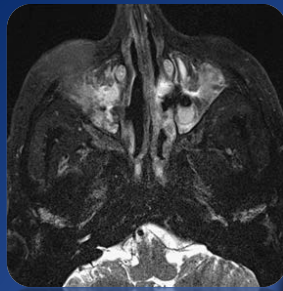


Erdheim-Chester Disease

A clonal histiocytic neoplasm
affecting multiple organ systems

ELAINE S. JAFFE, MD
National Cancer Institute

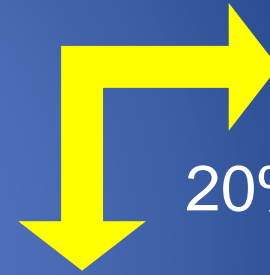
28%



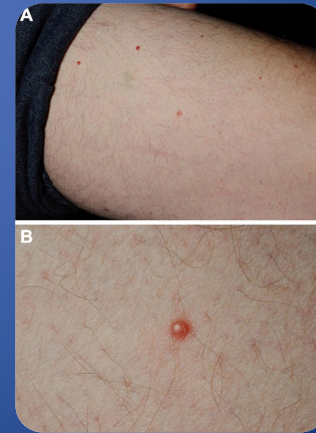
30%



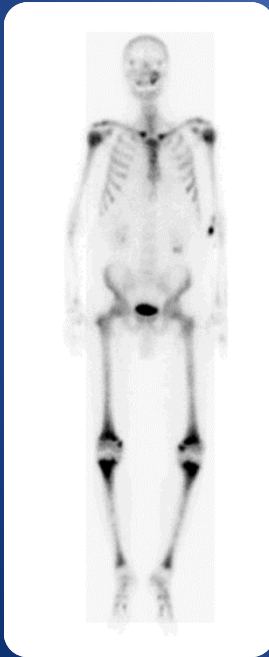
44%



20%

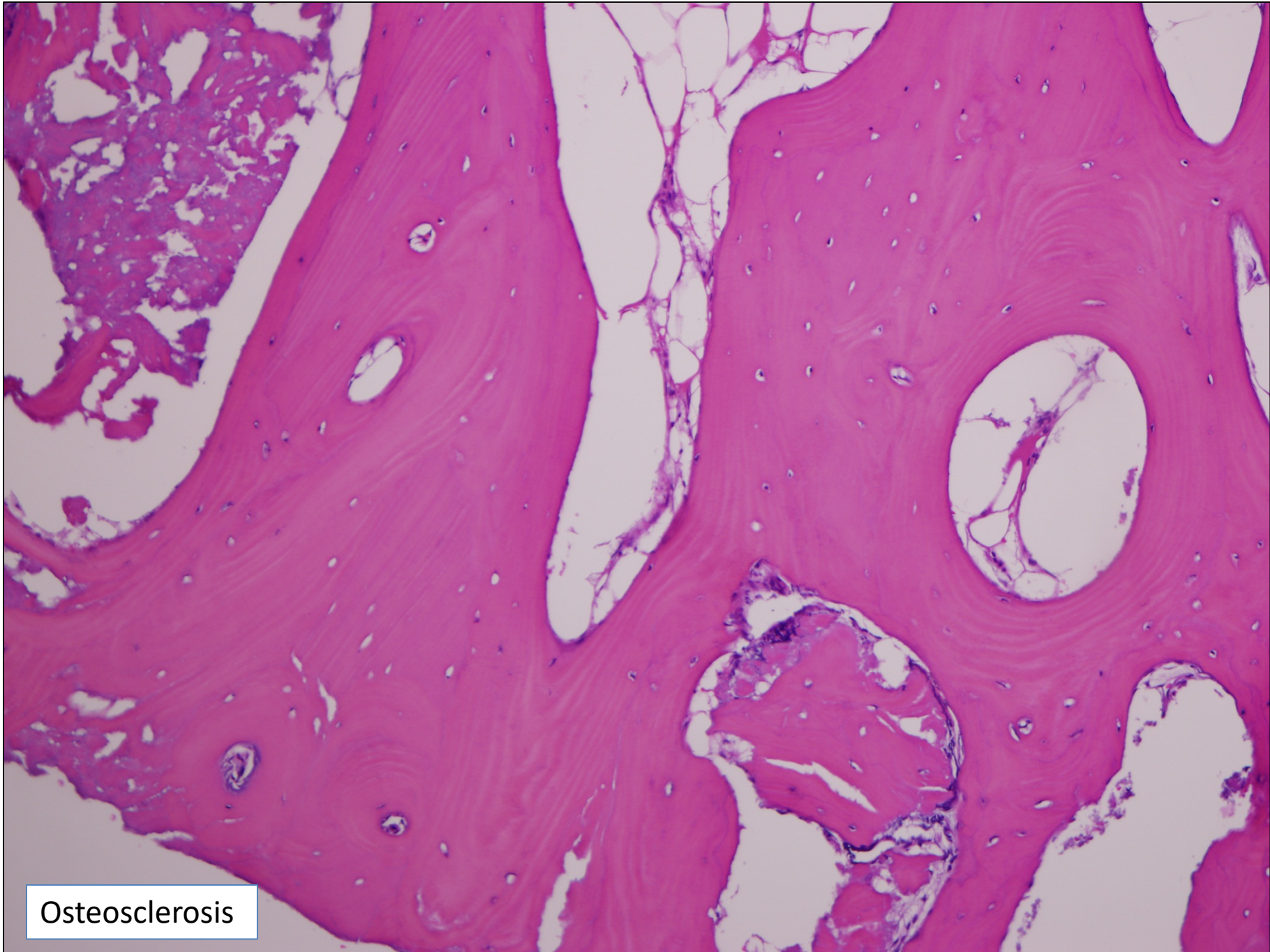


94%

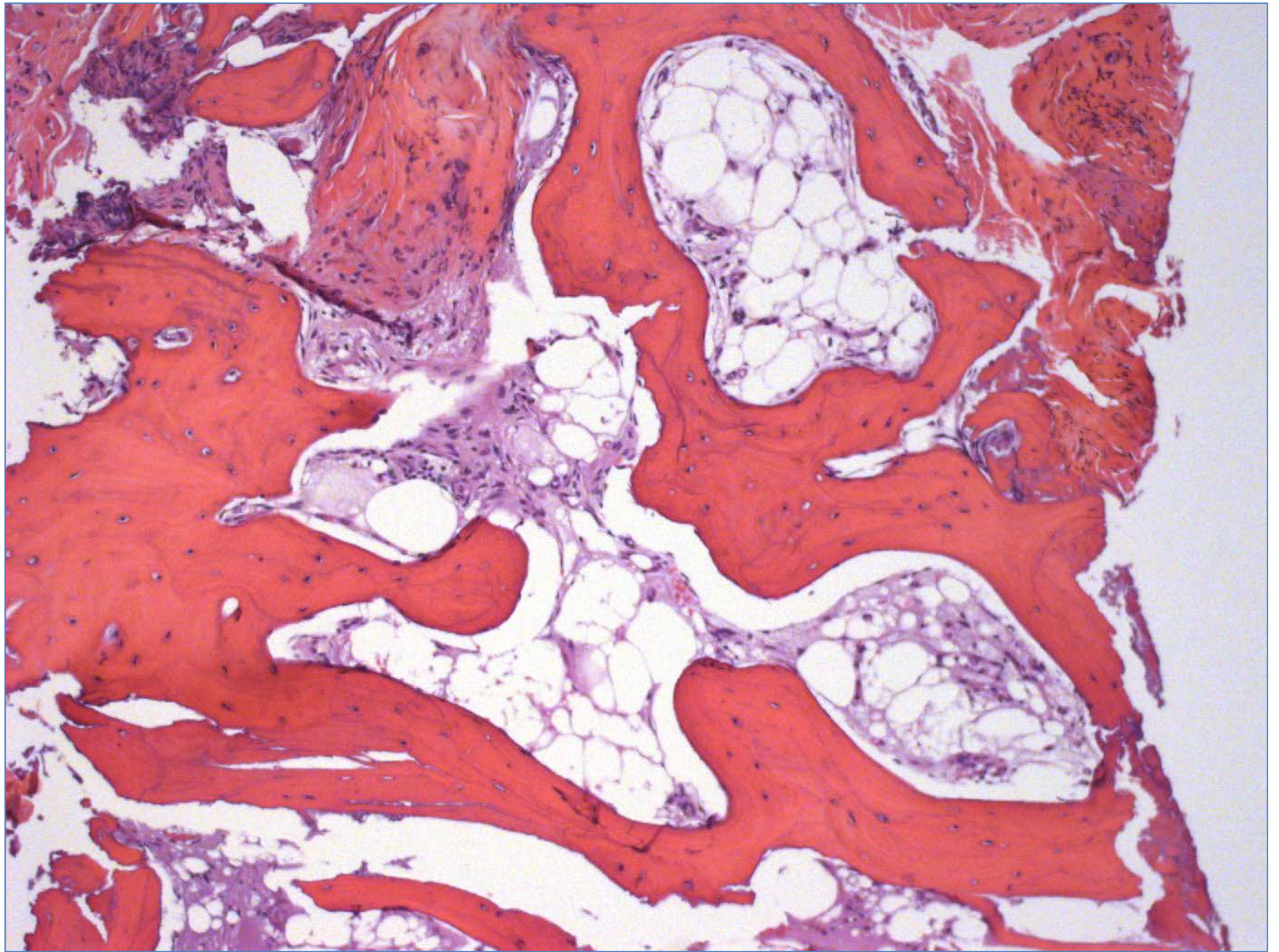


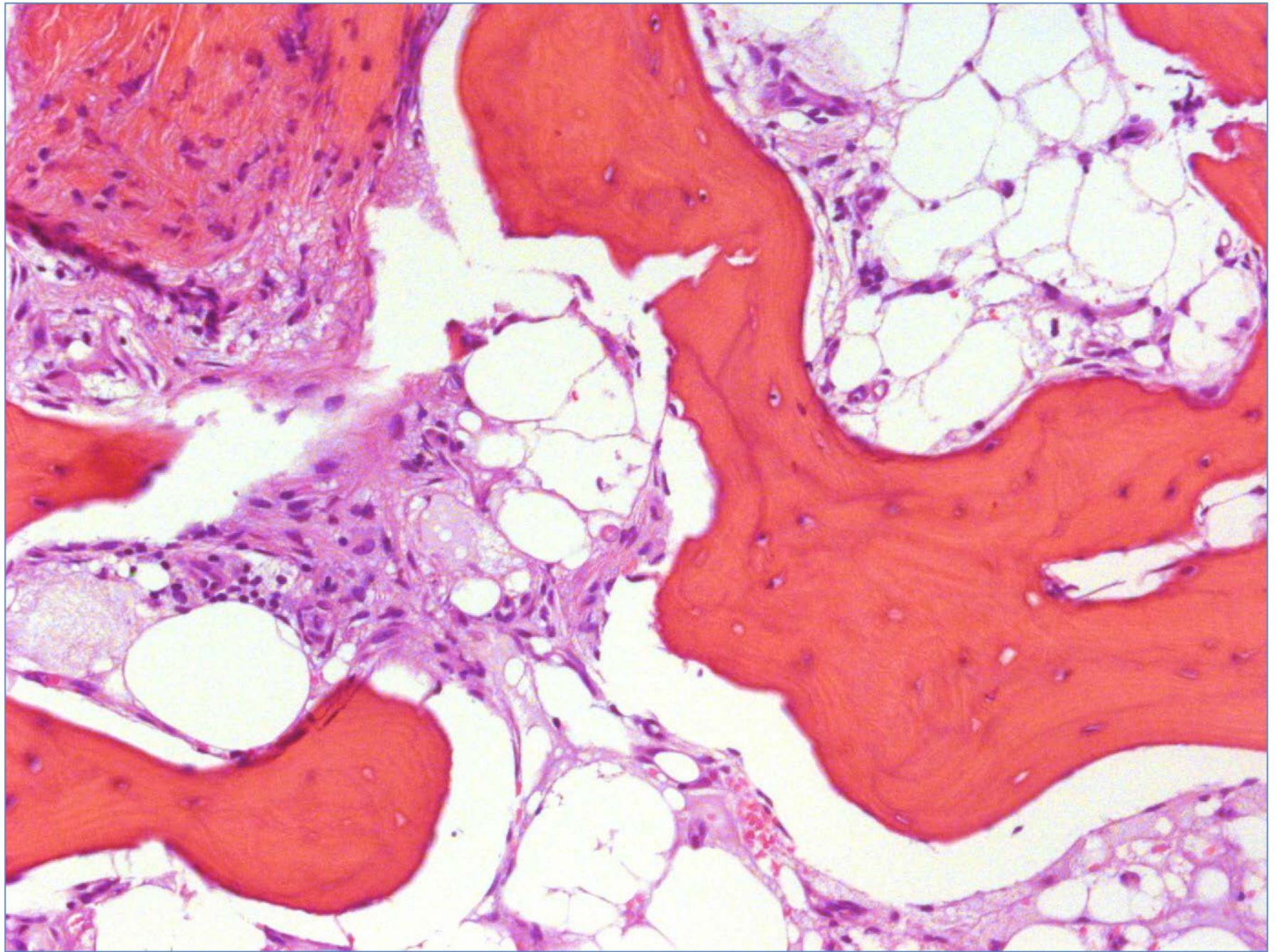
75 year old male
Bone pain, fatigue
Increased uptake in femur, tibia
CT of abdomen shows perirenal
stranding

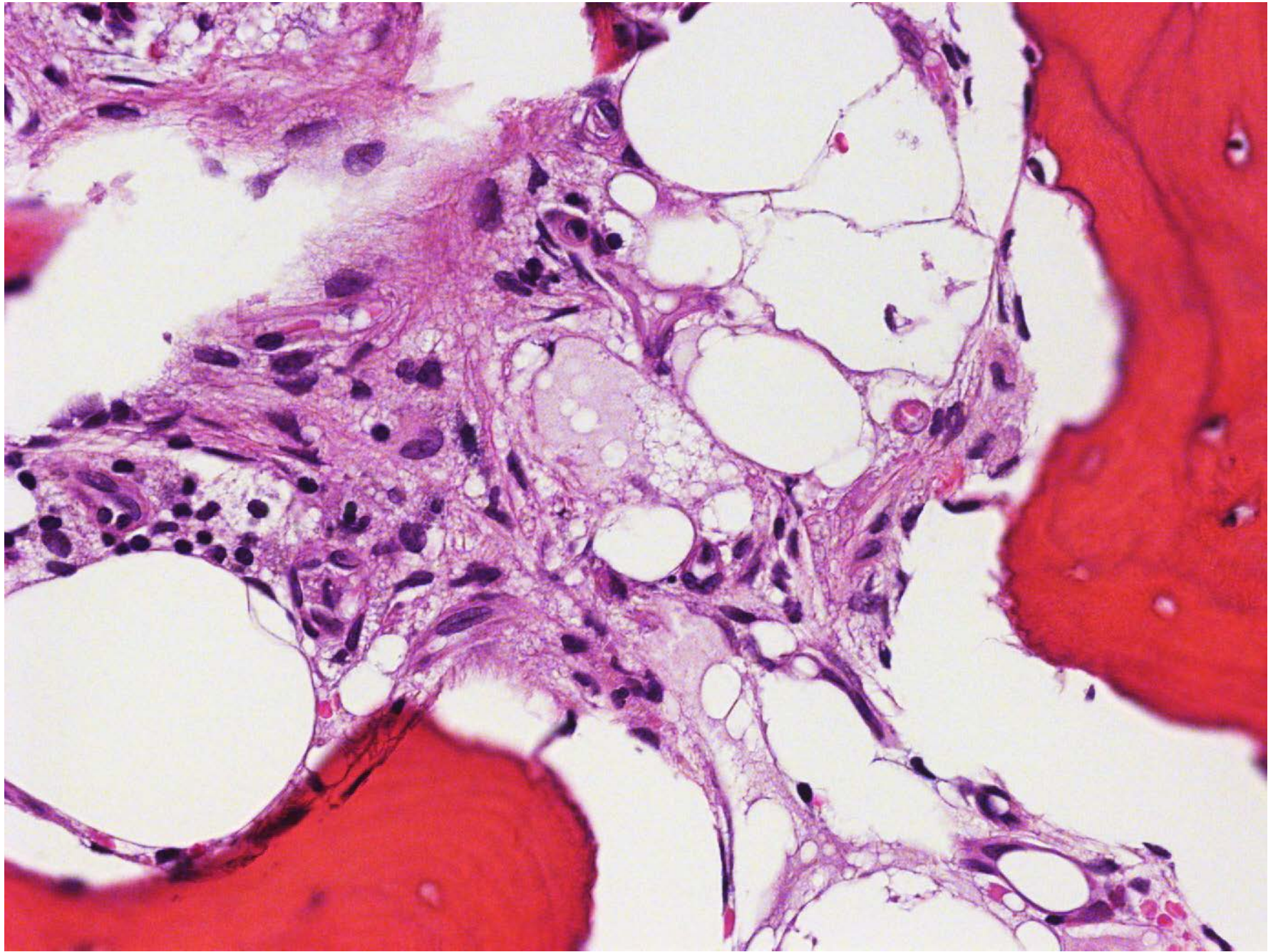
Biopsy, Left Distal Femur



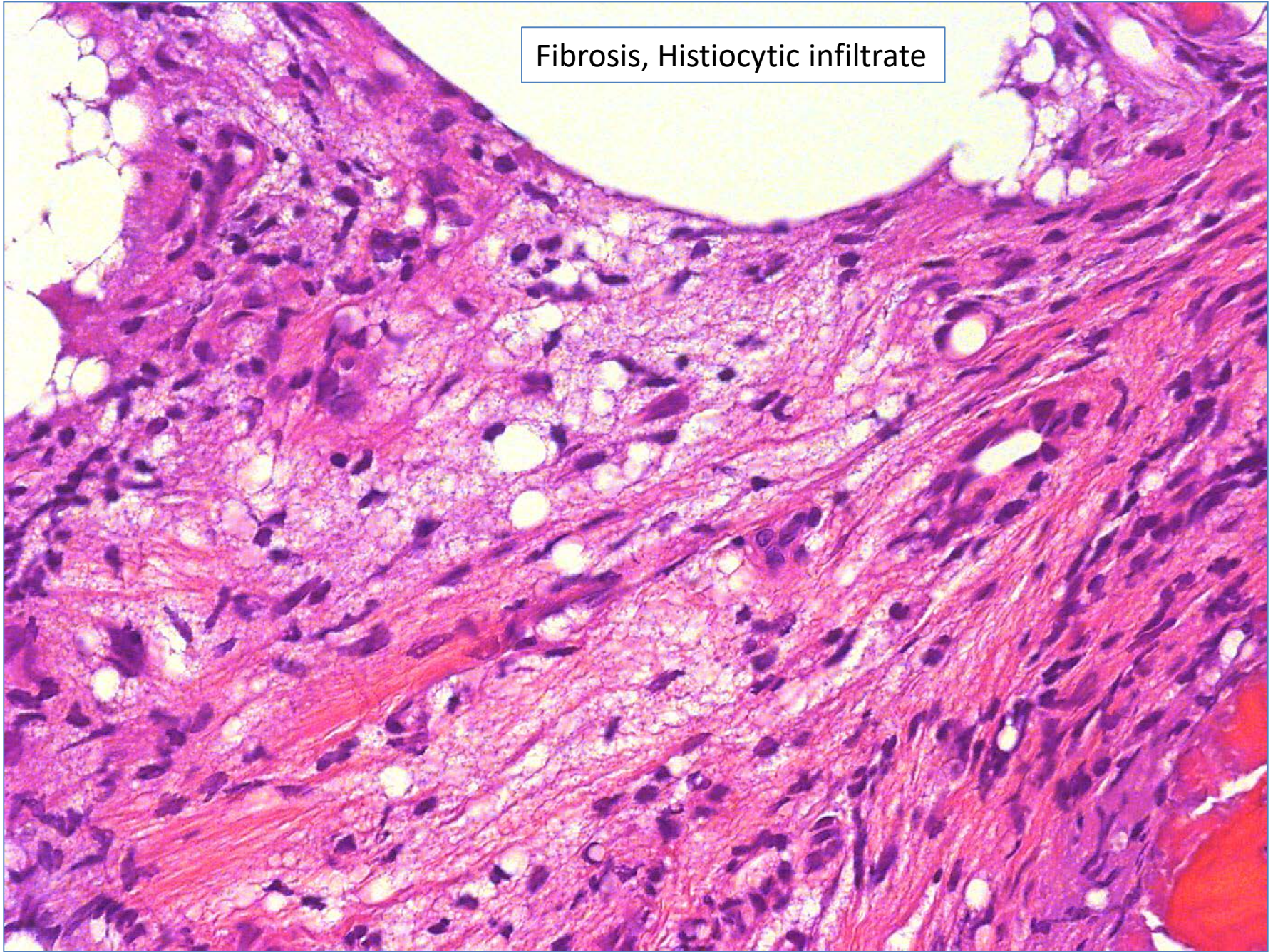
Osteosclerosis

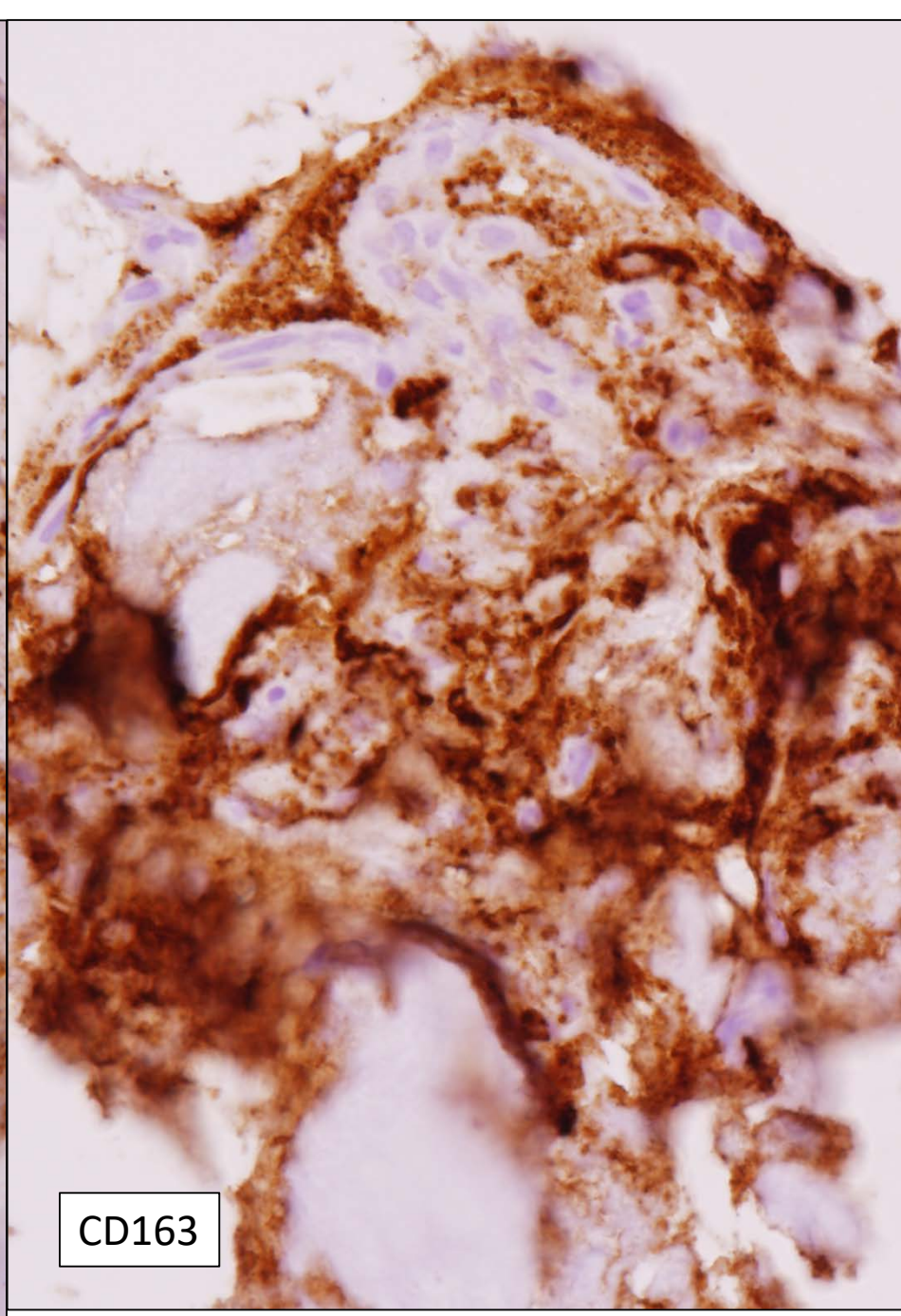
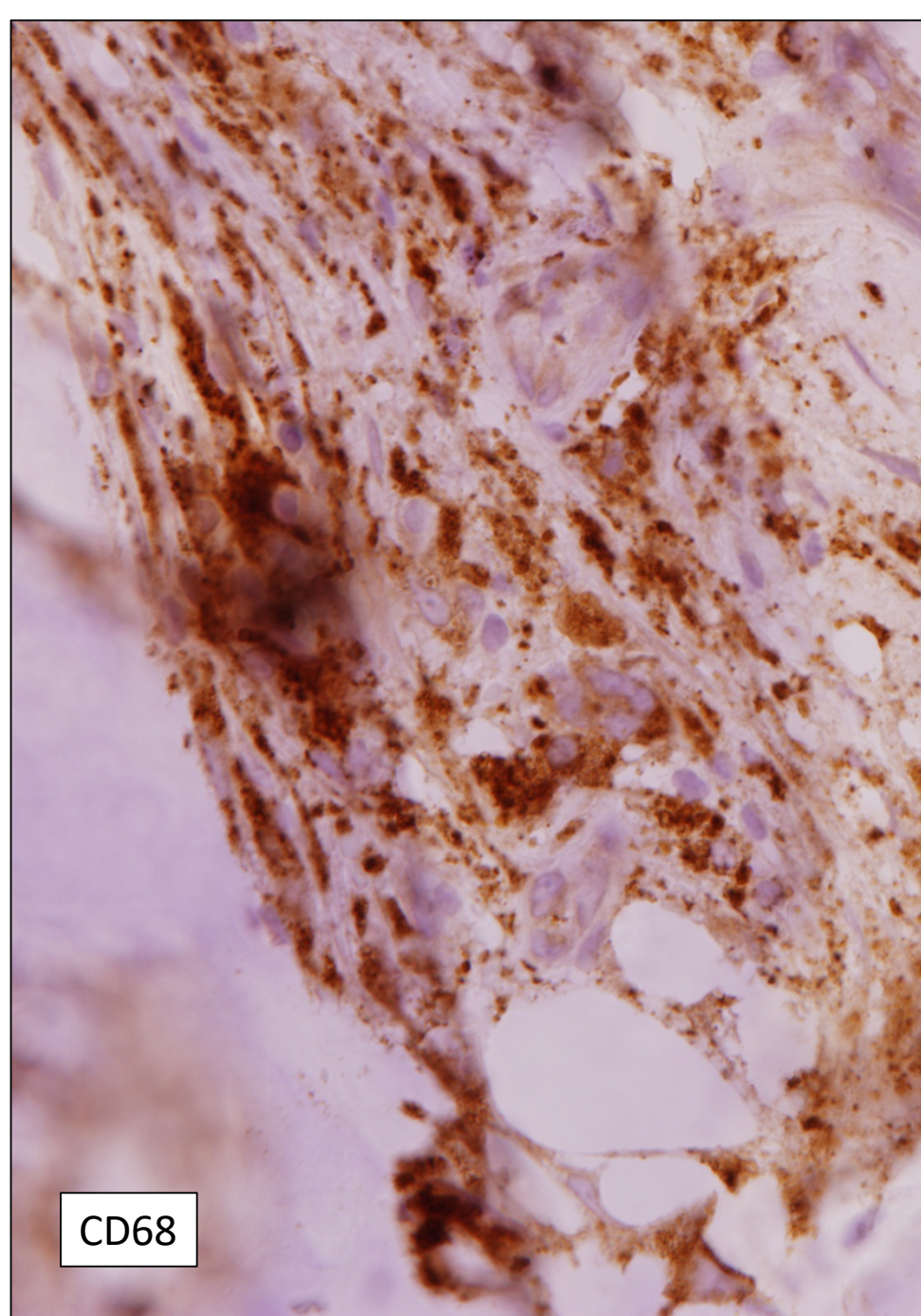




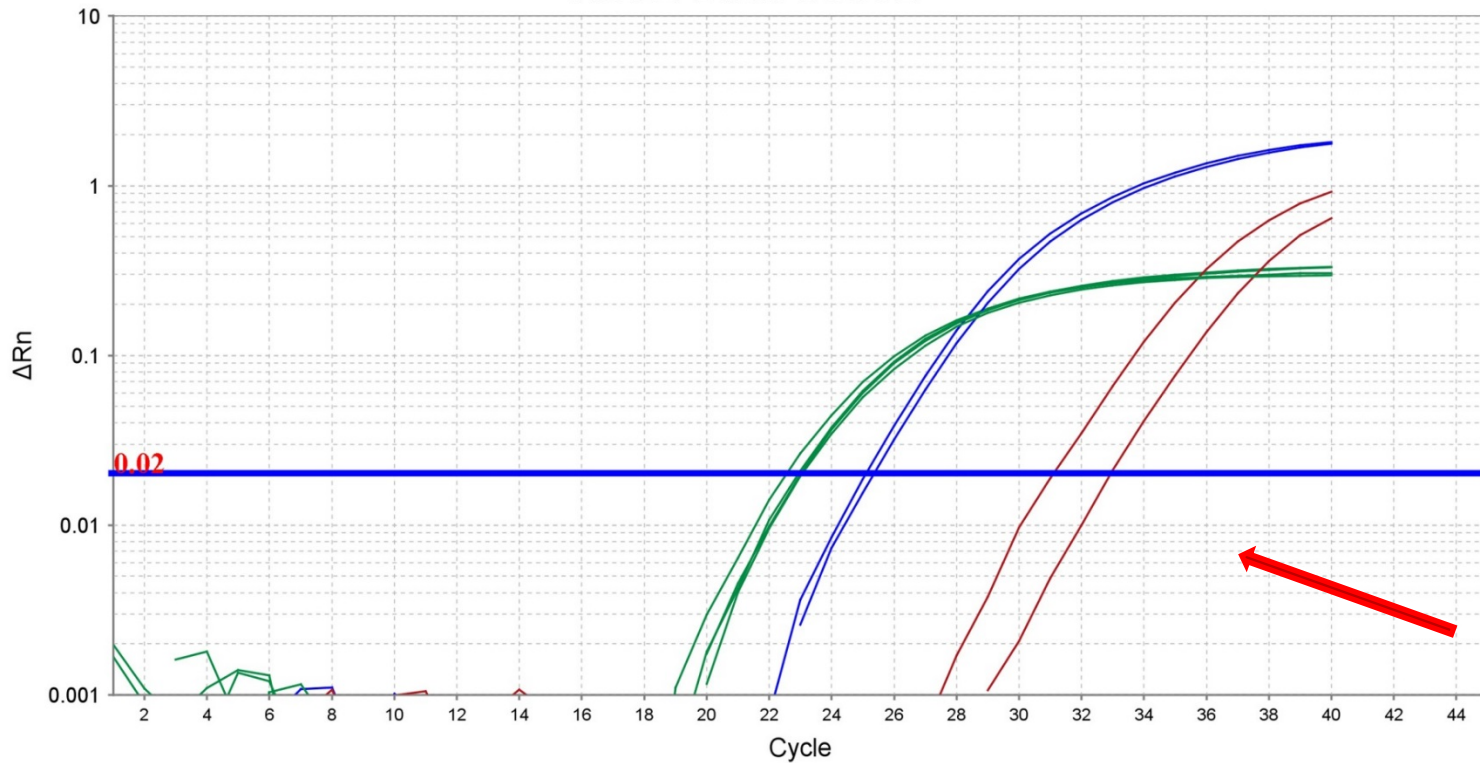


Fibrosis, Histiocytic infiltrate





M572-BRAFV600E



Legend

IPC BRAF_476_mu BRAF_rf

Positive for BRAF V600E mutation

Bone biopsies must be processed in EDTA – not formic acid for decalcification

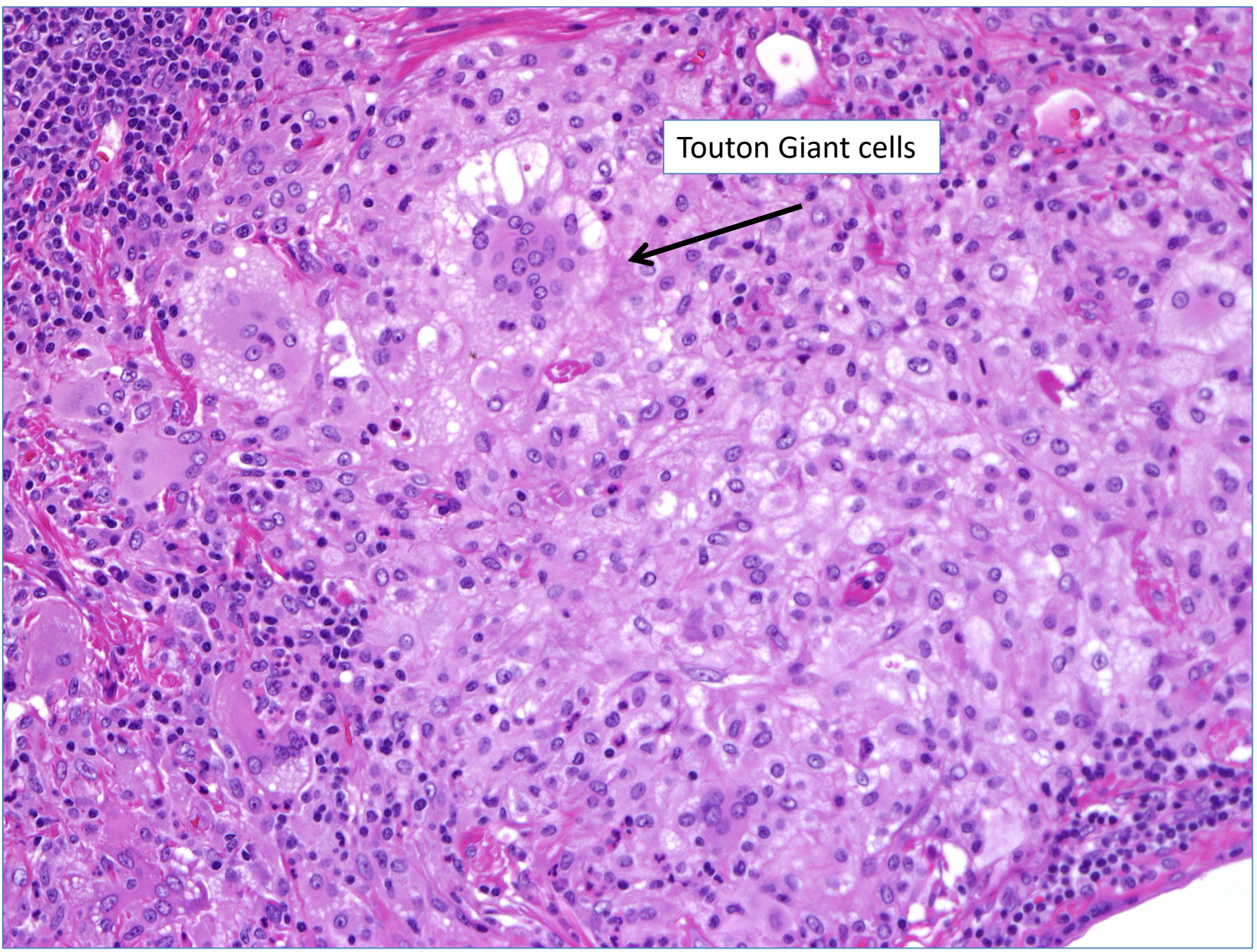
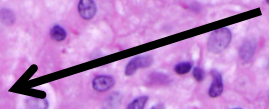
53 year old female
Progressive headache,
loss of balance

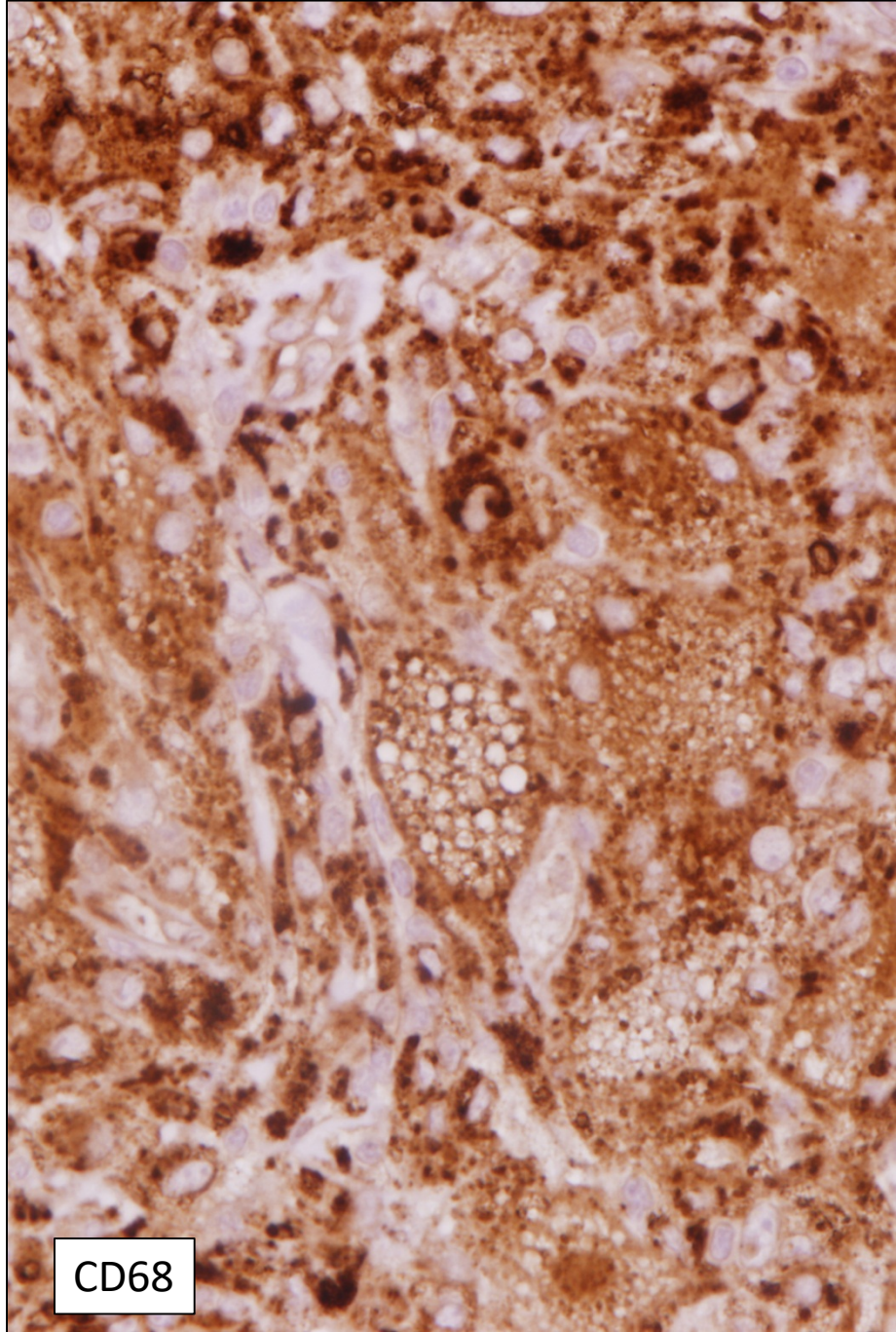
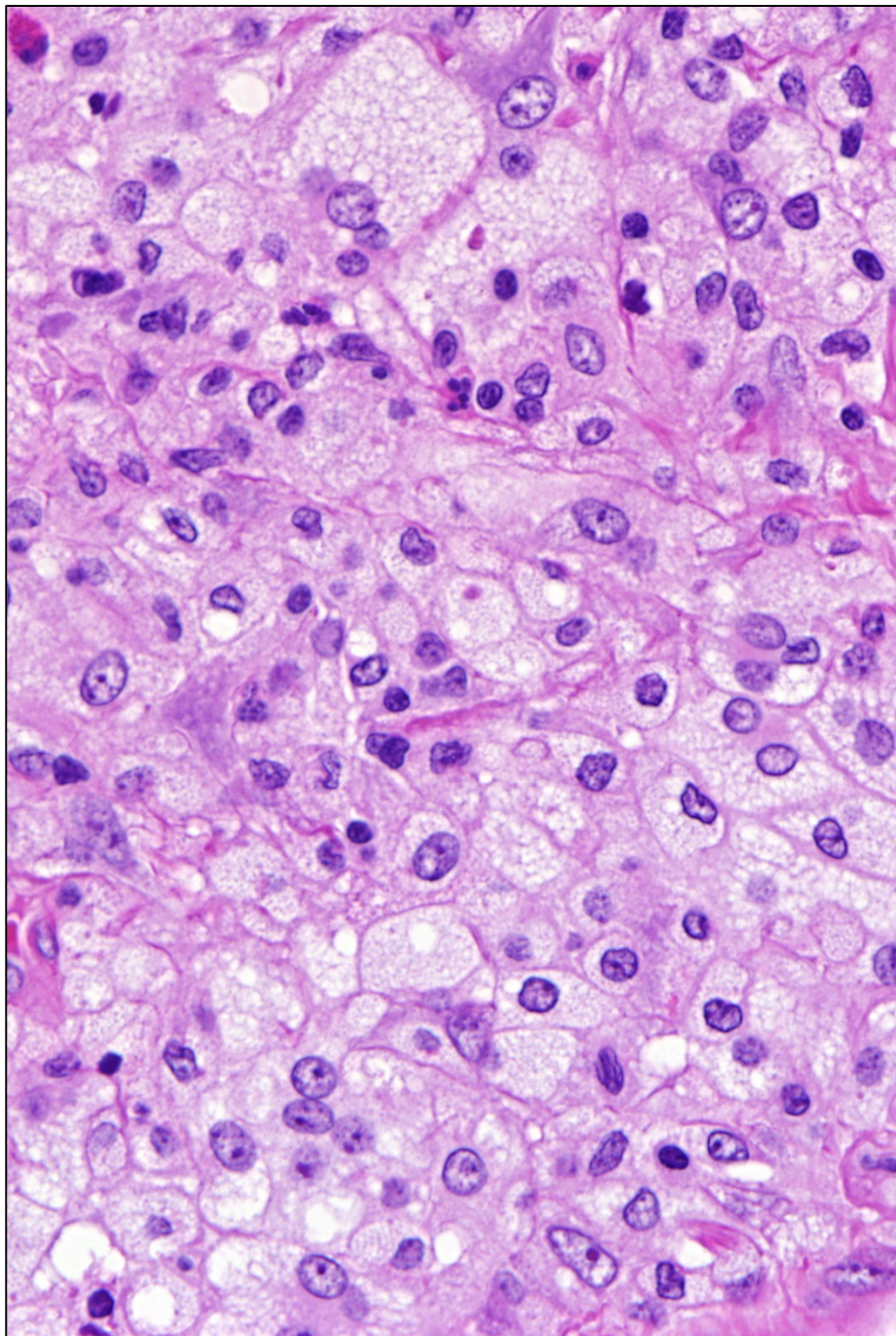
MRI showed cerebellar
lesion and multiple
lesions surrounding
fourth ventricle, pons,
left occipital lobe, right
parietal lobe

Biopsy of cerebellum



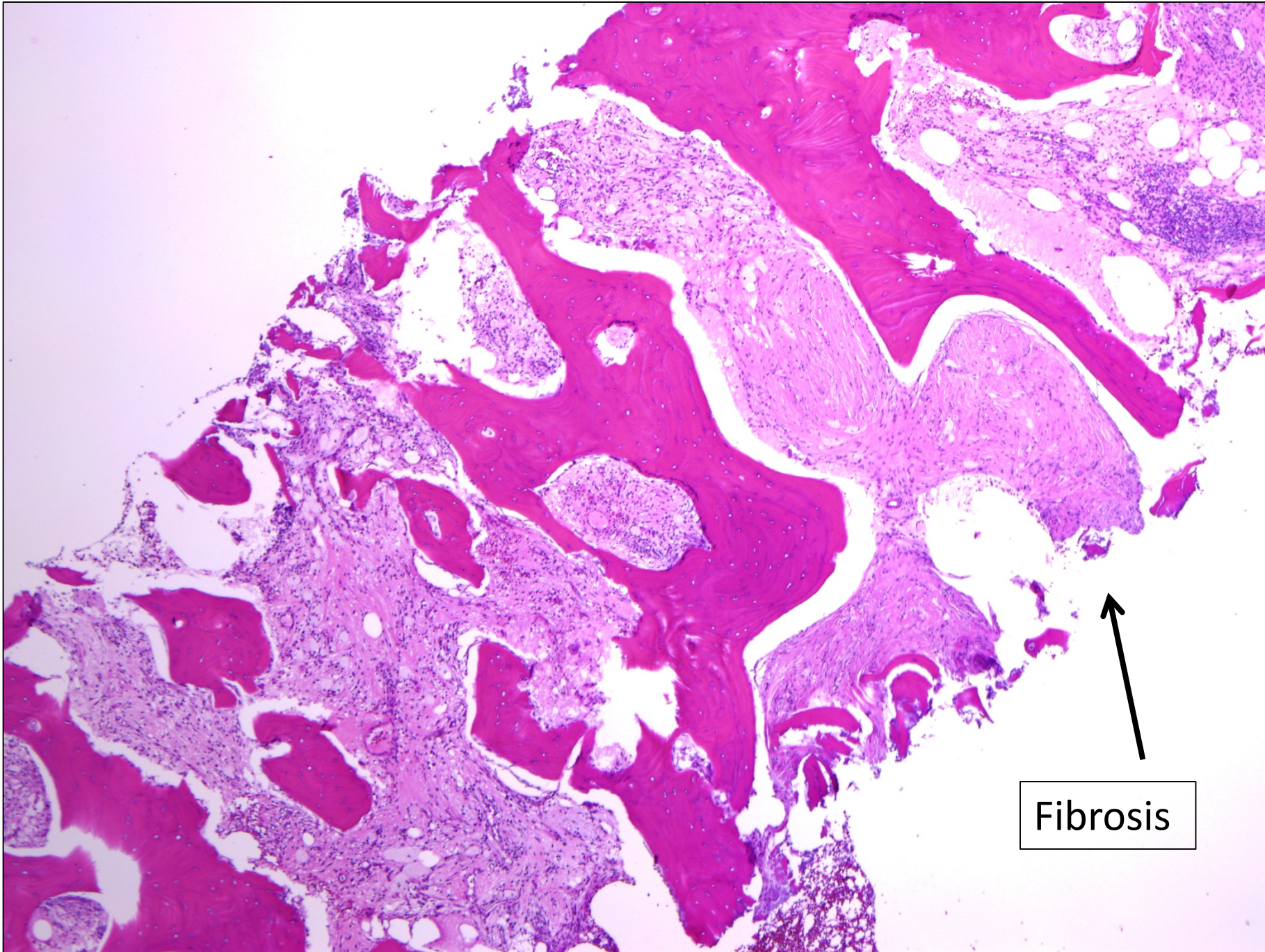
Touton Giant cells





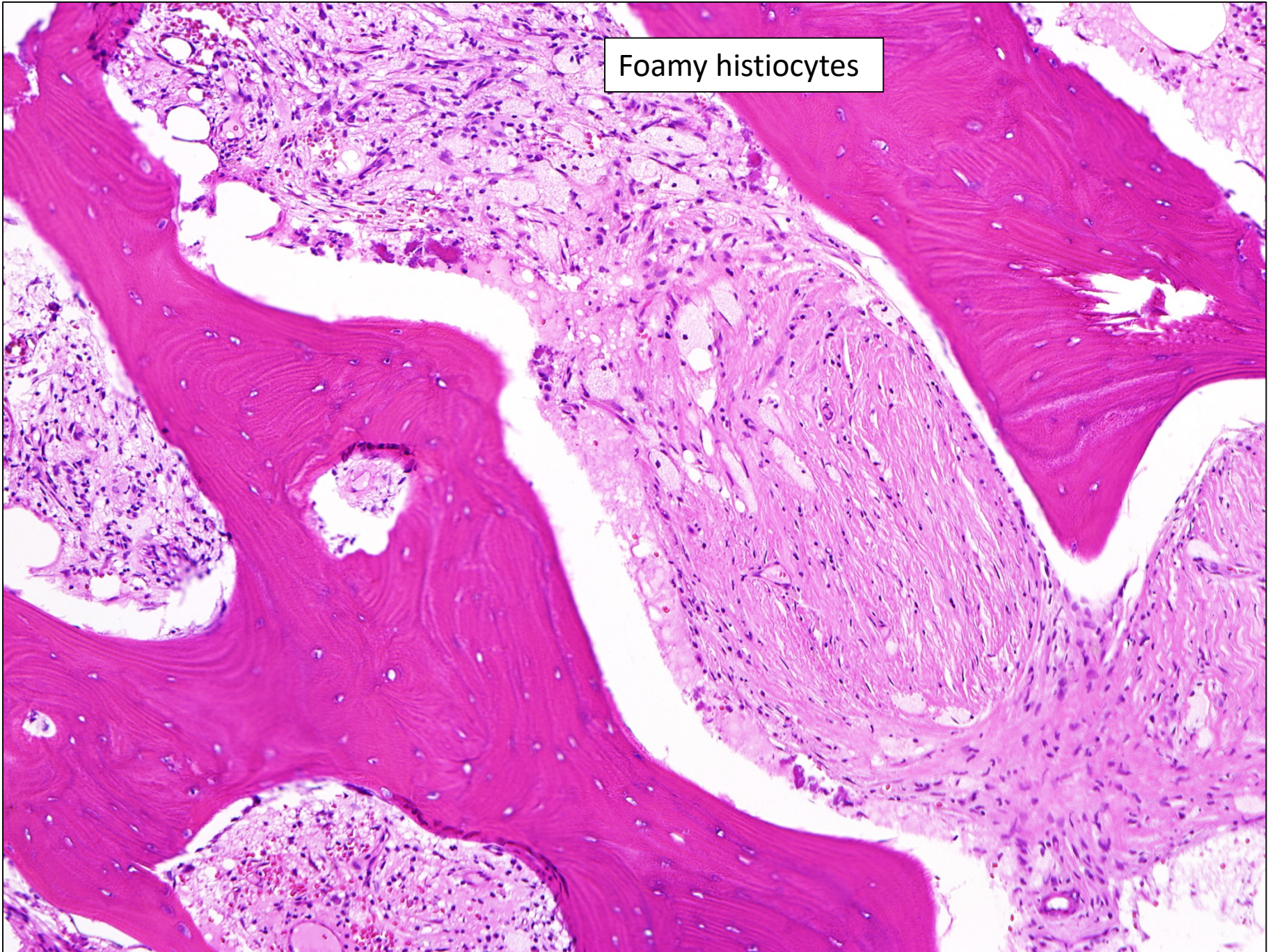
CD68

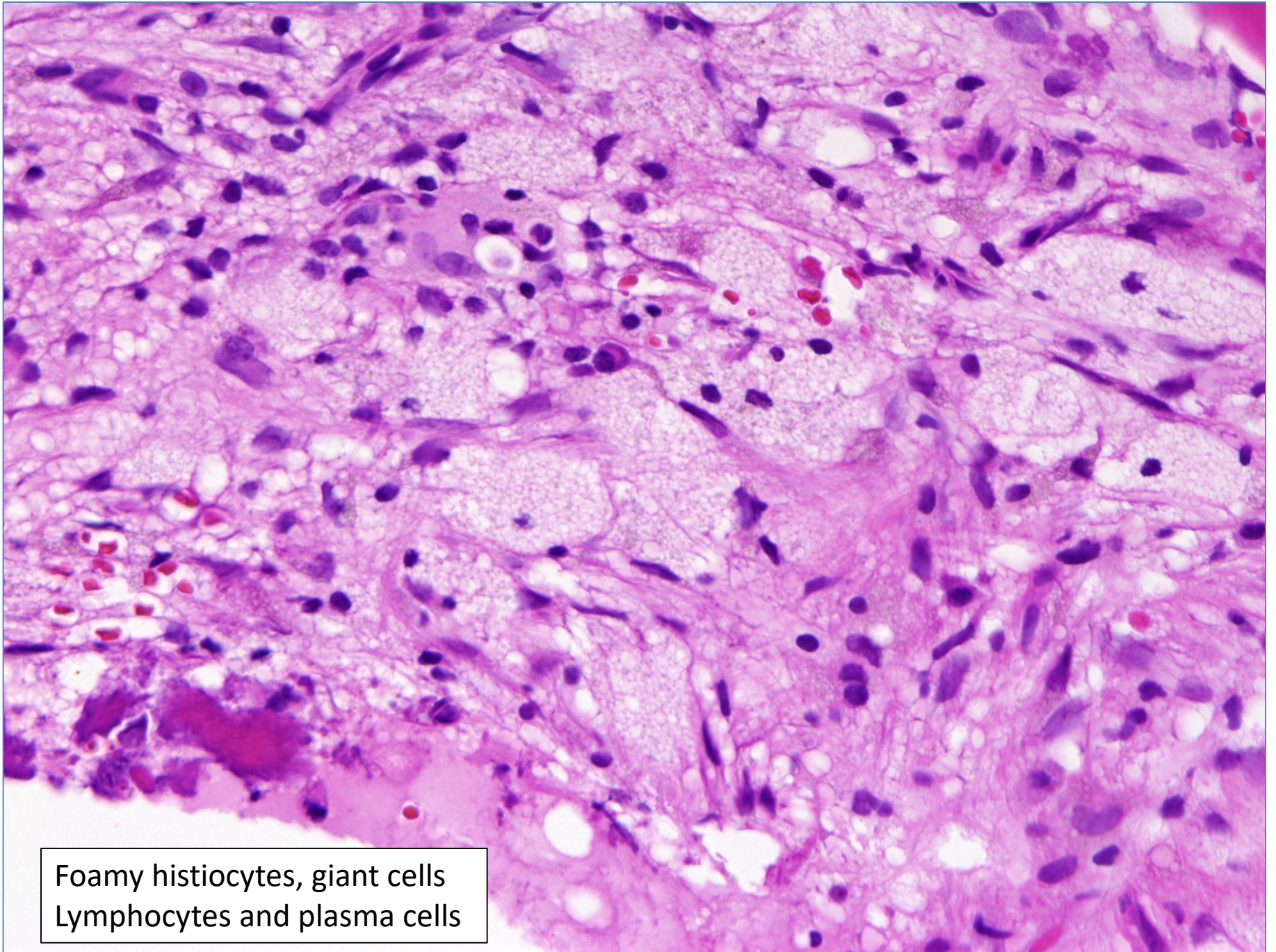
45 year old male with a sclerotic
lesion of the femur



Fibrosis

Foamy histiocytes





Foamy histiocytes, giant cells
Lymphocytes and plasma cells

CD68

CD163

