

All that glitters, is not gold

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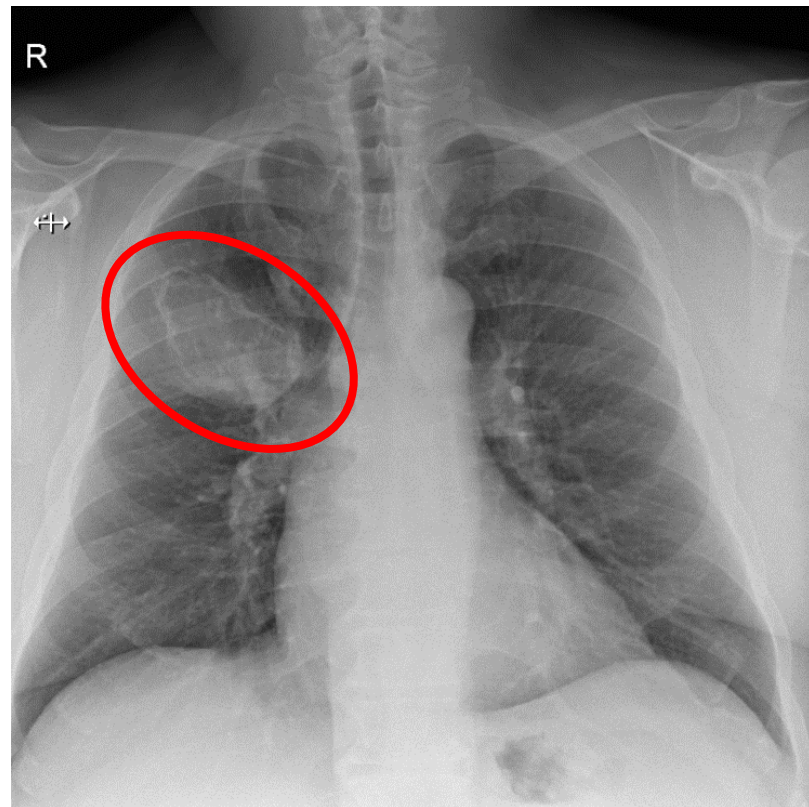


Case presentation

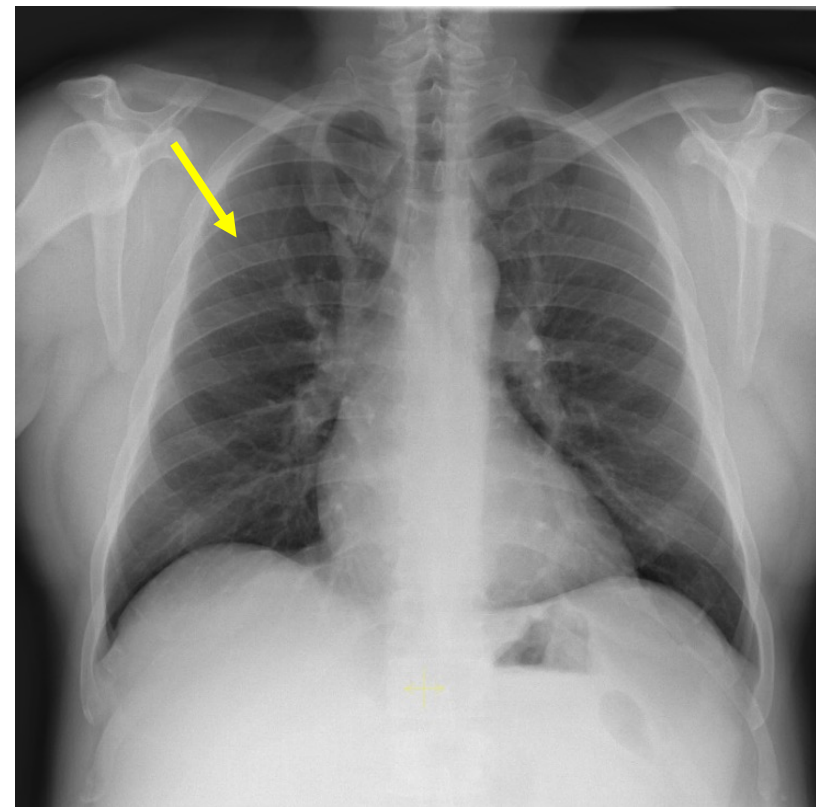
- 53Y - male
- Symptoms
 - Fatigue and dyspnea on exertion (3m)
 - Coughing with green mucus
- Clinical examination
 - Normal cardiac and pulmonary function
 - Pulse rate 100/min
- Biochemical examination
 - Increased leucocyte count ($9.66 \times 10^9/L$)
 - Limited elevation of transaminases and LDH
- Normal spirometry



Radiography of the thorax



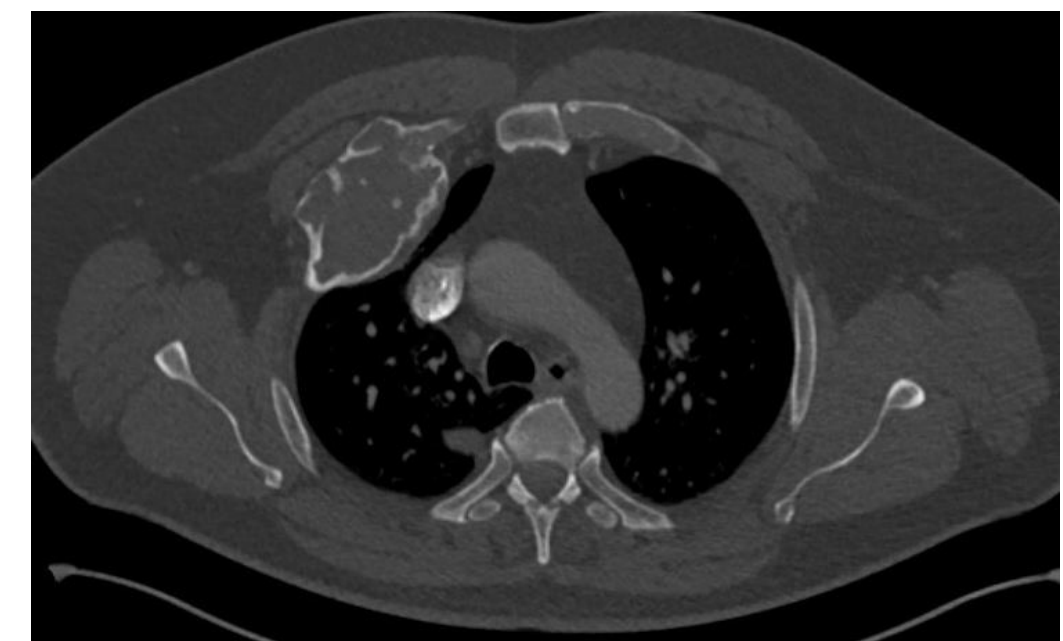
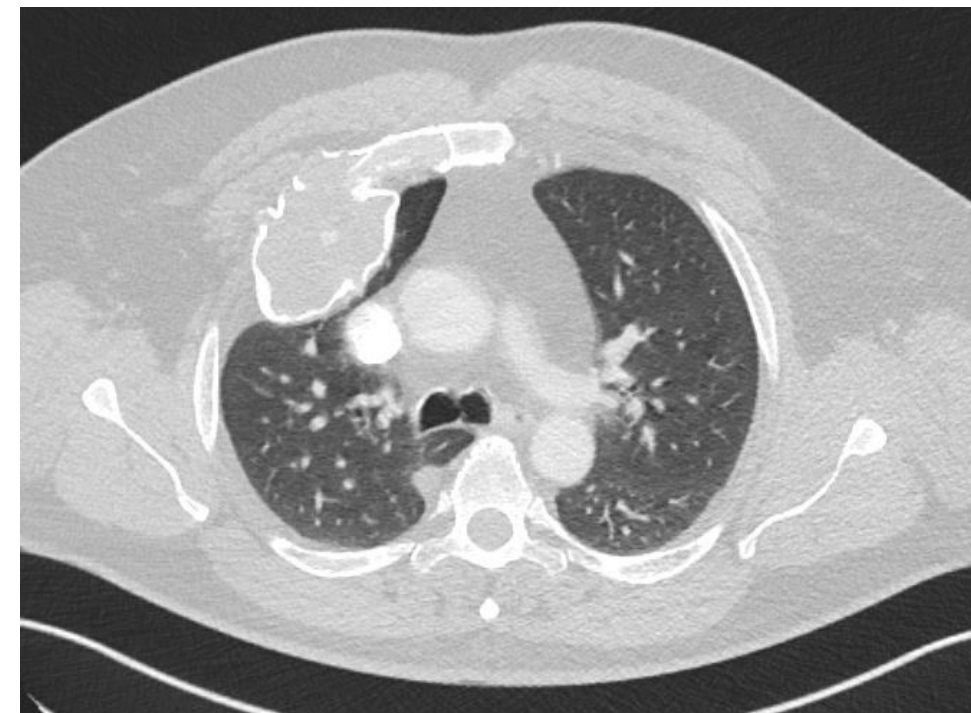
2022



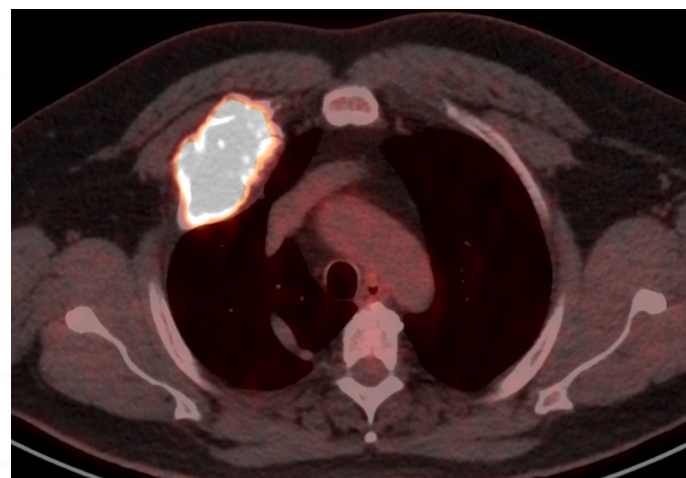
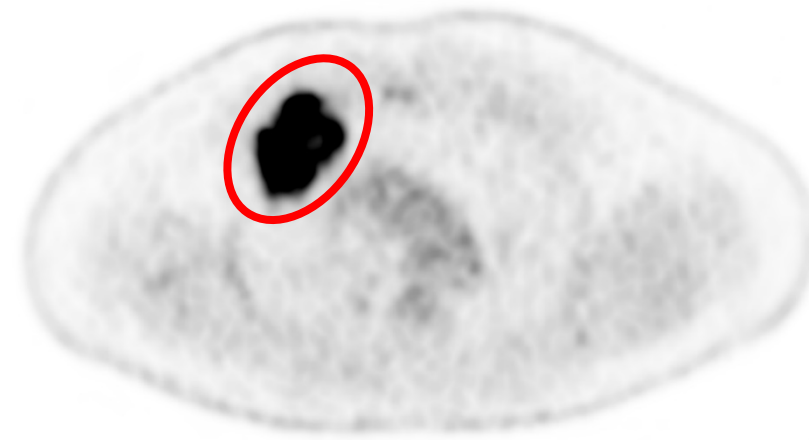
2008



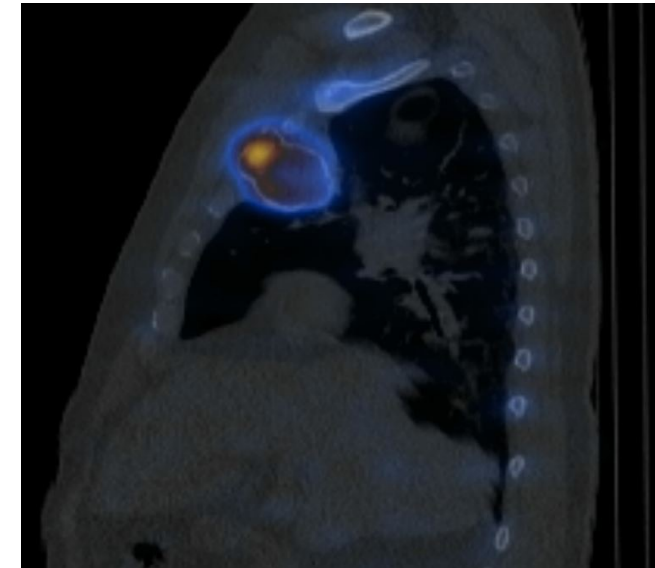
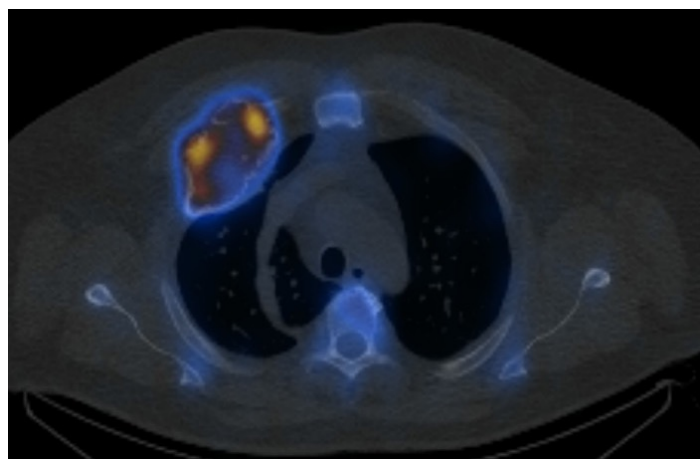
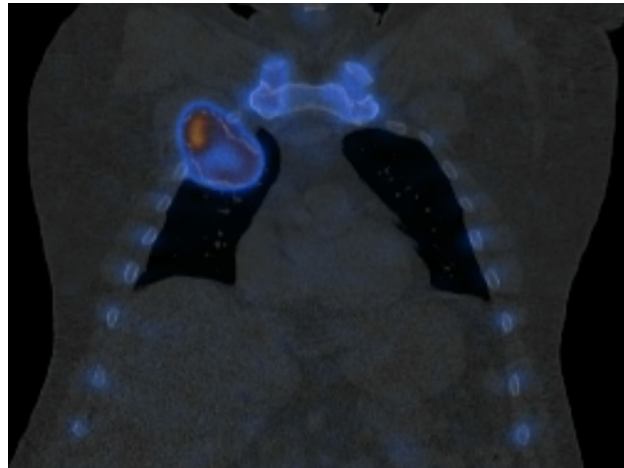
CT-scan of the chest with contrast



[¹⁸F]FDG PET/CT



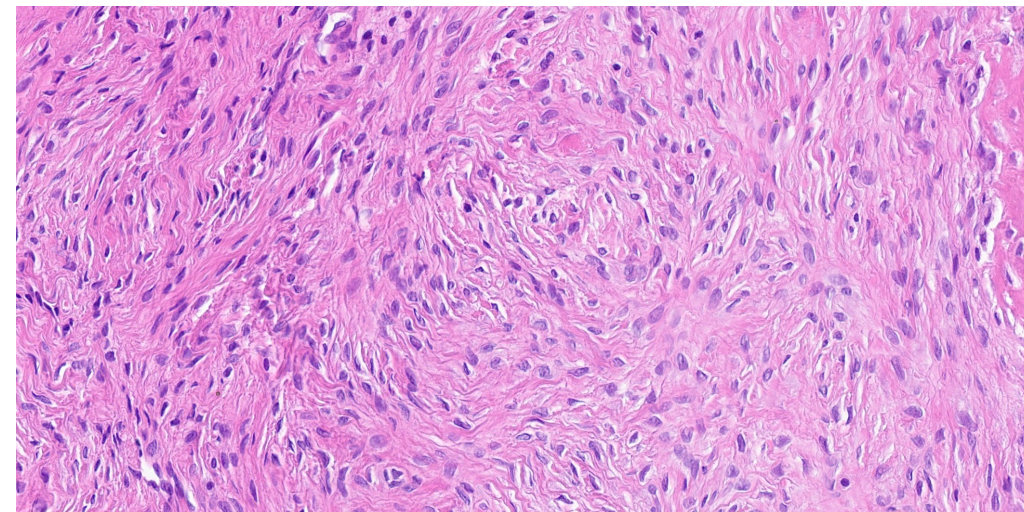
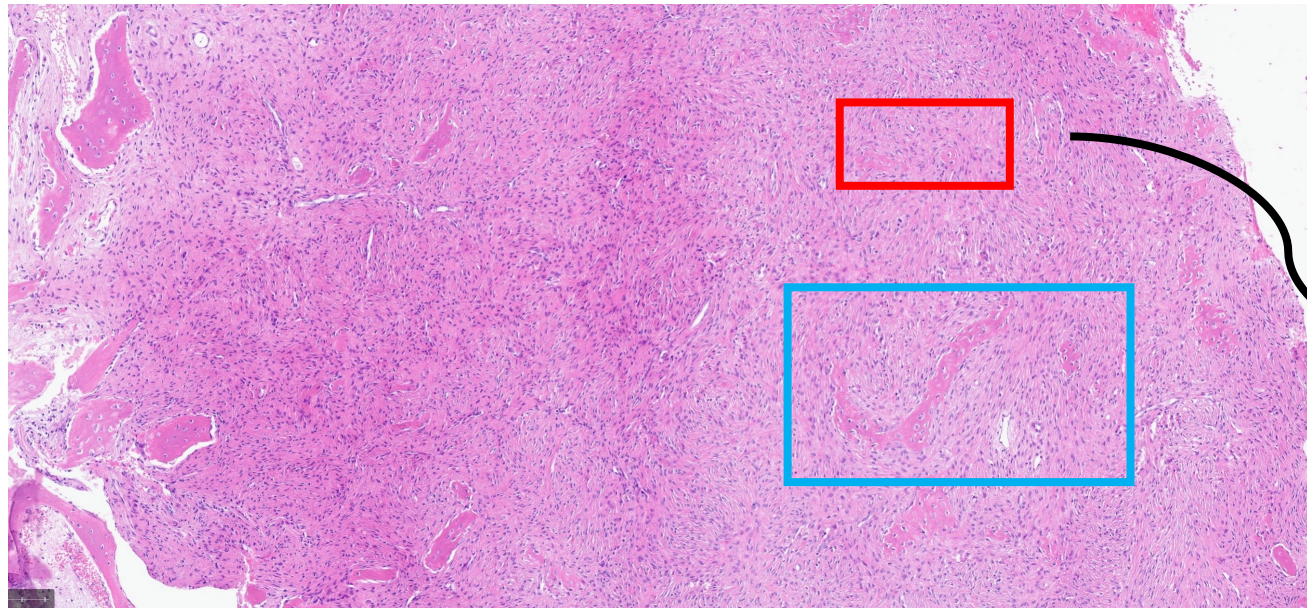
Bone scintigraphy



Biopsy



- Myofibroblasts
- Irregularly C- and S-shaped bone trabeculae
- Osteocytes without cytologic atypia (Ki-67 < 2%)



Fibrous dysplasia (1)

- **What?**

- Abnormal proliferation of bone fibroblasts and the formation of woven bone

- **When?**

- Prevalance 1/100,000 – 1/1,000,000
- 7% of benign bone tumors

- **Why?**

- Genetic, non-hereditary disorder, due to a somatic mutation in the GNAS gene (alpha subunit of the Gs protein)
 - cAMP ↑ → proliferation of skeletal progenitor cells

- **Where?**

- Single skeletal location (monostotic FD)
- Multiple skeletal locations (polyostotic FD)
- McCune-Albright Syndrome
 - Café au-lait skin hyperpigmentation
 - Precocious puberty
 - Endocrinopathies



Fibrous dysplasia (2)

- Complications
 - Neurological and ophtalmological manifestations (craniofacial skeletal dysplasia)
 - Vision loss, hearing loss, headache, migraine and orofacial pain
 - Bone pain
 - Fractures
 - Vertebral deformities
 - Scoliosis: pulmonary compromise and death
- Malignant transformation
 - 0,4% up to 4% in McCune-Albright Syndrome
 - Osteosarcoma (55%)
 - Fibrosarcoma (30%)
 - Chondrosarcoma (15%)
- Therapy
 - Surgery
 - Biphosphonates
 - Denosumab (RANK-ligand)



Baseline

6 months

Tuesday, April 4, 2023

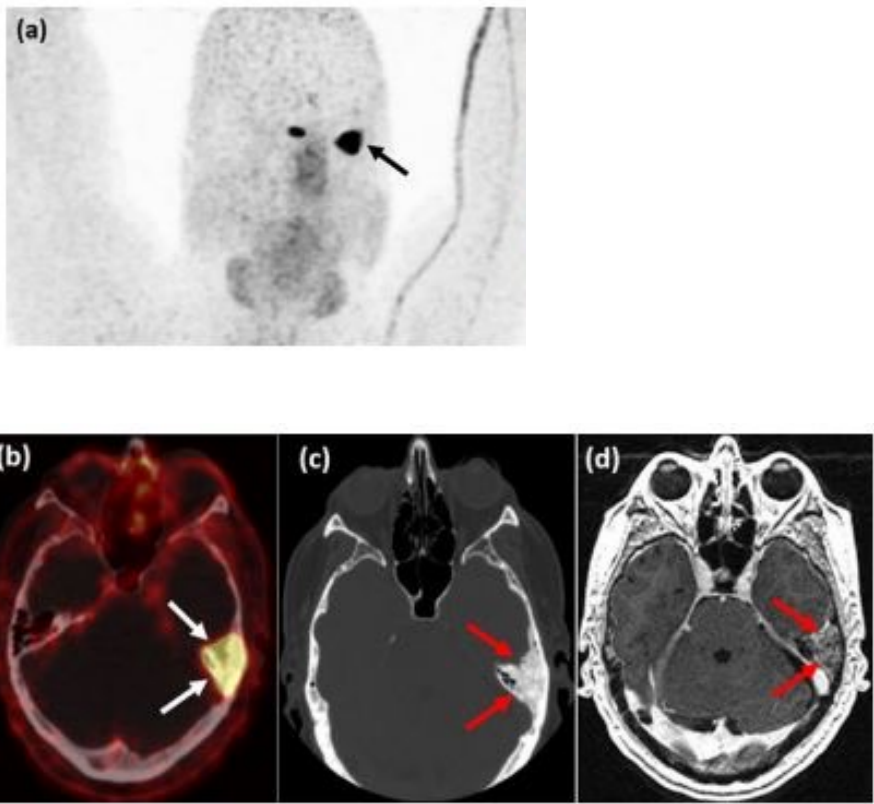
Therapy for Rare Bone Disorder Shows Promise in NIH Clinical Trial



The great imitator

Fibrous dysplasia mimicking malignancy on ⁶⁸Ga-DOTATATE PET/CT

Georgios Z. Papadakis, MD^a, Corina Millo, MD^b, Samira M. Sadowski, MD^{c,d}, Apostolos H. Karantanas, MD, PhD^e, Ulas Bagci, PhD^f, and Nicholas J. Patronas, MD^a

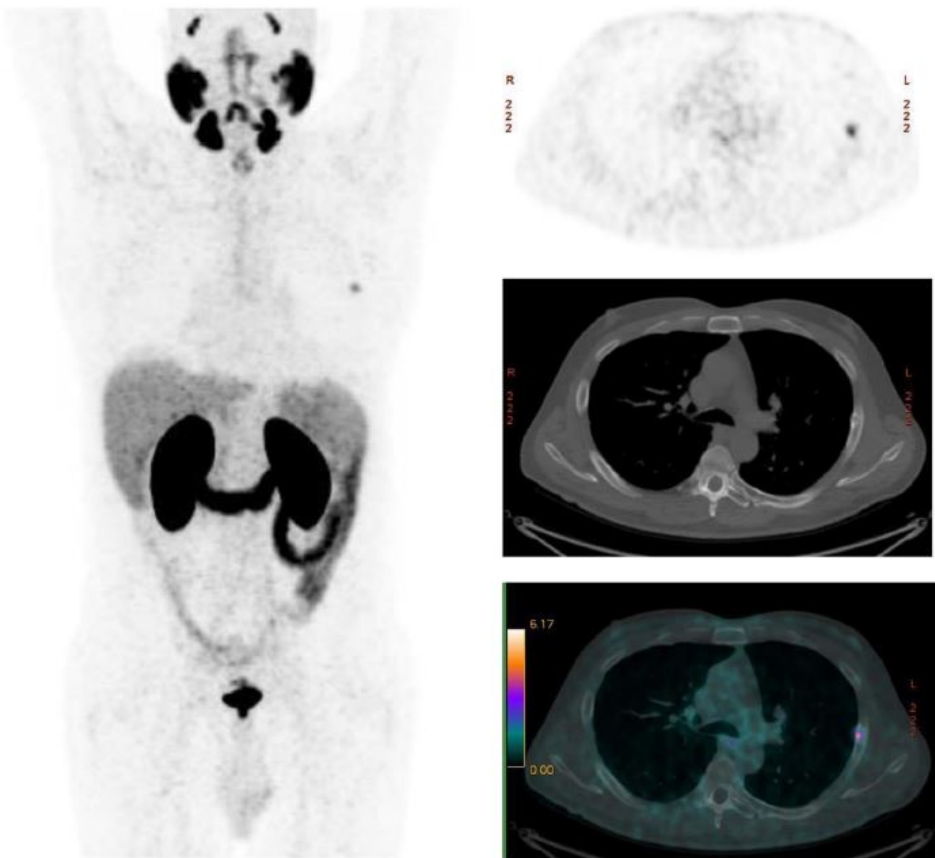


CNM, Volume 42, Number 3, March 2017

INTERESTING IMAGE

⁶⁸Ga-PSMA Uptake in Fibrous Dysplasia

Maria Lucia Reale, MD,* Consuelo Buttigliero, MD,* Marcello Tucci, MD,*
Roberto Giardino, MD,† and Carlo Poti, MD‡

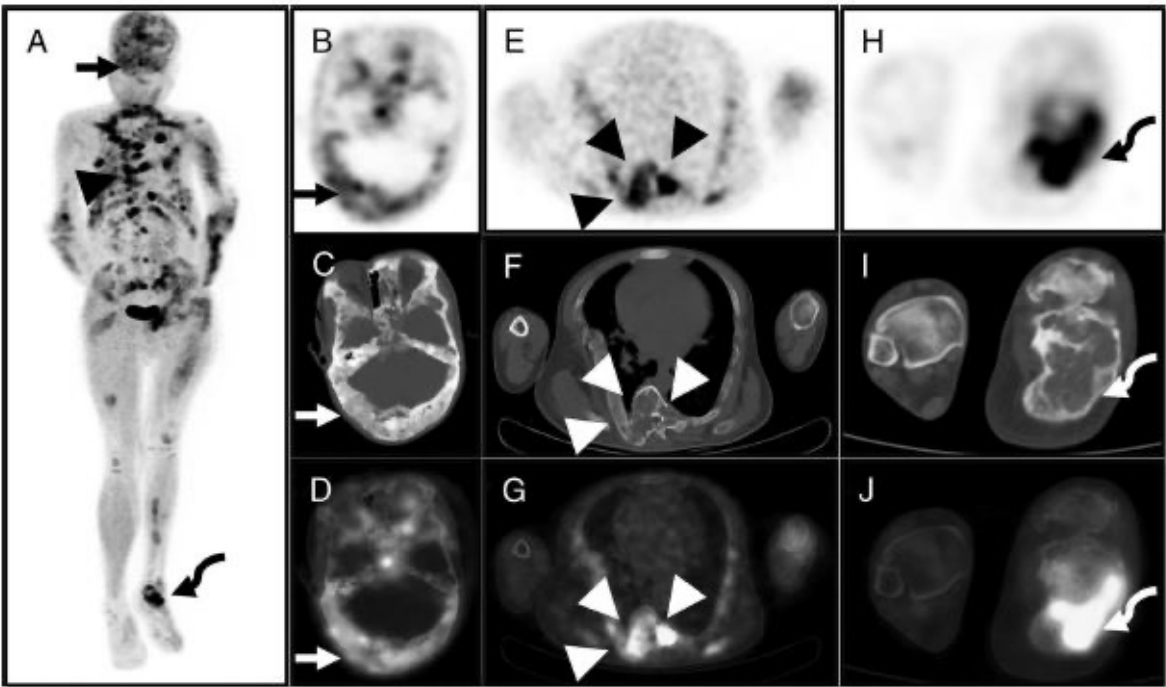


CNM, Volume 44, Number 6, June 2019

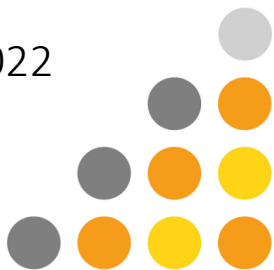
INTERESTING IMAGE

⁶⁸Ga-FAPI-04 PET/CT Imaging for Fibrous Dysplasia of the Bone

Yingwei Wang, MD, Junhao Wu, MD, Lin Liu, MD, DengSai Peng, MD, and Yue Chen, MD



CNM, Volume 47, Number 1, January 2022



Tip of the iceberg



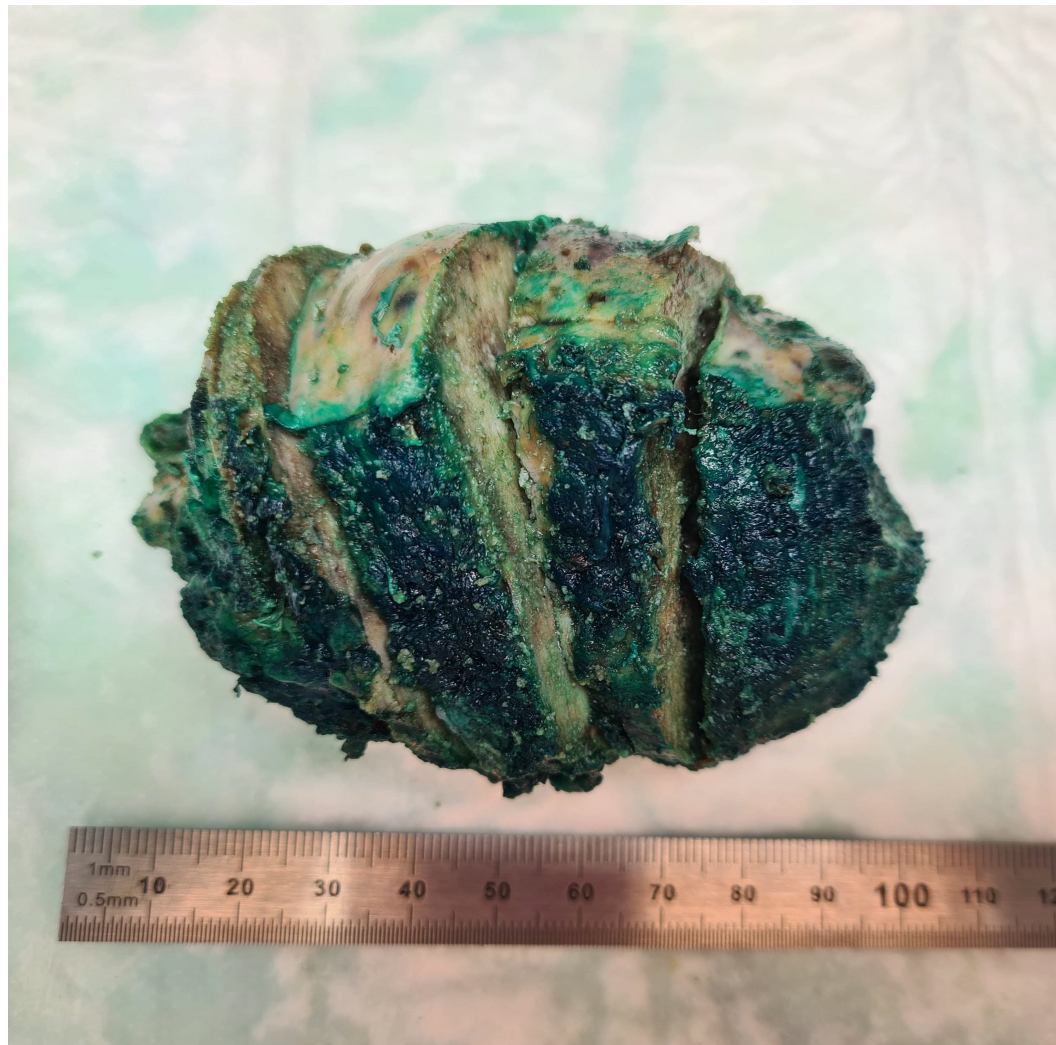
“The highest SUVmax reported in the literature for a benign fibrous dysplasia lesion was a SUVmax of 11.4, reported by Su et al.”



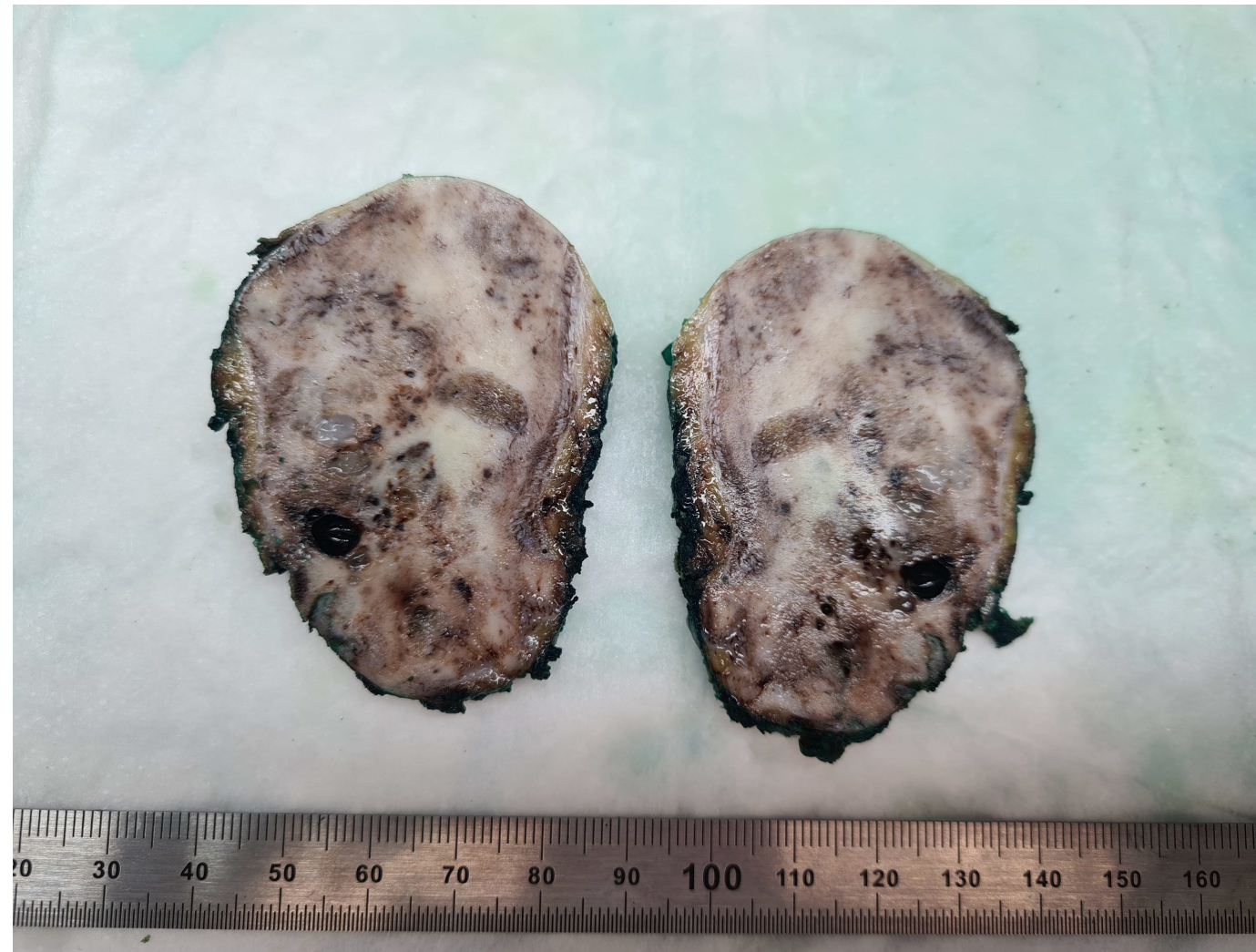
SUVmax 12.5 !!!



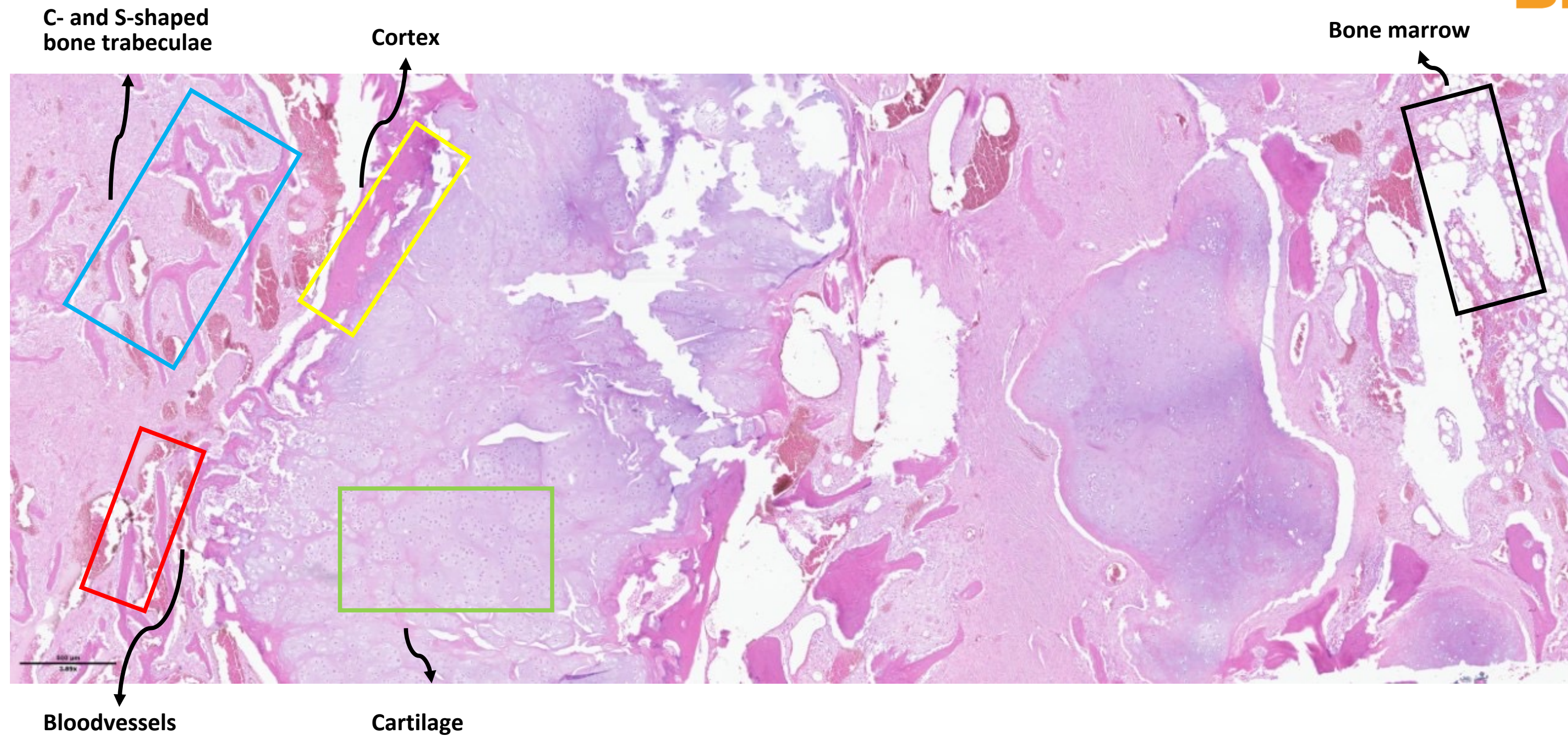
Resection



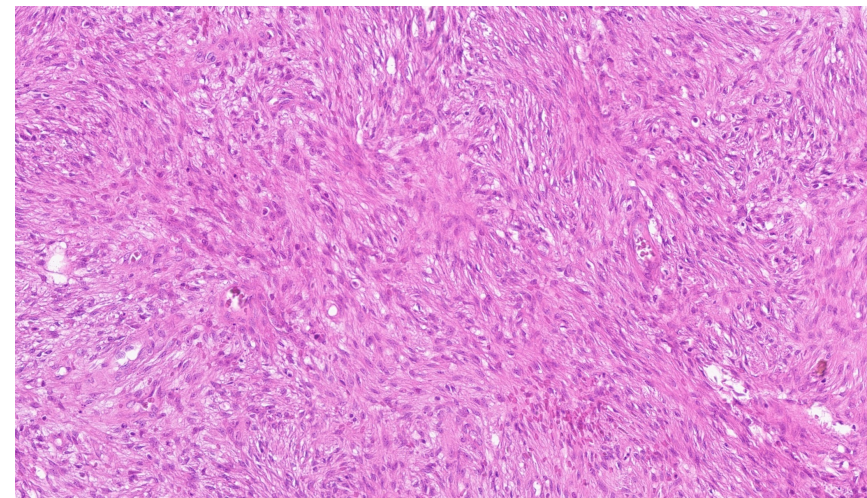
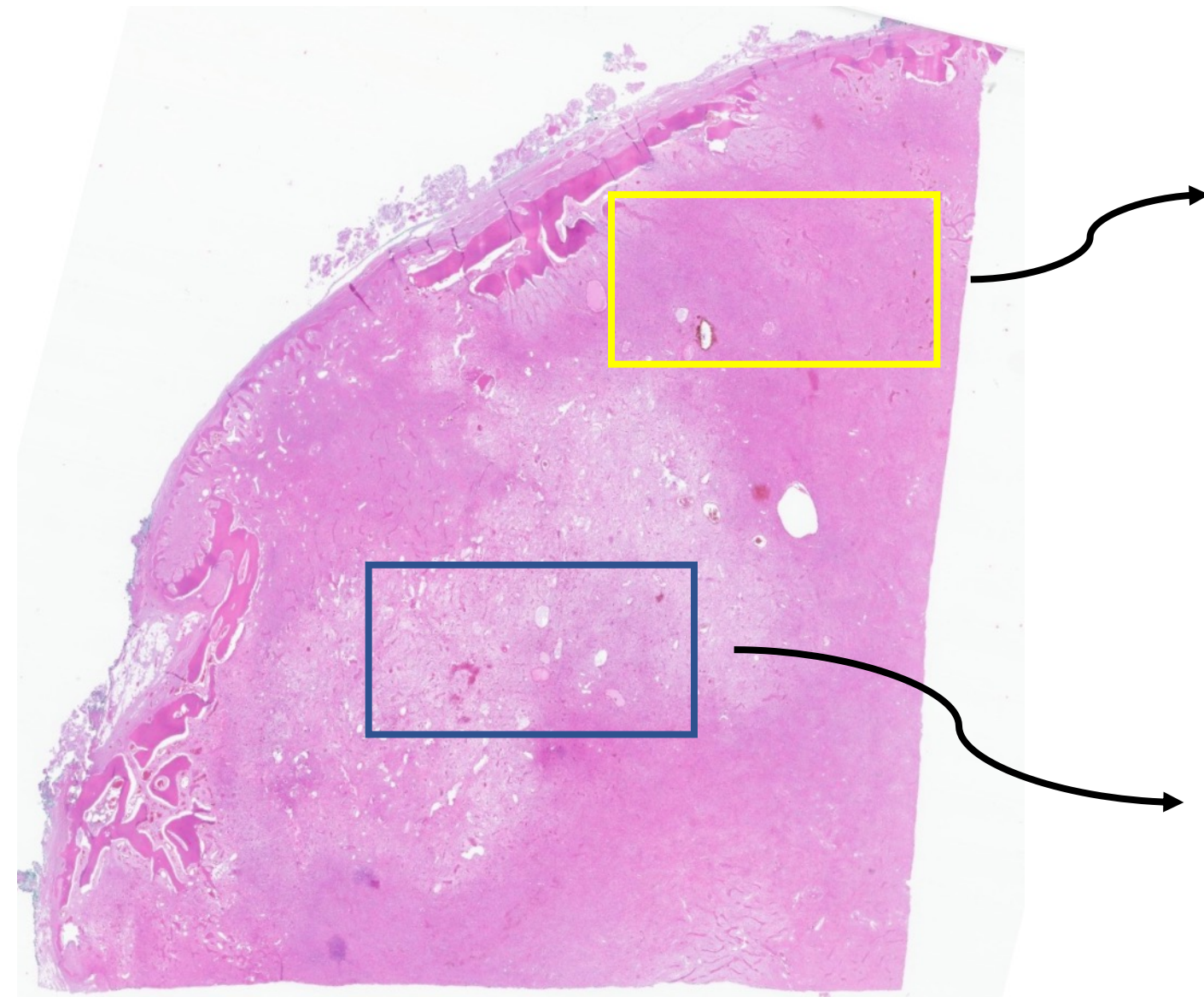
10 x 6,5 x 5,5 cm



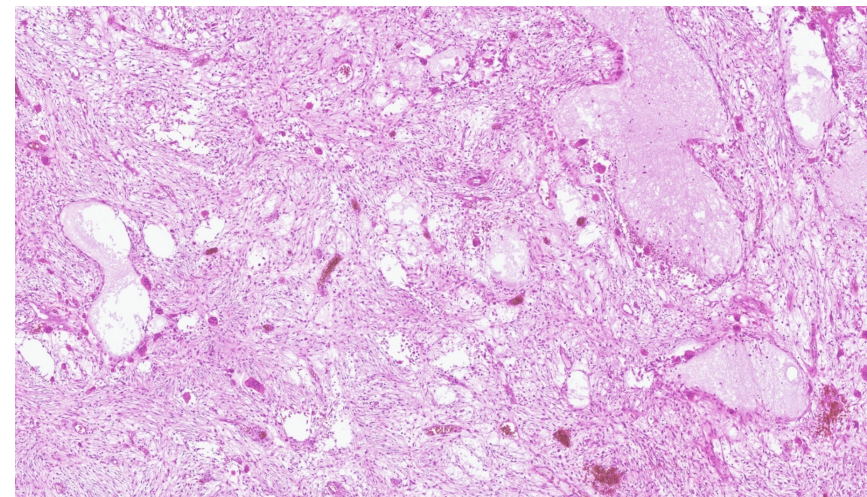
Histopathology



Hypermetabolism?



Inflammation



Vascularization



Take home messages: fibrous dysplasia

- (A)symptomatic disease: therapy can be necessary. Malignant transformation is rare.
- Increased tracer uptake on bonescan, FDG-, SSR-, PSMA- and FAPI-PET imaging. Potential pitfall mimicking malignancy.
- Histopathology is golden standard for diagnosis.
- Nuclear imaging is useful in evaluation of extensiveness of the disease, activity of the lesions and response to therapy.

