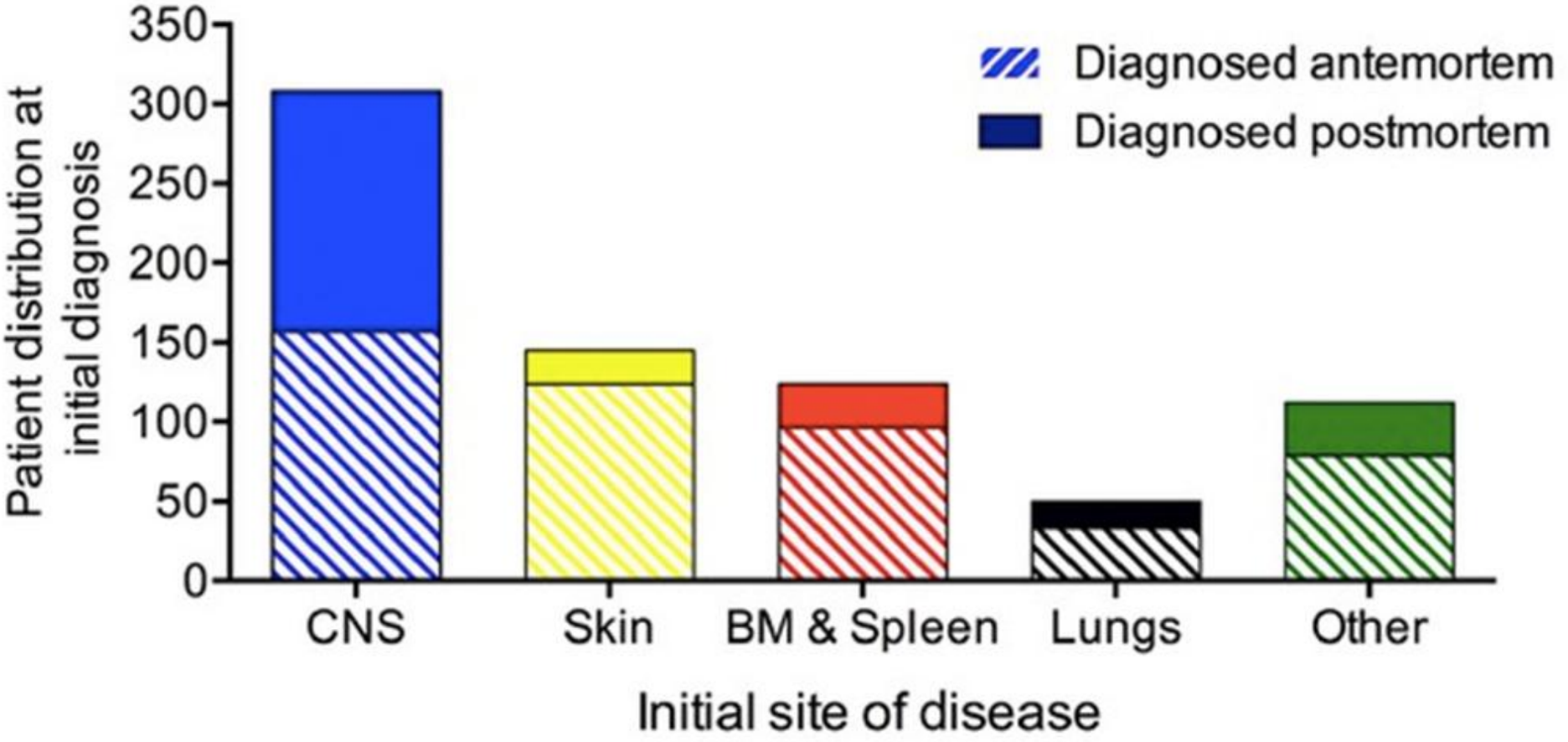
Purpuric lesions on arms of deceased 70-year-old patient with IVL

The diagnosis of IVL is made postmortem in over 50% of the cases.

IVL can affect virtually any organ in the body:

- CNS - 60%.
- Skin - 8%.
- Bone marrow and spleen - 11%.
- Lung - 7%.

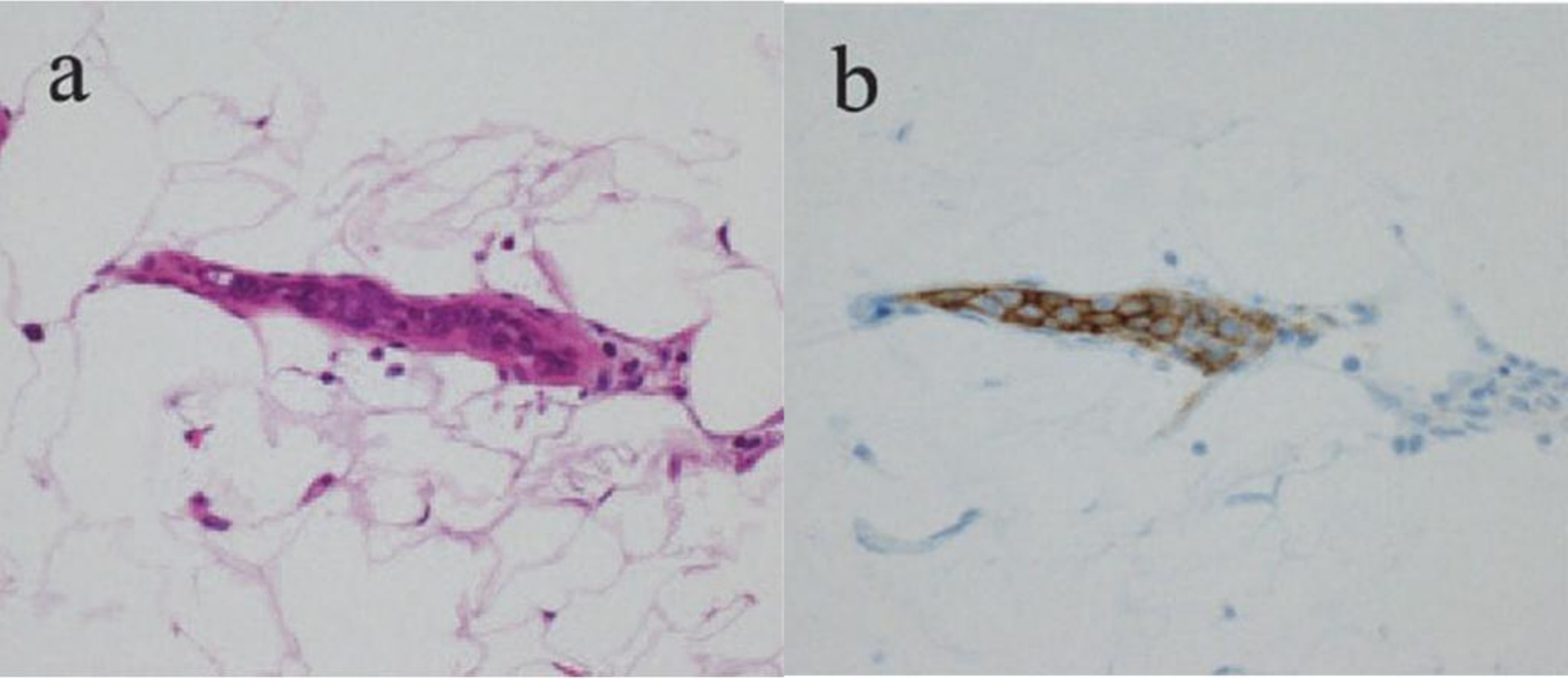
Orwat DE, 2012; Fonkem E, 2014



Patient distribution at initial diagnosis.

Among the newly diagnosed IVL patients, 308 (41%) presented in the CNS, 145 (20%) in skin, 124 (17%) in BM and spleen, 50 (7%) in lungs, 101 (13%) in other sites and 12 (2%) unknown. Among the 250 patients diagnosed at postmortem, IVL affecting the CNS was highest compared to other organ sites: 151 (60%) CNS, 21 (8%) skin, 28 (11%) BM and spleen, and 17 (7%) lung and 33 (13%) other organs.

Fonkem E, 2014



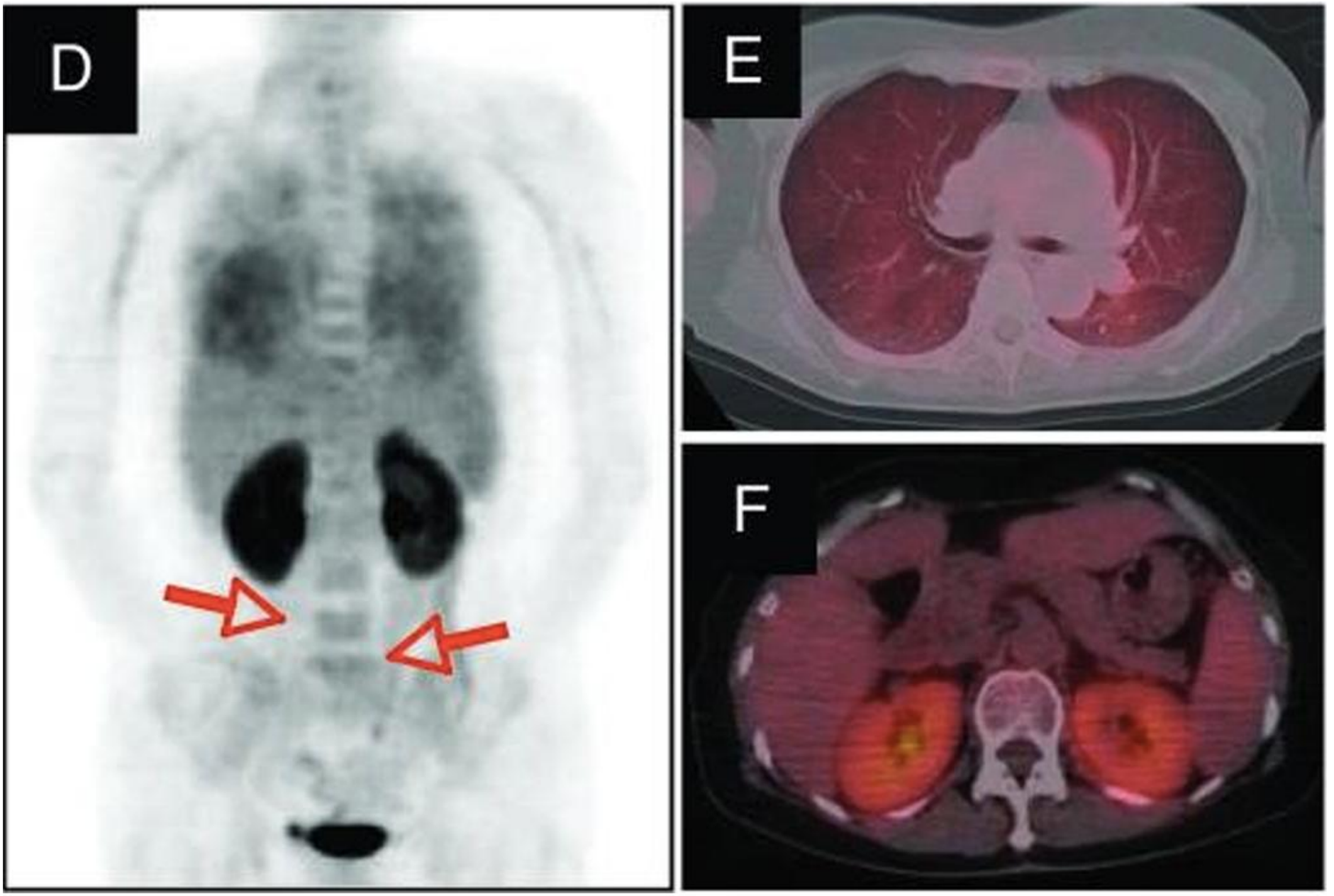
Histopathological analysis of skin biopsy from the left thigh.
(a) Large lymphoma cells in the small vessels (hematoxylin and eosin staning).
(b) Tumor cells were positive for CD20 (CD20 immunostain)

Shiiba M, 2014

FDG-PET findings
demonstrated diffuse FDG uptake in the lung (47.8%).

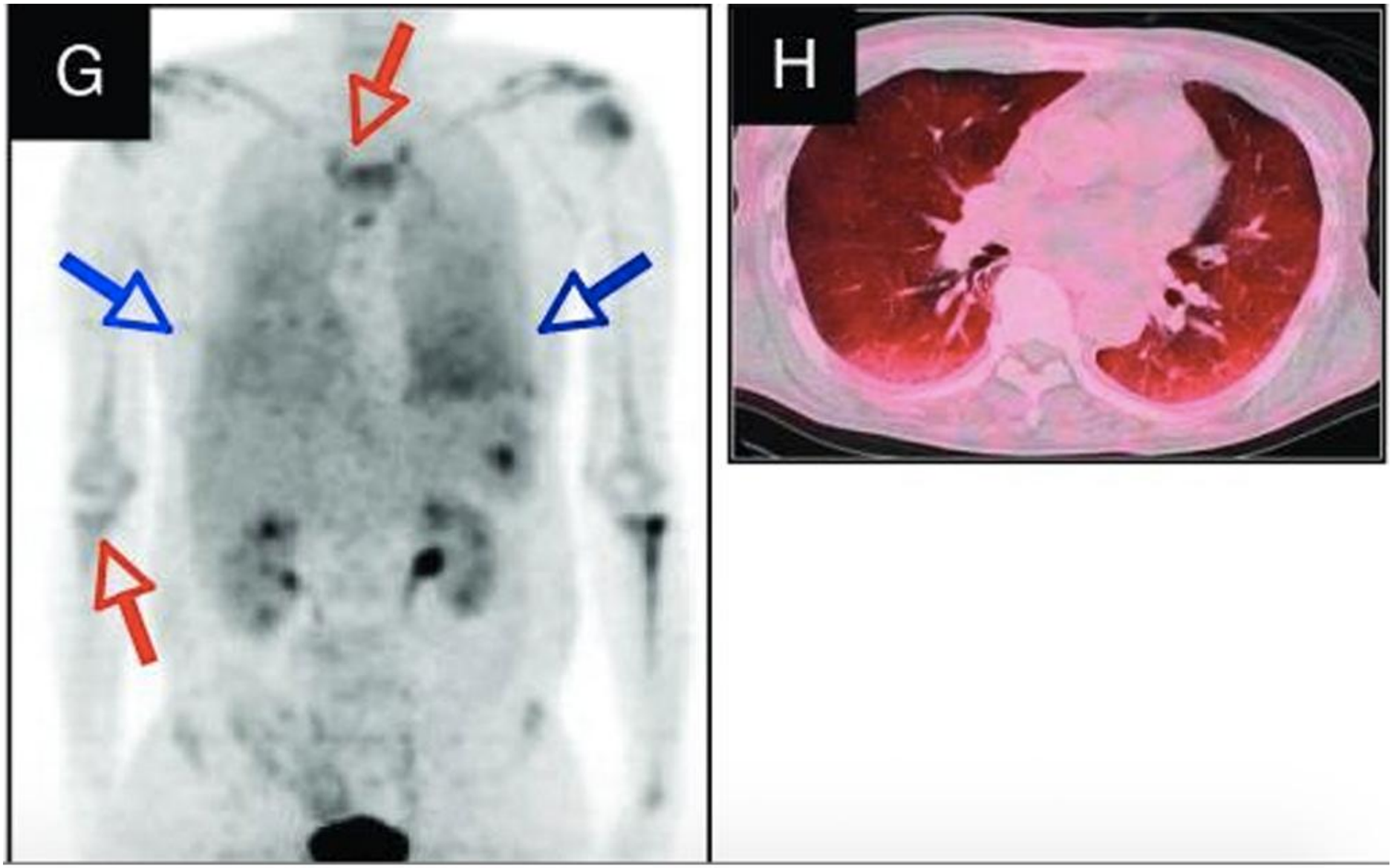
The other reported abnormal foci of FDG uptake were in the bone marrow (57.0%), spleen (30.0%), renal cortex (22.0%), uterus/vagina (17.4%), adrenals (13.0%), lymph nodes (8.7%) and stomach (4.0%).

Shiiba M, 2014



D-F, Uptake of FDG with diffuse and intense accumulation in the bilateral lung of a 69-year-old woman (D and E), accumulation in the bilateral renal cortex (F), and multiple accumulations in the bones (D [red arrows]).

Miura Y, 2010



G-H, Uptake of FDG with diffuse accumulation in the bilateral lung of a 64-year-old man, especially in the lower lung field (G [blue arrows] and H), and multiple accumulations in the bones (G, red arrows). FDG uptake was not observed in the bilateral renal cortex (G).

Miura Y, 2010