diaphyseal leg; partial

21 yo ♂ **1st referral: October 27th, 2015**

HPI:

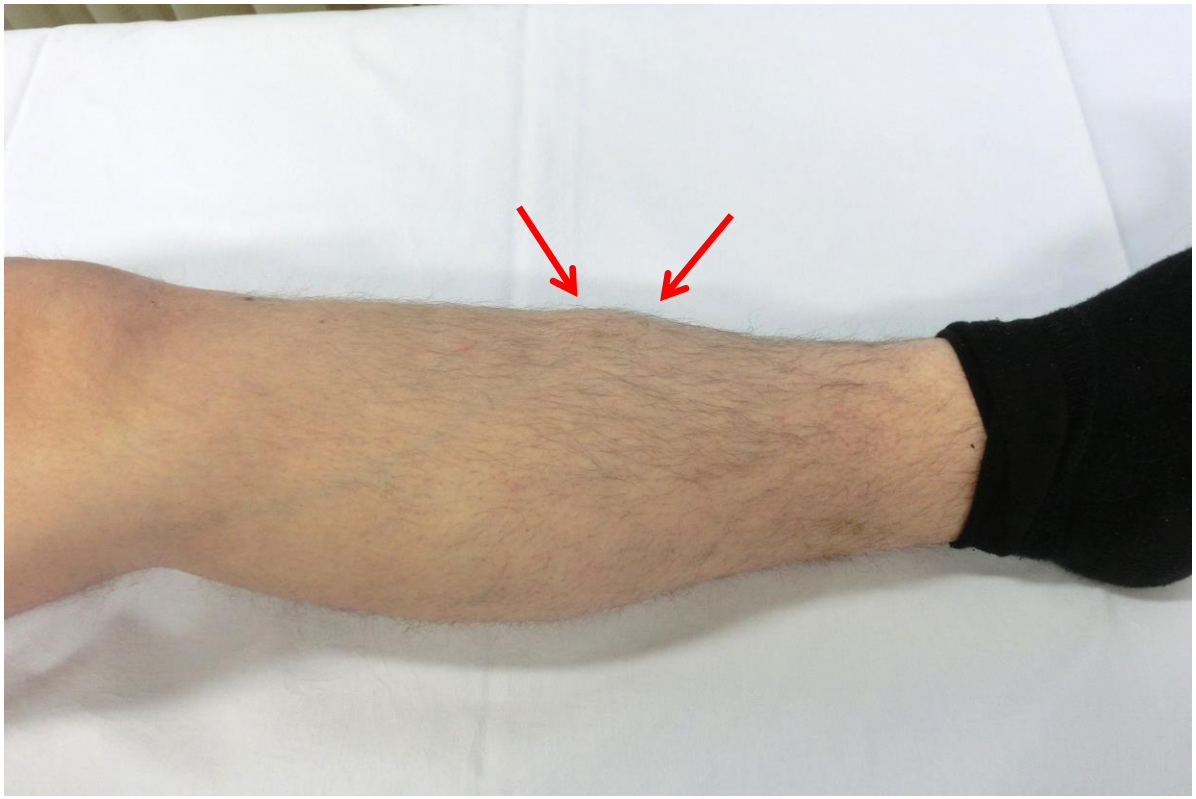
- First notice of a slightly increasing swelling at anterior shin 3 ½ yrs ago
- Pain upon exertion at left shin since summer 2014
- Pain causes sleeping disorders

PMH:

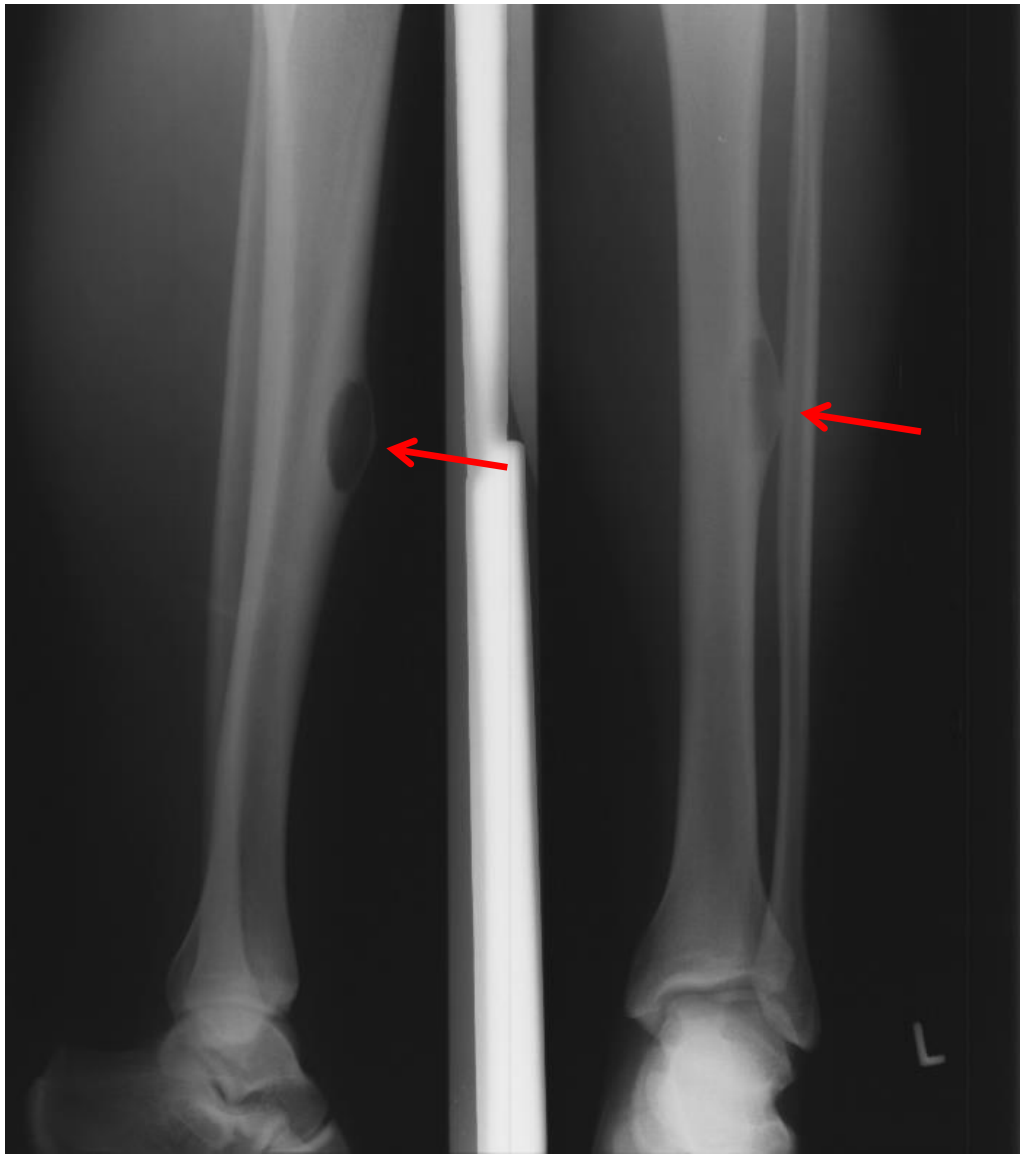
- Bronchial asthma

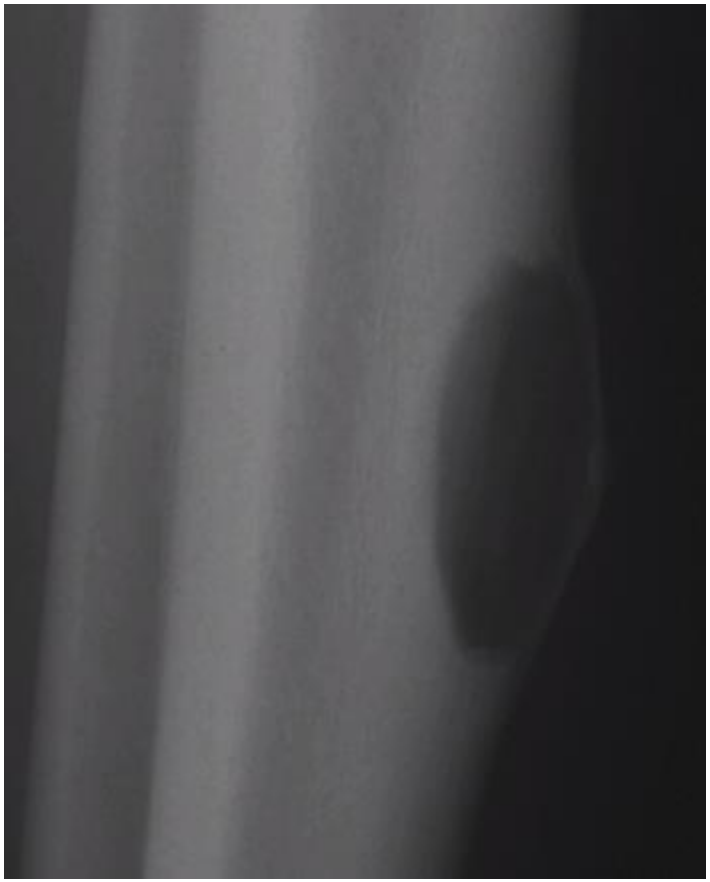
diaphyseal leg; partial
Clinical findings on November 25th, 2015

- Bony prominence at anterior shin (3.5 x 3.5 cm)
- Painful upon palpation, hyperthermic
- No sensomotoric deficits



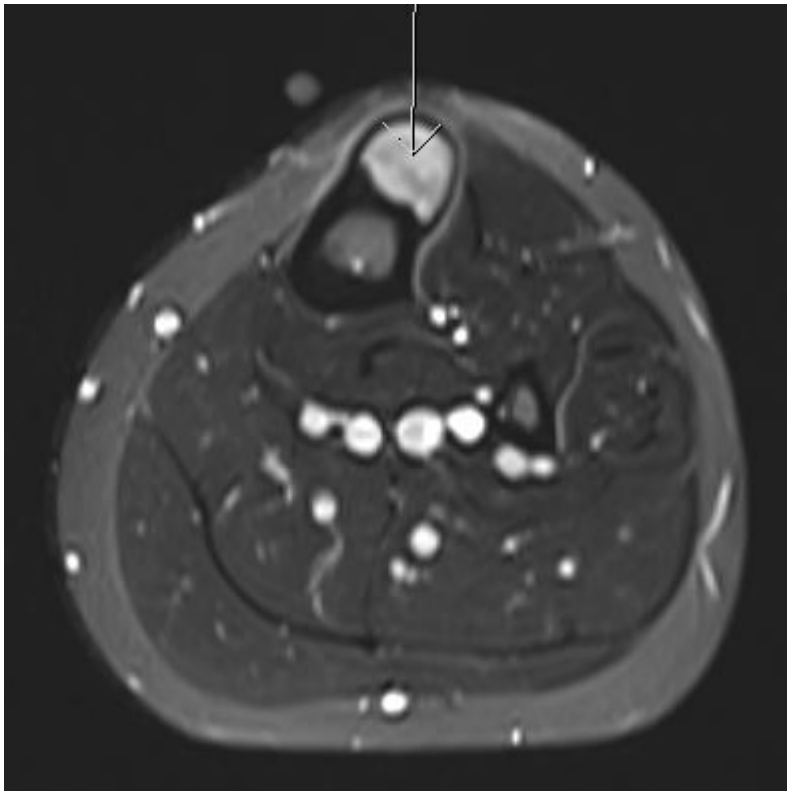
diaphyseal leg; partial
XR October 6th, 2015



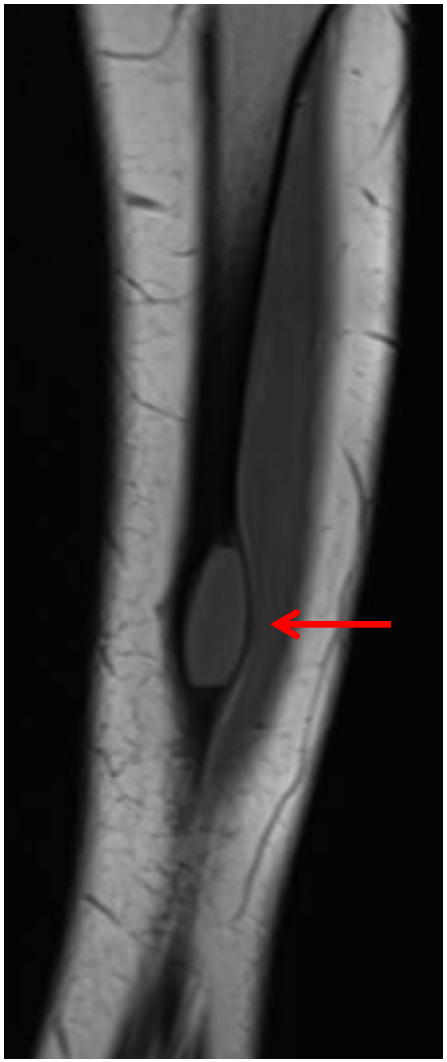


SarcOmaSurgery

diaphyseal leg; partial
MRI October 8th, 2015



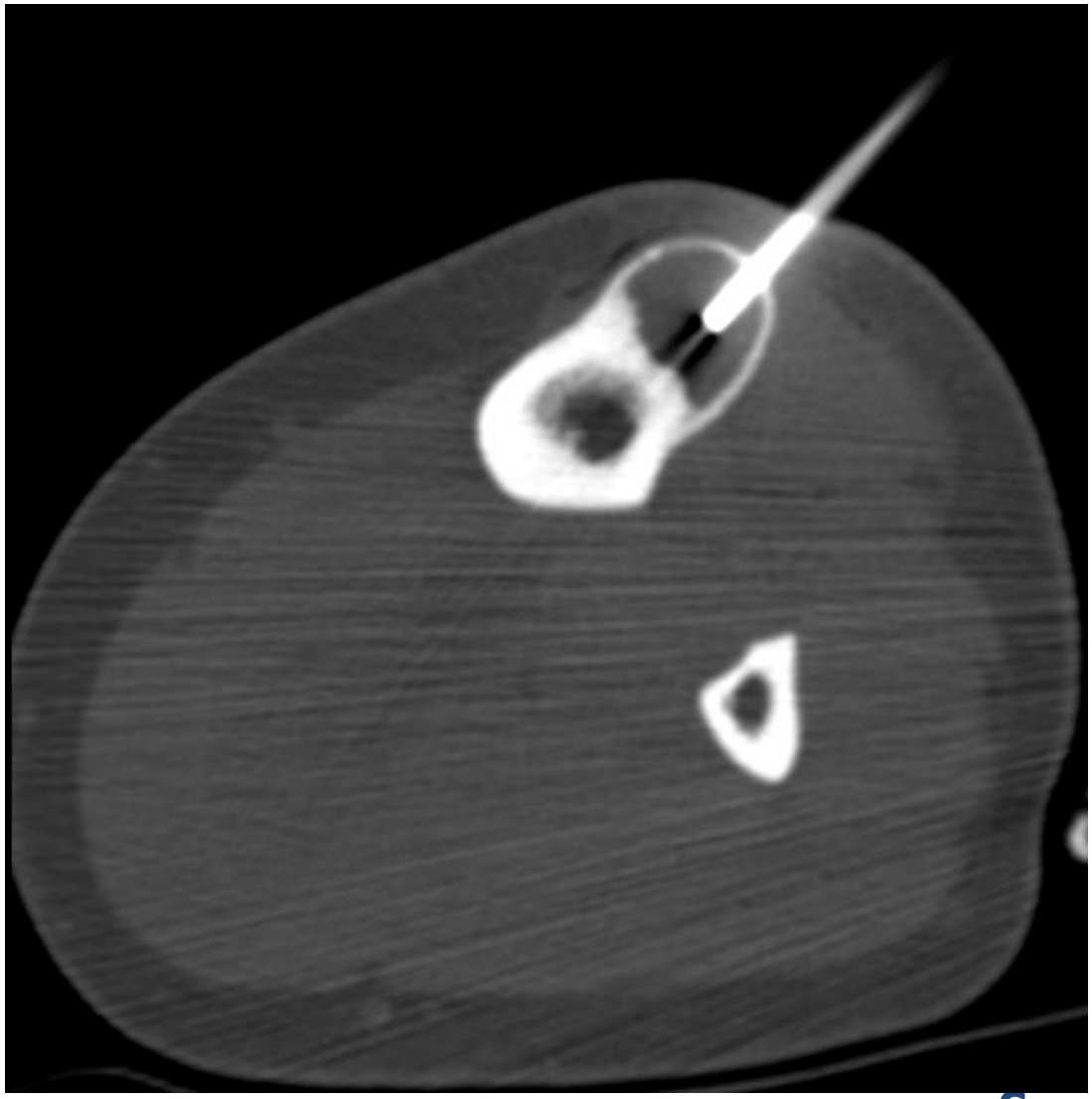
T1_TIRM_ax



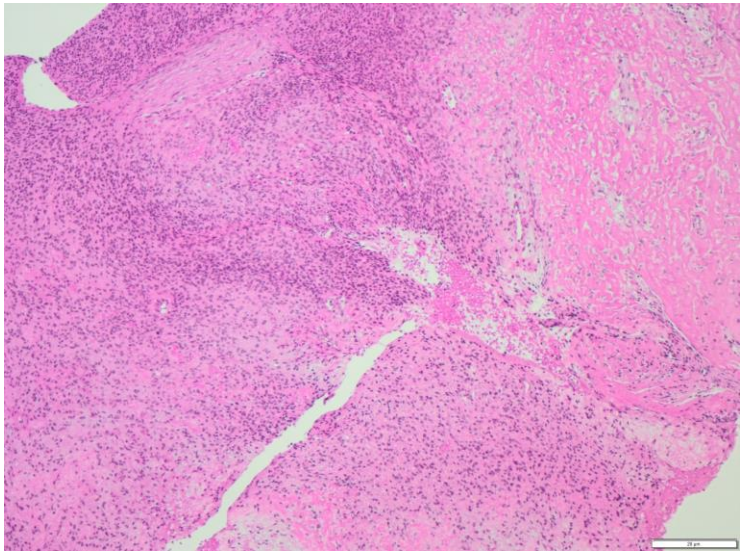
T1_TSE_cor

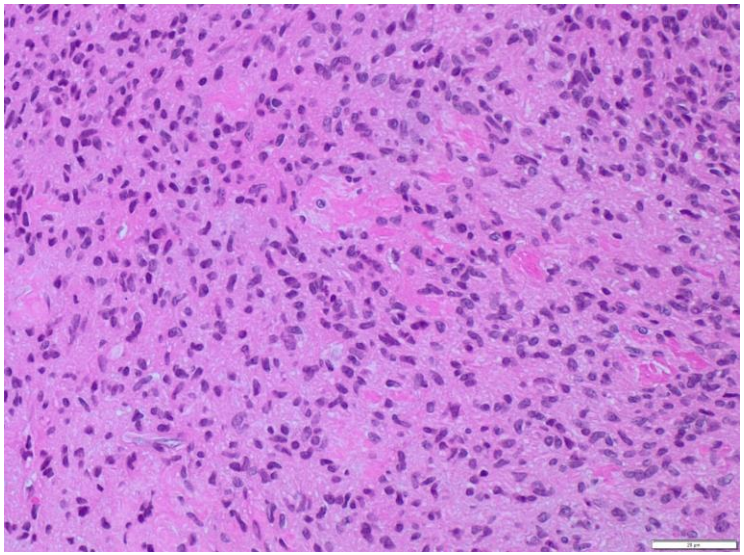
SarcOmaSurgery

diaphyseal leg; partial
CT-guided needle biopsy November 25th, 2015



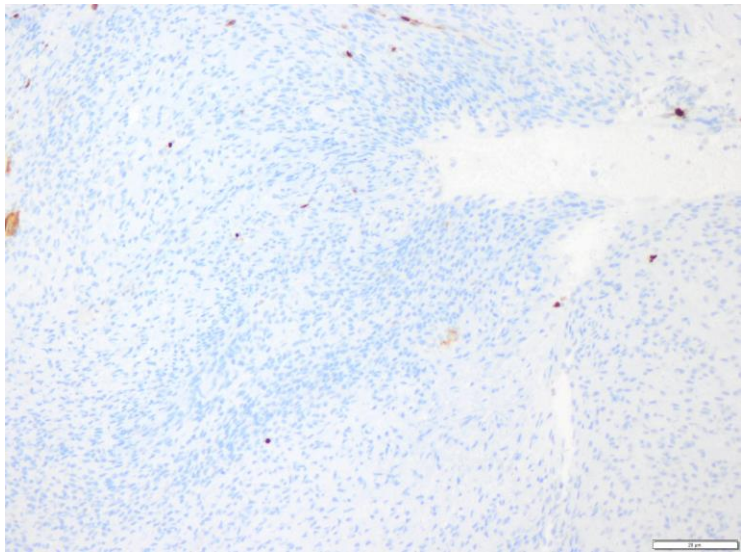
diaphyseal leg; partial
Histological Findings November 25th, 2015





SarcOmaSurgery

diaphyseal leg; partial
Histological Findings November 25th, 2015



CK – Neg

Keine osteofibröse Dysplasie
Kein Adamantimom

Kein high grade Tumor

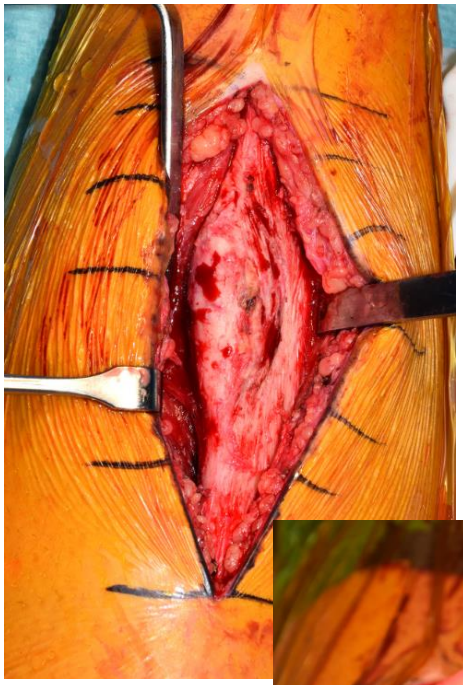
DD

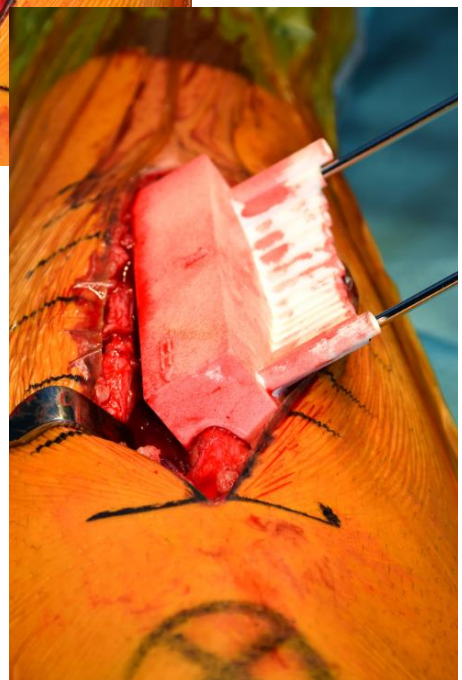
-Chondromyxoid-Fibrom
-Phosphaturischer mesenchymaler Tumor

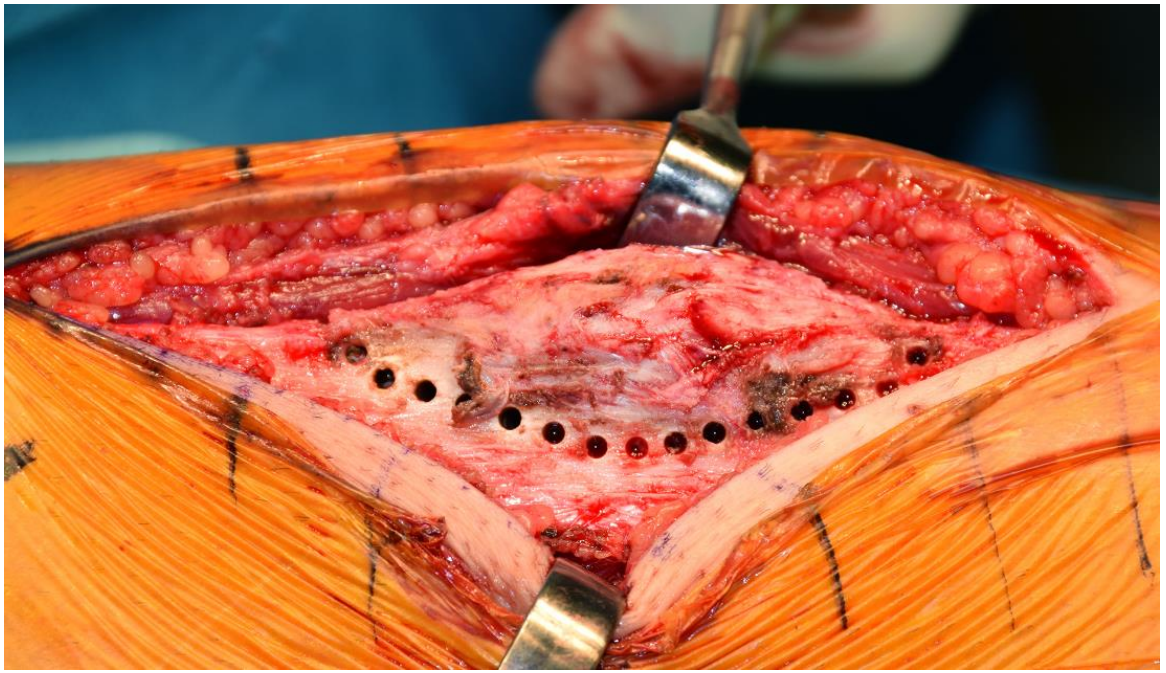
KONSIL
-DD low grade (intrakortikales??) OSA?

SarcOmaSurgery

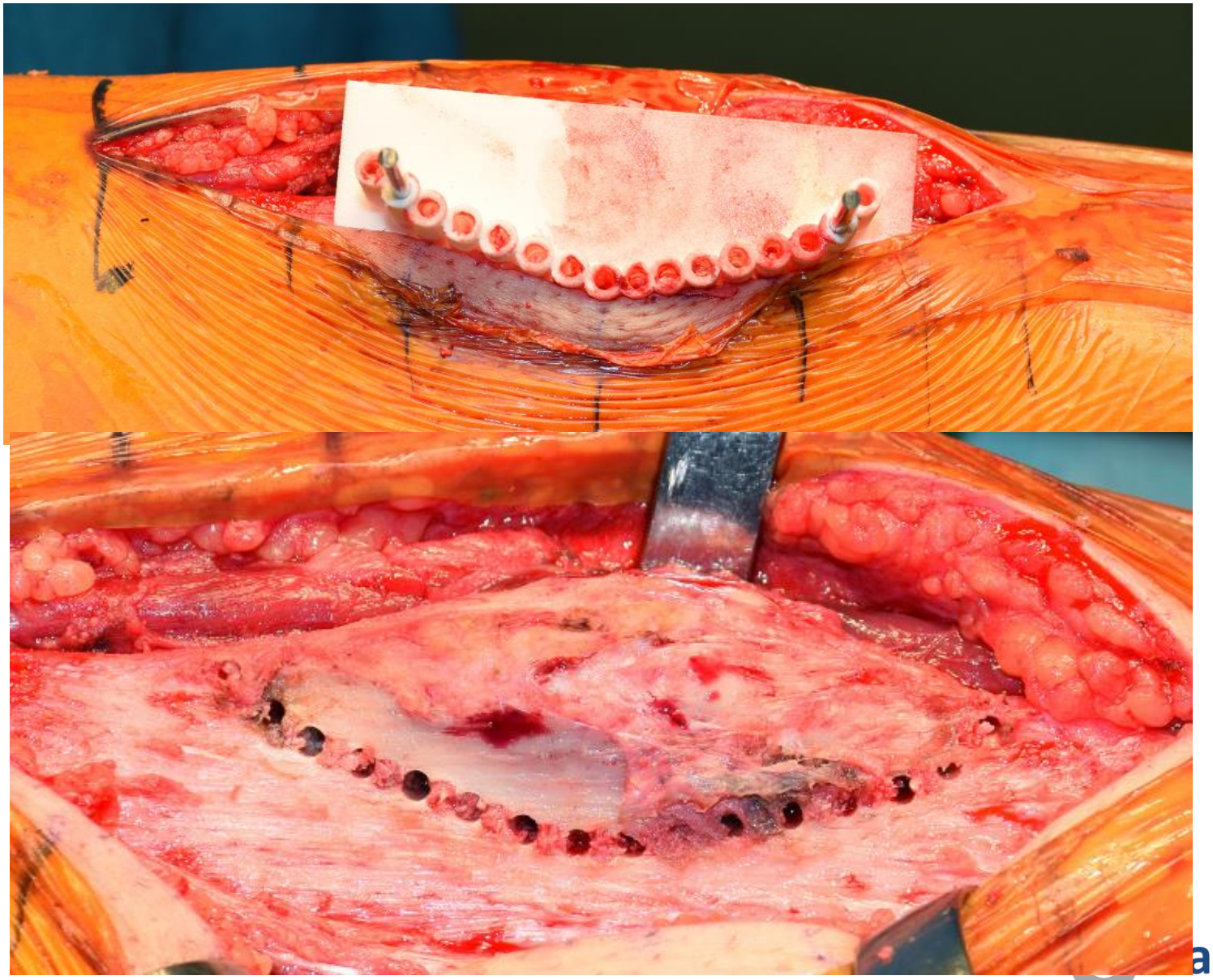
diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016



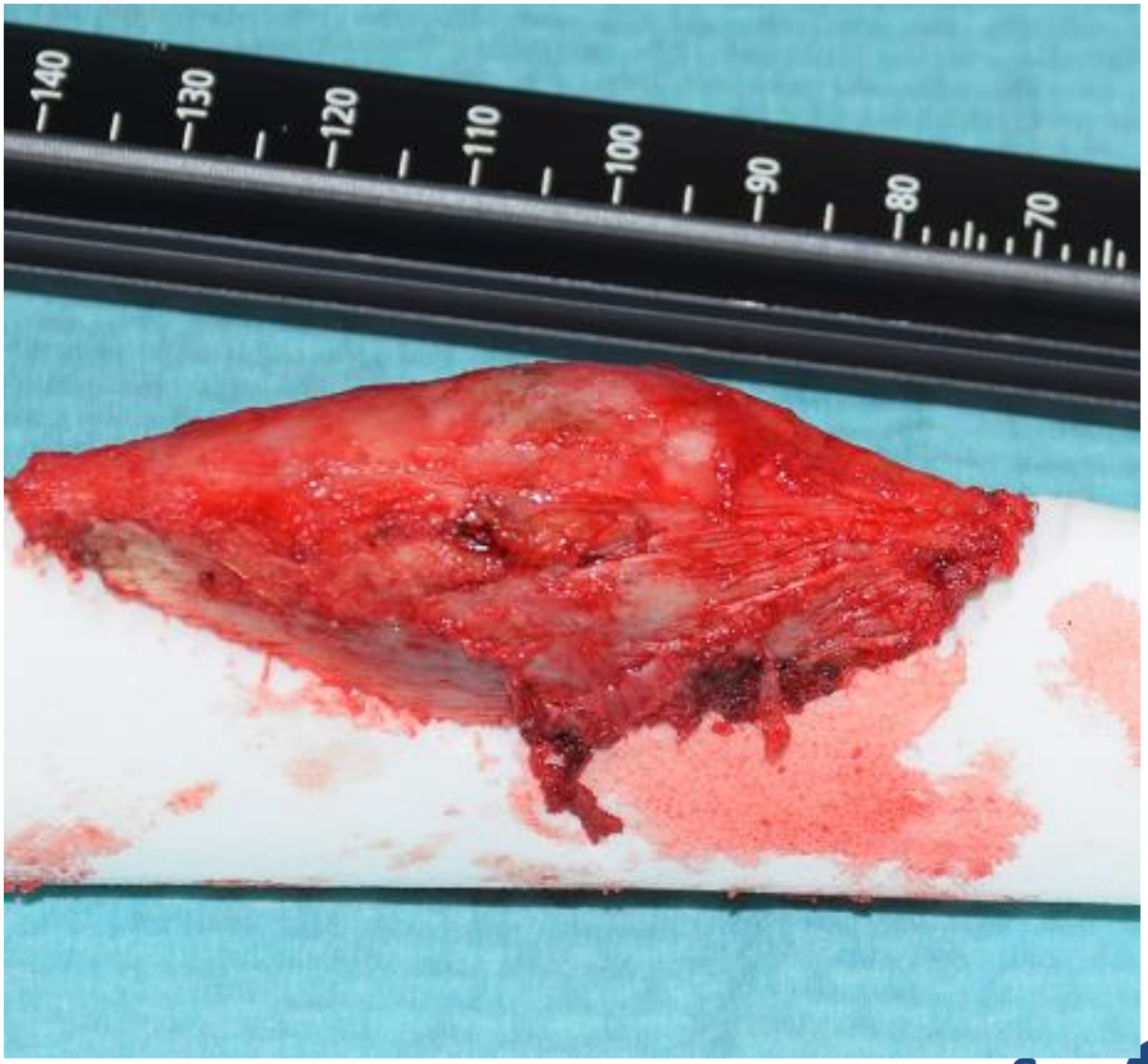




diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016

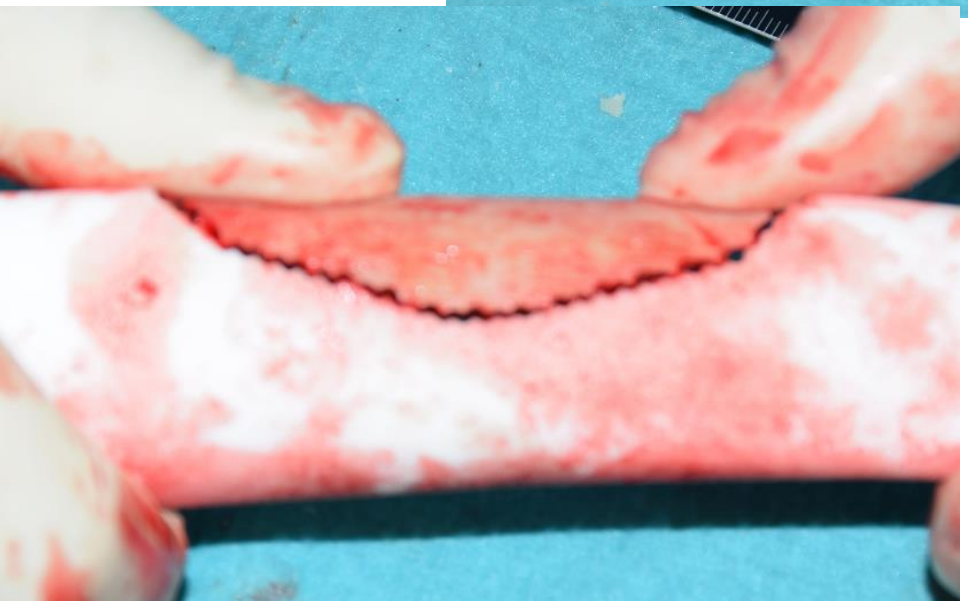


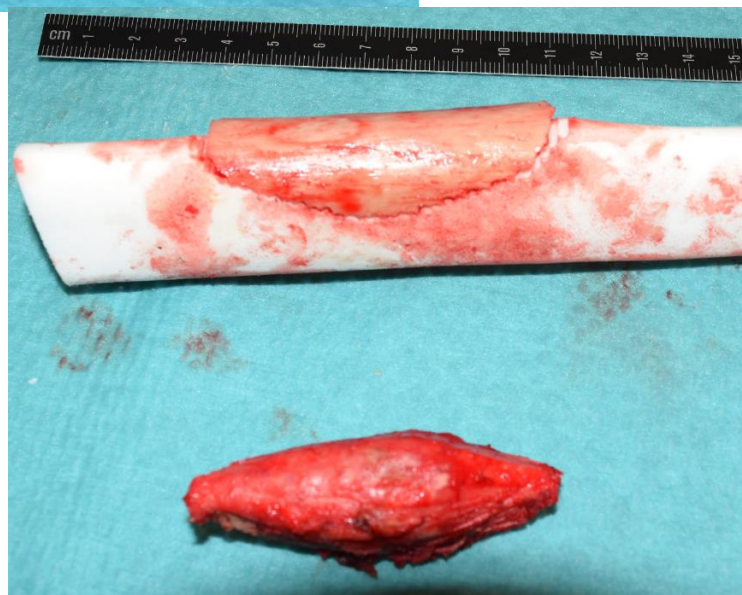
diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016



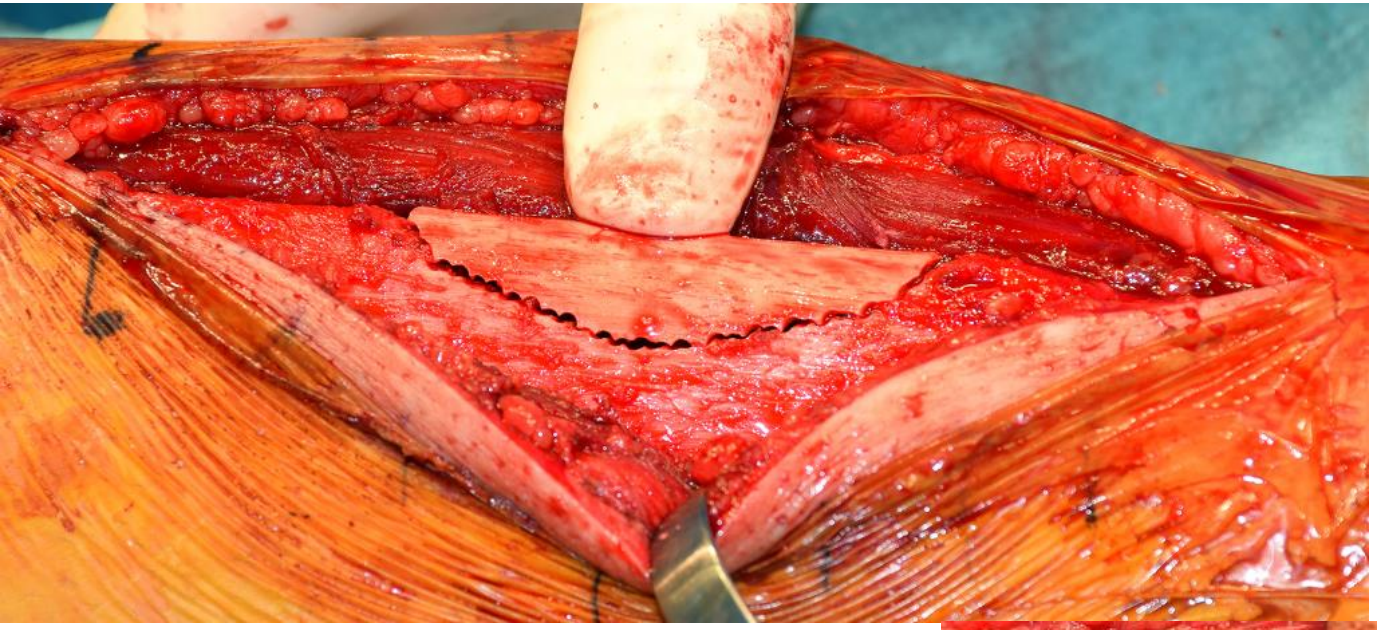
SarcOmaSurgery

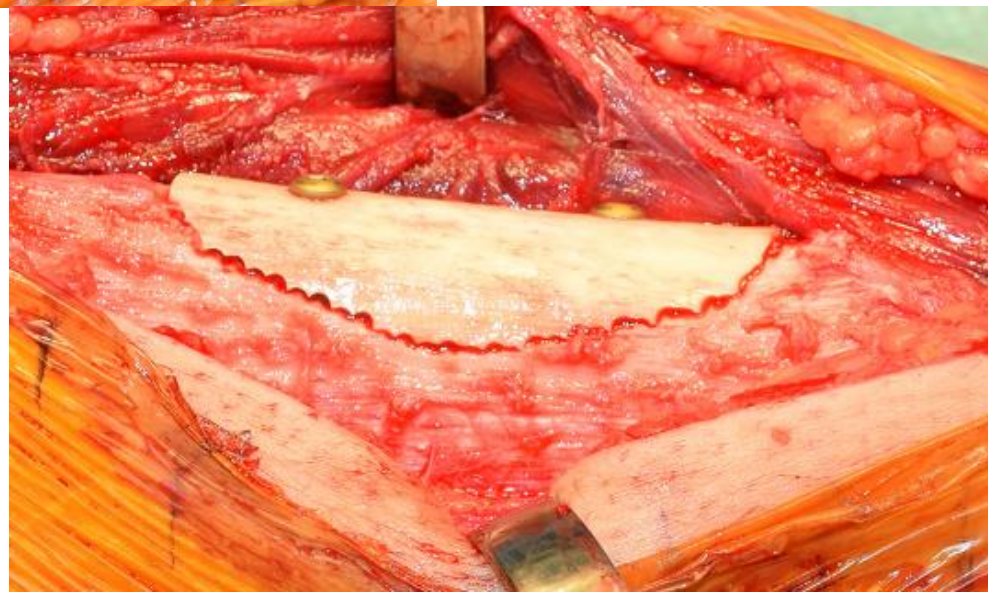
diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016



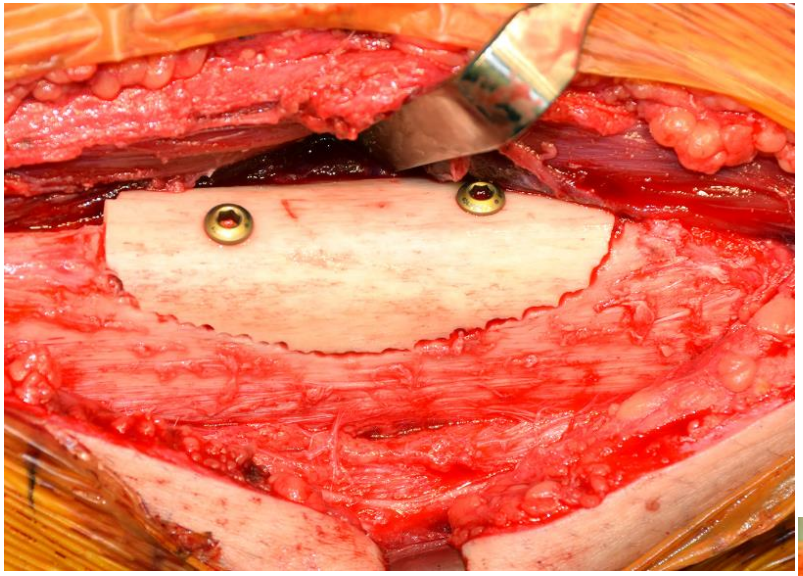


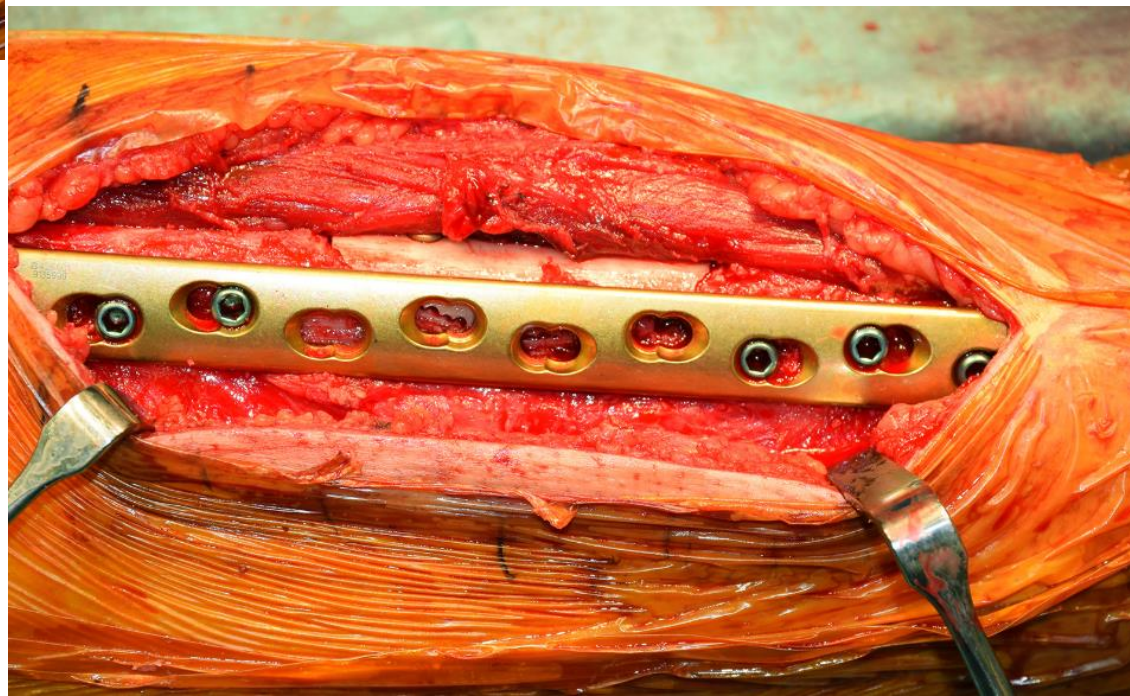
diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016



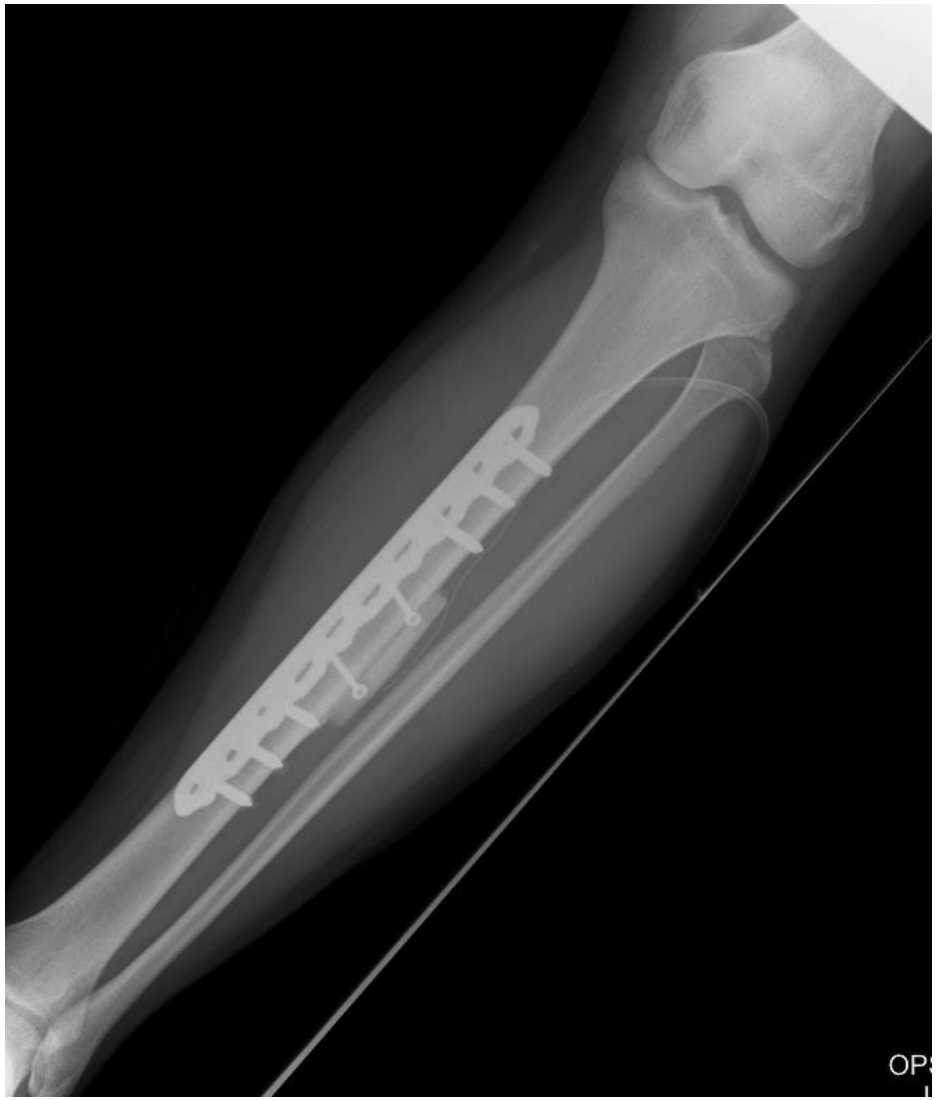


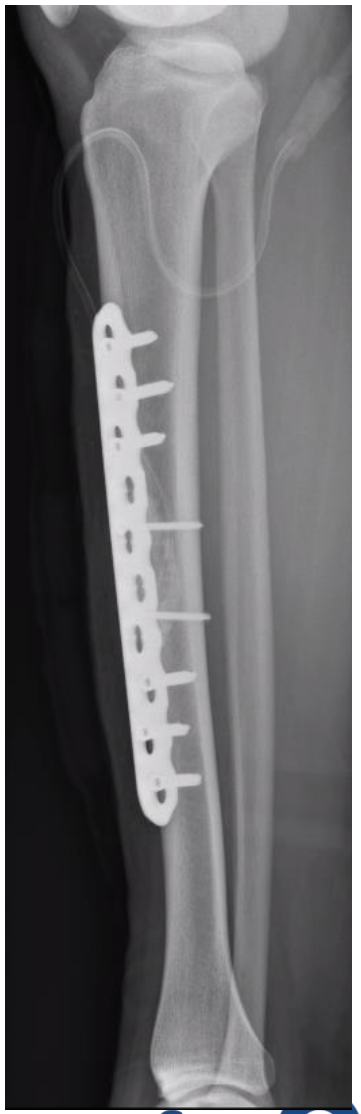
diaphyseal leg; partial
Surgery: Tumorresection March 15th 2016



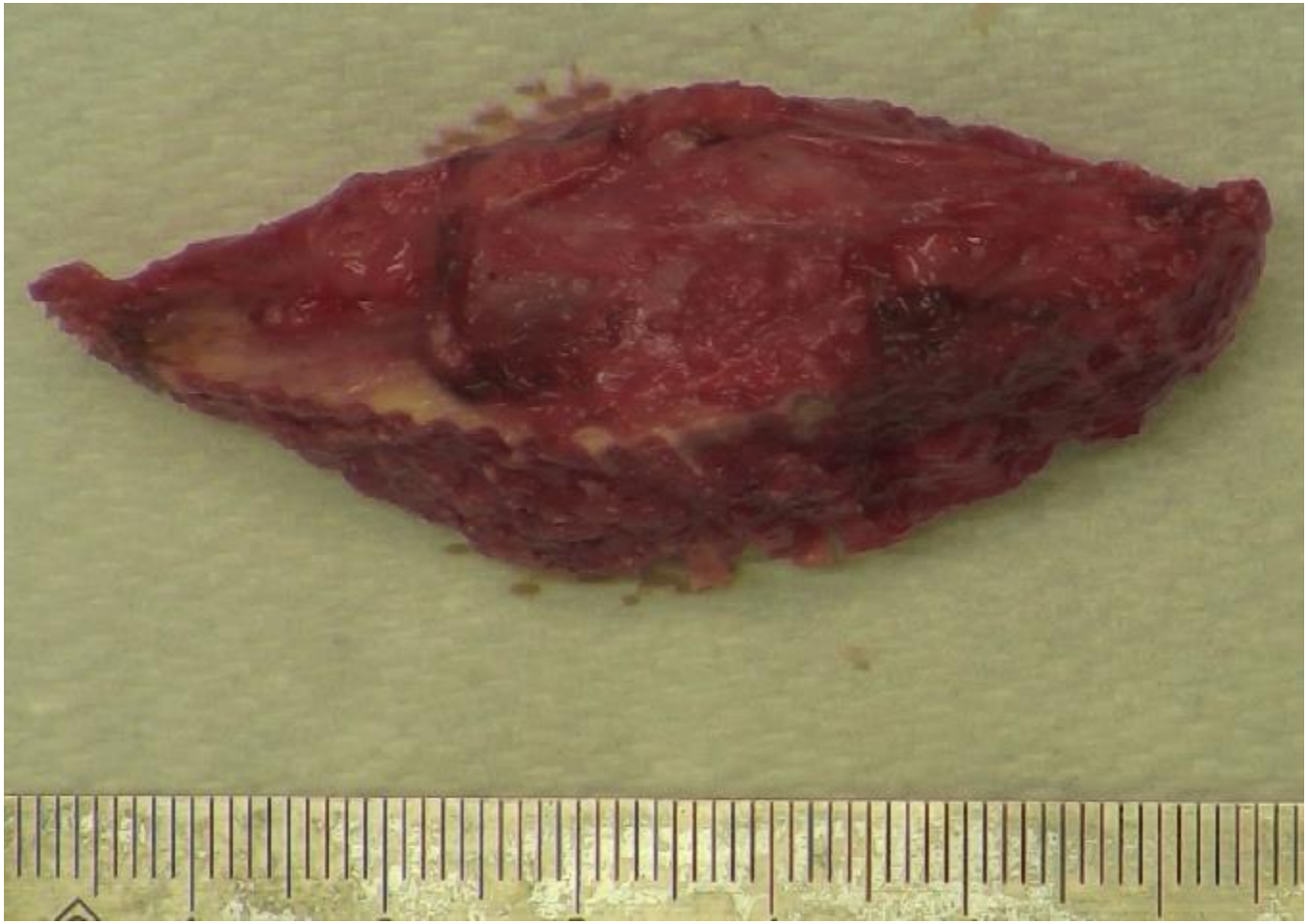


diaphyseal leg; partial
Postoperativ X-Rays 15.03.2016



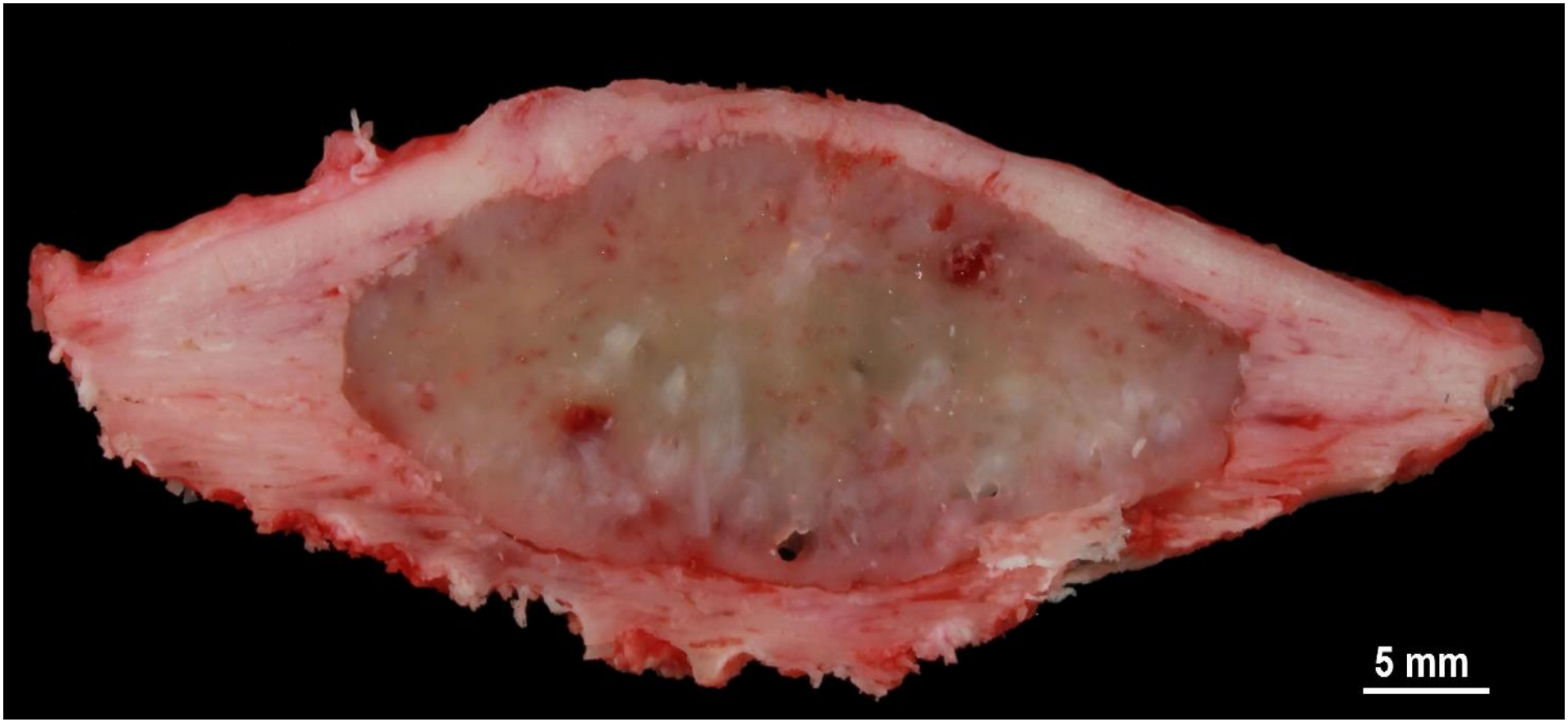


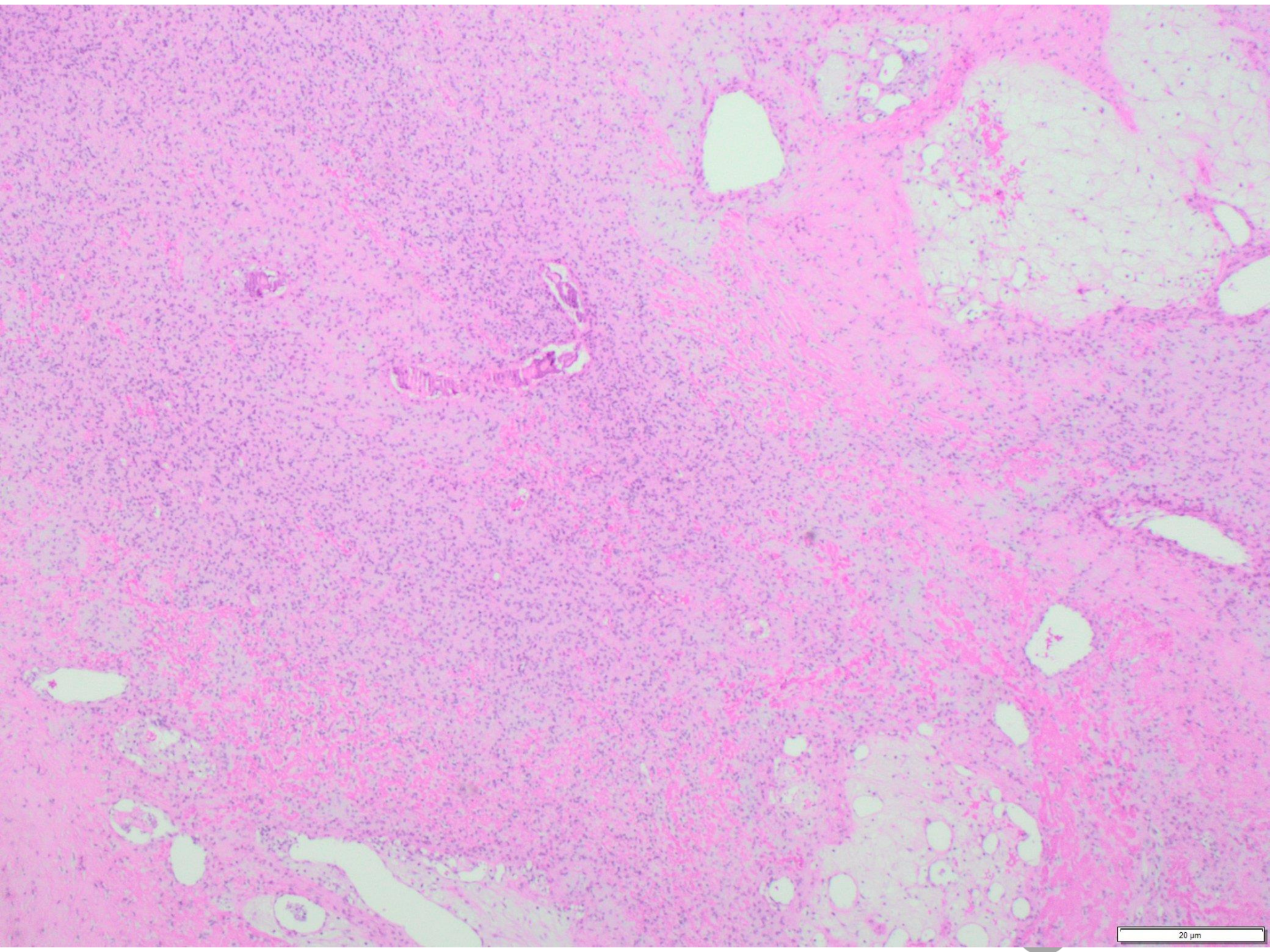
diaphyseal leg; partial
Histological Results March 15th 2016

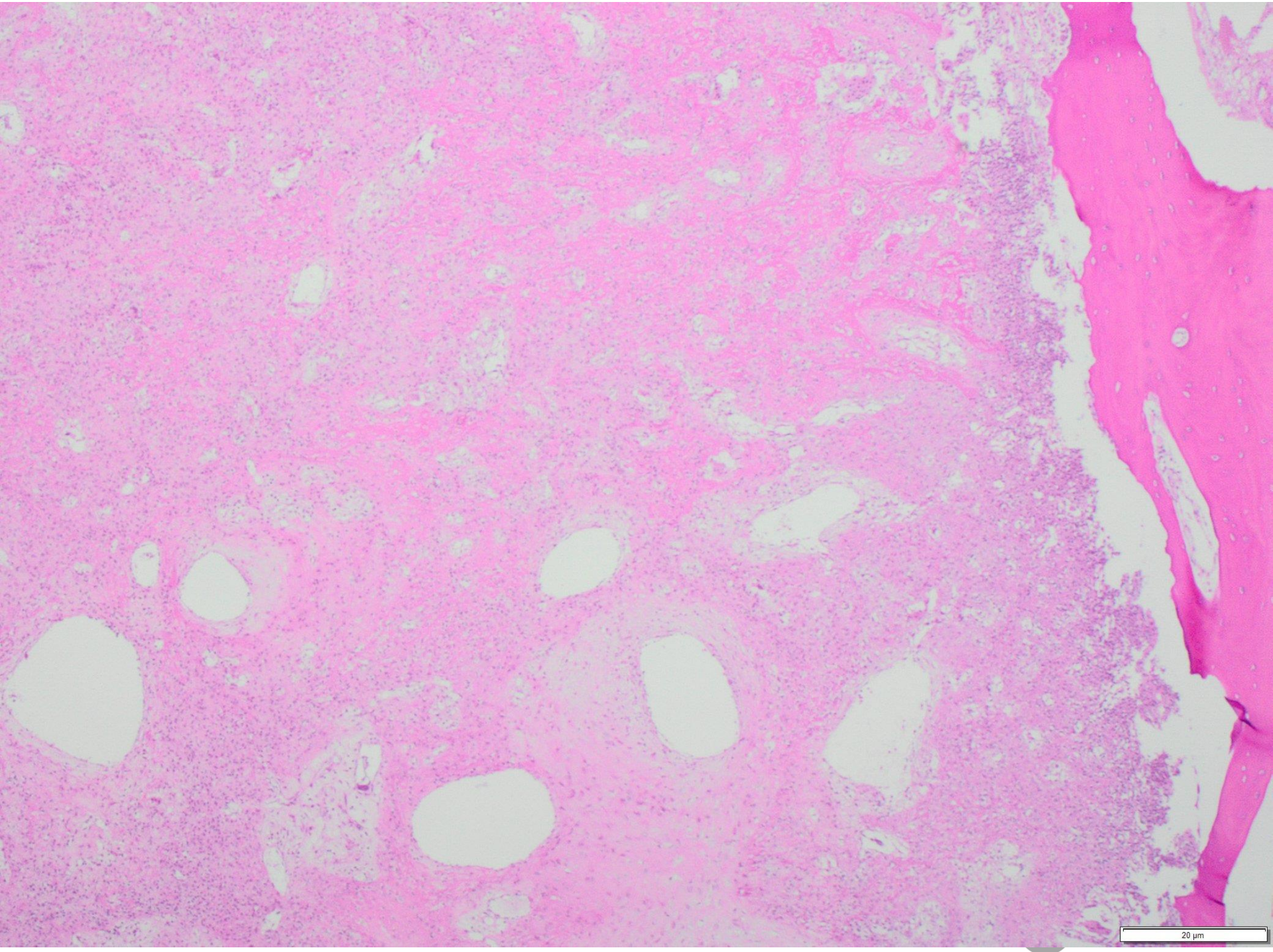


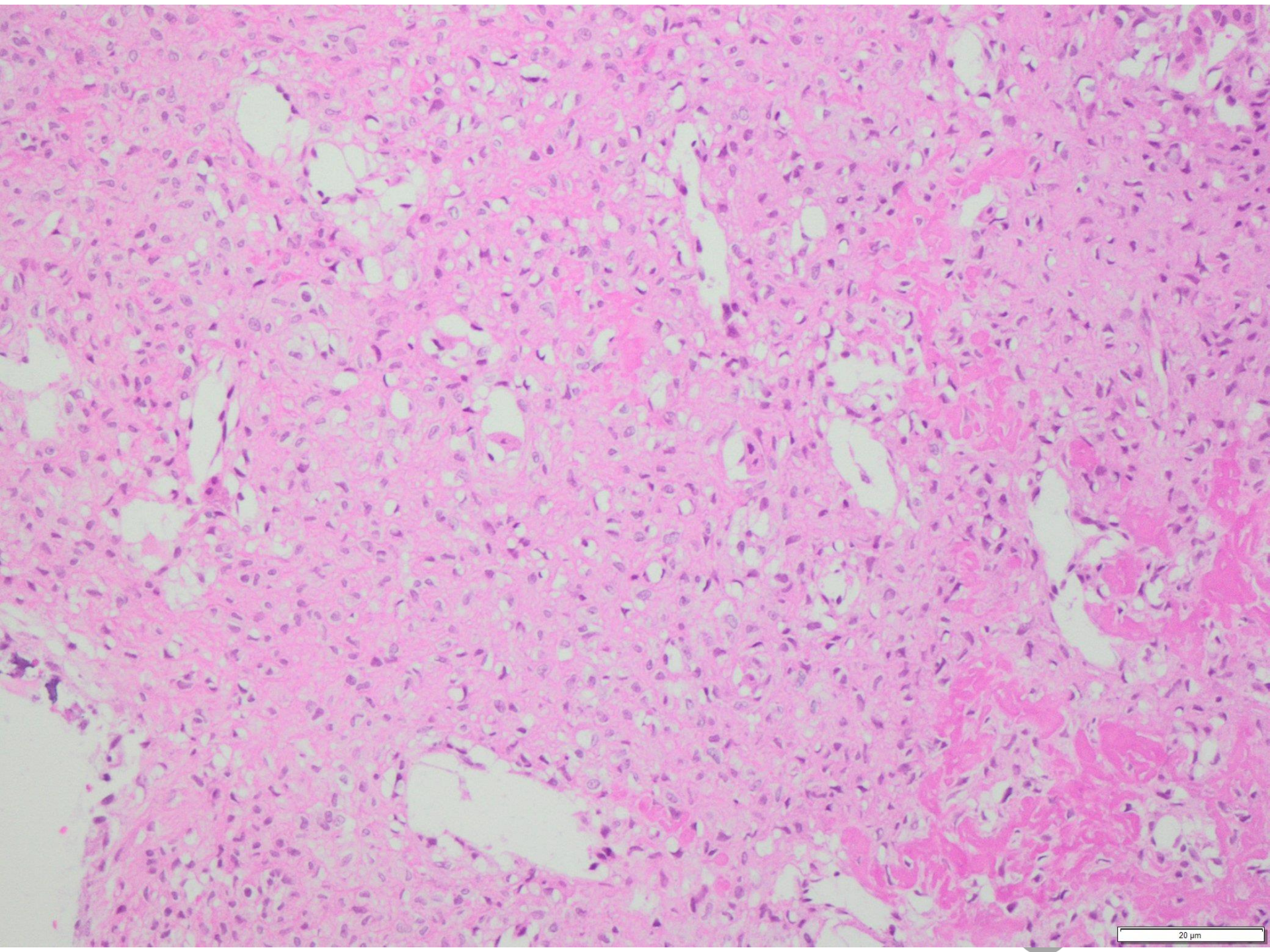
SarcomaSurgery

diaphyseal leg; partial
Histological Results March 15th 2016




20 µm


20 µm


20 µm

Phosphaturic mesenchymal tumour

A.L. Folpe

Definition

Phosphaturic mesenchymal tumours are morphologically distinctive neoplasms that produce tumour-induced osteomalacia (TIO) in most affected patients, usually through production of fibroblast growth factor 23 (FGF23).

ICD-O code

Phosphaturic mesenchymal tumour 8990/0
Phosphaturic mesenchymal tumour, malignant 8990/3

Synonym

Phosphaturic mesenchymal tumour, mixed connective tissue type

Epidemiology

Phosphaturic mesenchymal tumours are exceptionally rare, with fewer than 250 reported cases {140,245,595,884,1494,2919}. They occur most frequently in middle-aged adults {140,884}, although they have been reported in infants {1352} and the elderly.

Sites of involvement

Phosphaturic mesenchymal tumours may involve essentially any soft-tissue location {140,245,884,2919}. They are extremely rare in the retroperitoneum, viscera and mediastinum {2506,2797}.

Clinical features

Most tumours present as small, inapparent lesions that may require careful clinical examination and radionuclide scans for localization {946,2555}. A long history of osteomalacia is usually, but not always, present. Phosphaturic mesenchymal tumours appear to be responsible for the overwhelming majority of previously reported cases of mesenchymal tumour-associated TIO, although many such cases have been reported with other diagnoses {884}. Some tumours may be identified before osteomalacia becomes clinically evident {140}.

Macroscopy

Most phosphaturic mesenchymal tumours present as nonspecific soft tissue or bone masses, often with a component of fat. Some may be highly calcified.

Histopathology

These tumours are usually composed of

Prognostic factors

The overwhelming majority are histologically and clinically benign. Morphologically benign cases frequently recur locally, but are cured with complete excision, with resolution of osteomalacia {595, 884,968,1494}. Malignant tumours may metastasize and cause death from disease.