1

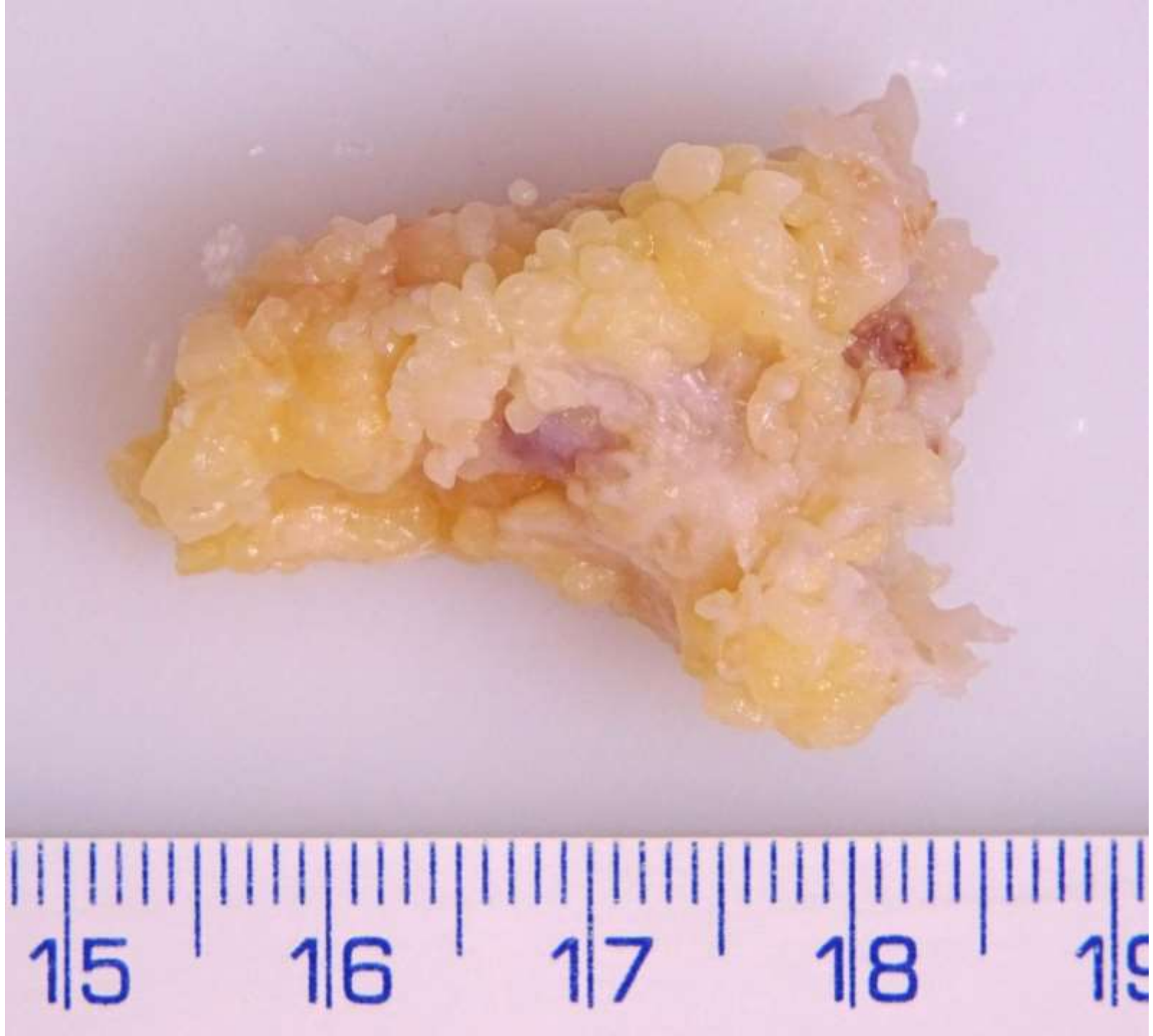
L2025-1

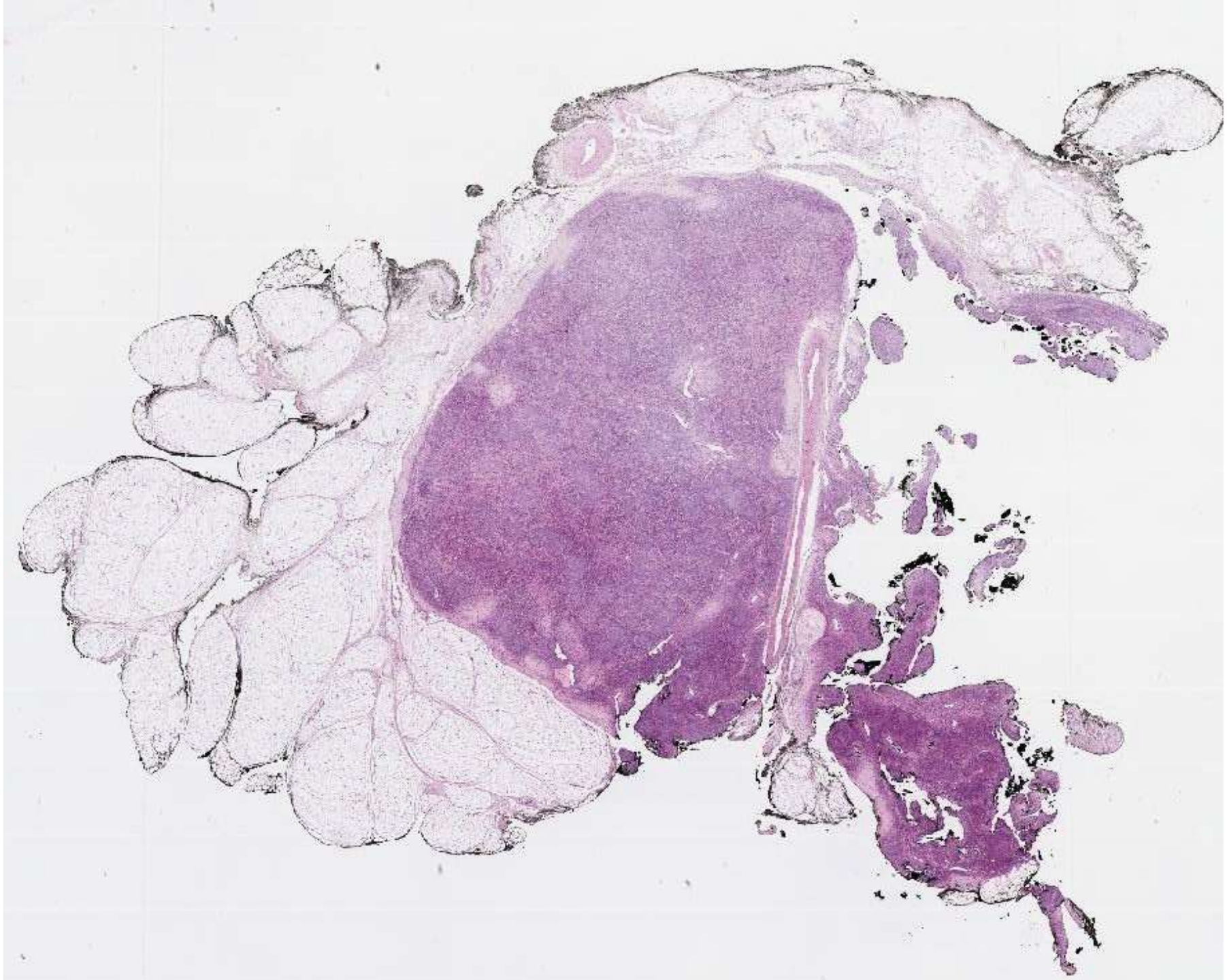
Tomáš Rozkoš

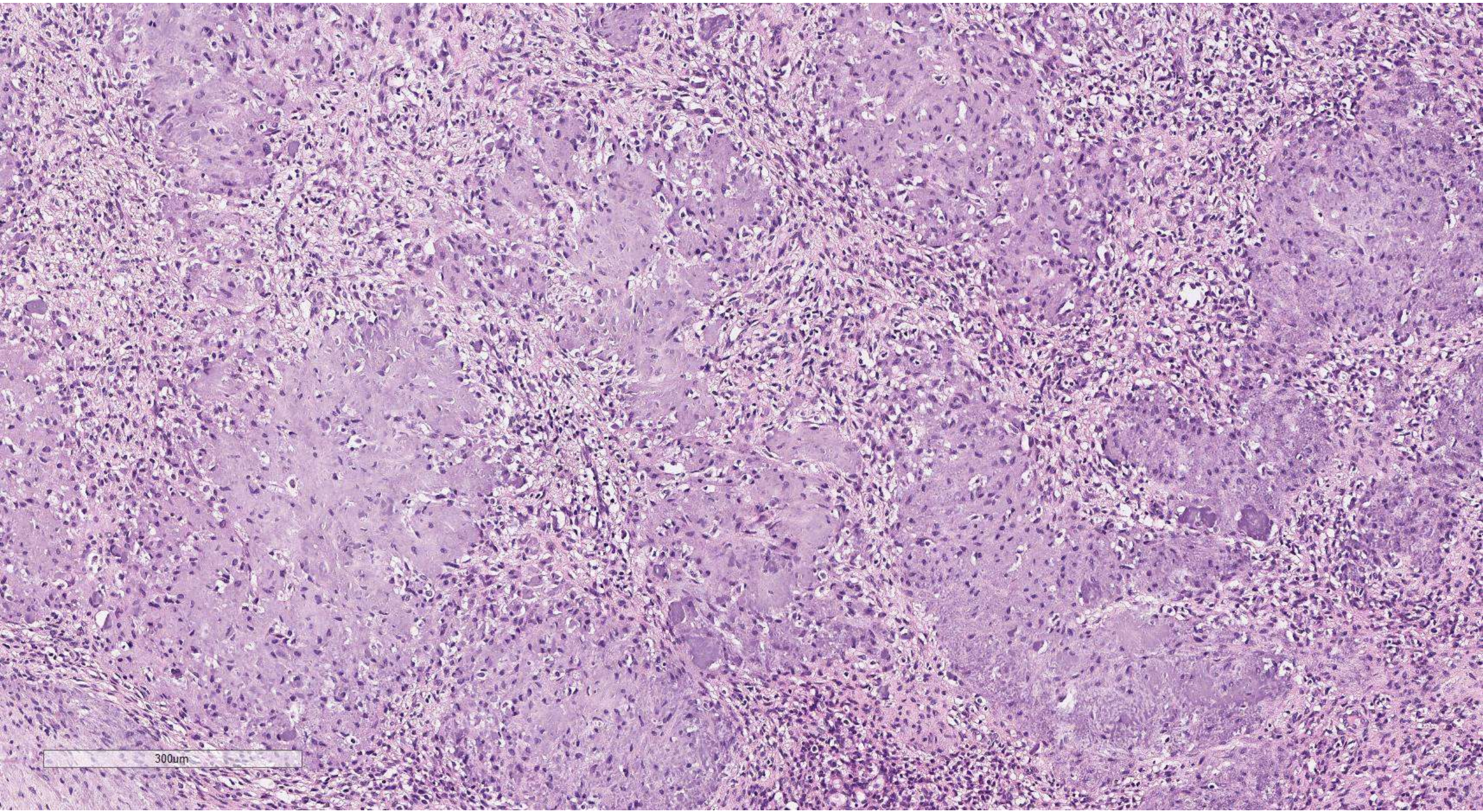
případ č.1

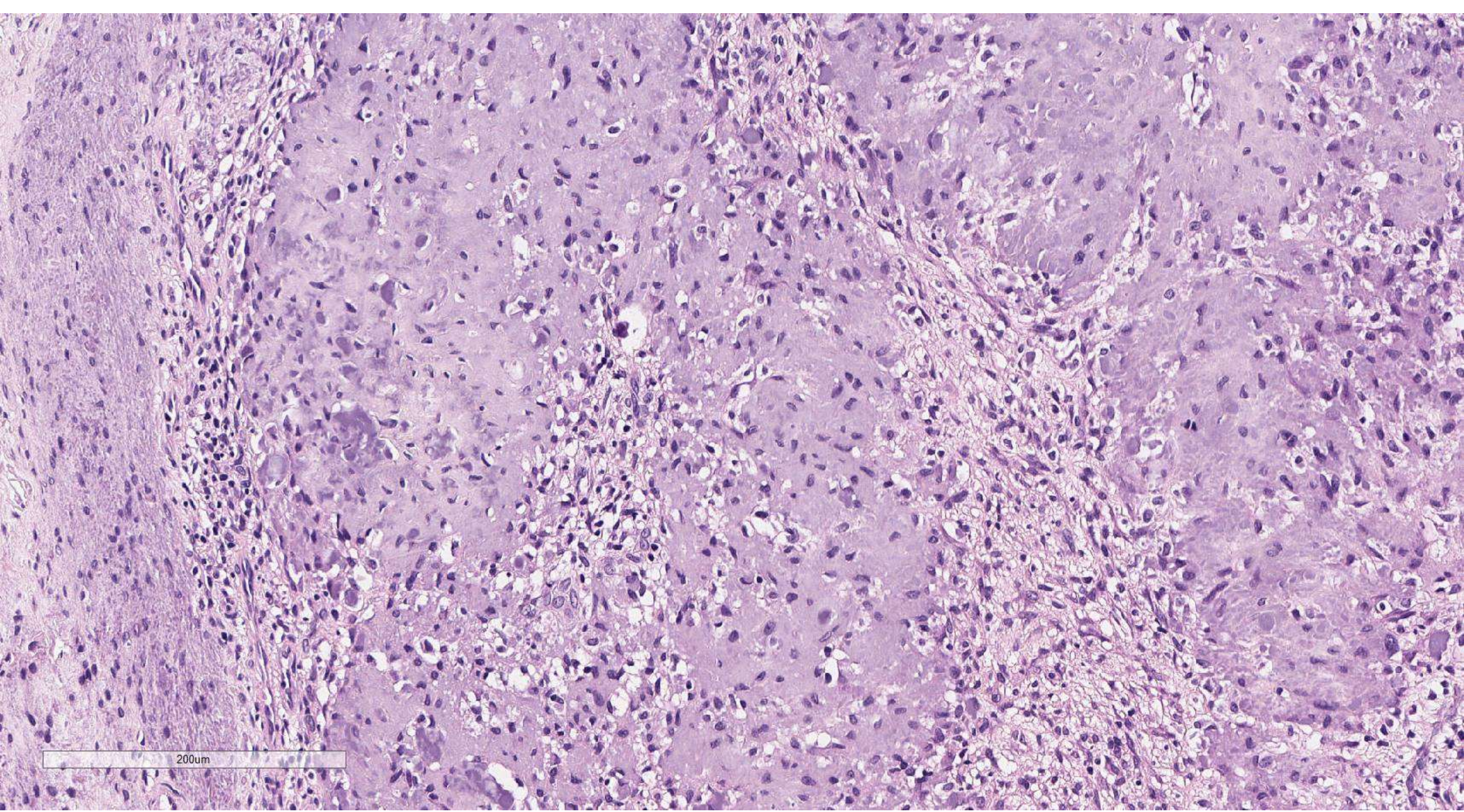
Klinické údaje a makropopis

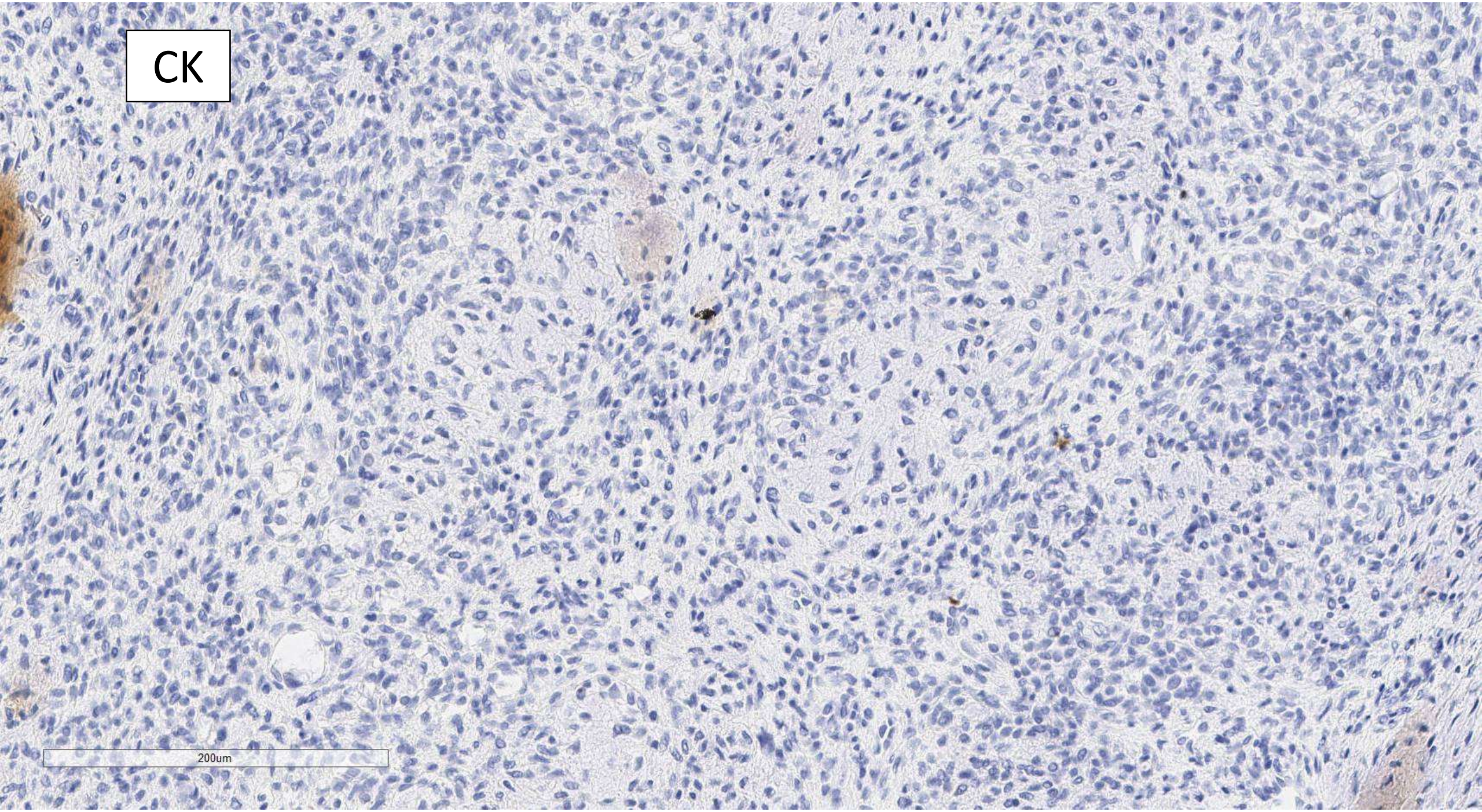
- žena, 37 let
- tumor v podkoží plosky levé nohy
- potrhaná, tukově vazivová částice 35 x 15 x 15 mm, na řezu s ložiskově prokrvácenýmtumorem šedé barvy, do průměru 15 mm, který zasahuje do potrhaných resekčních okrajů

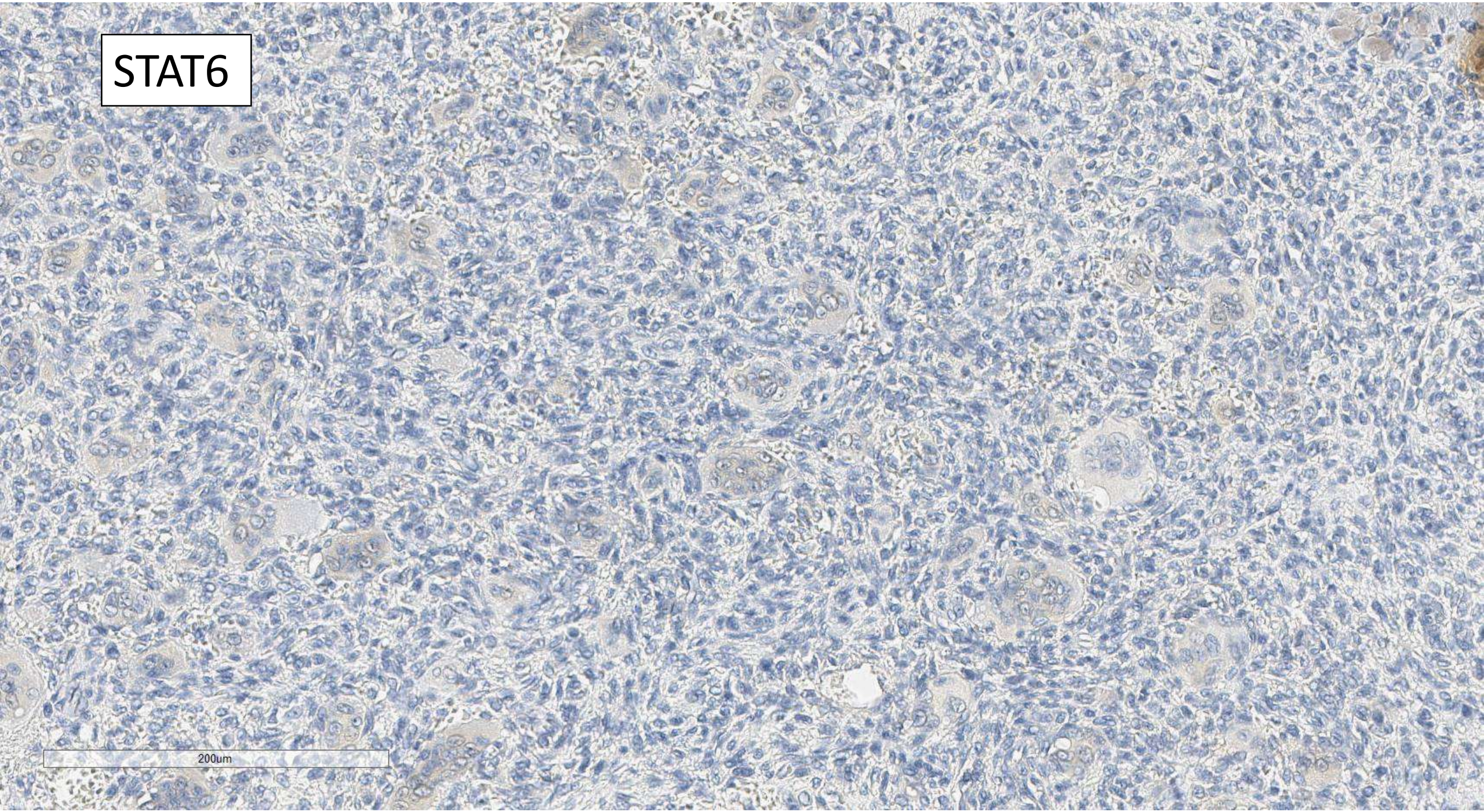

15
16
17
18

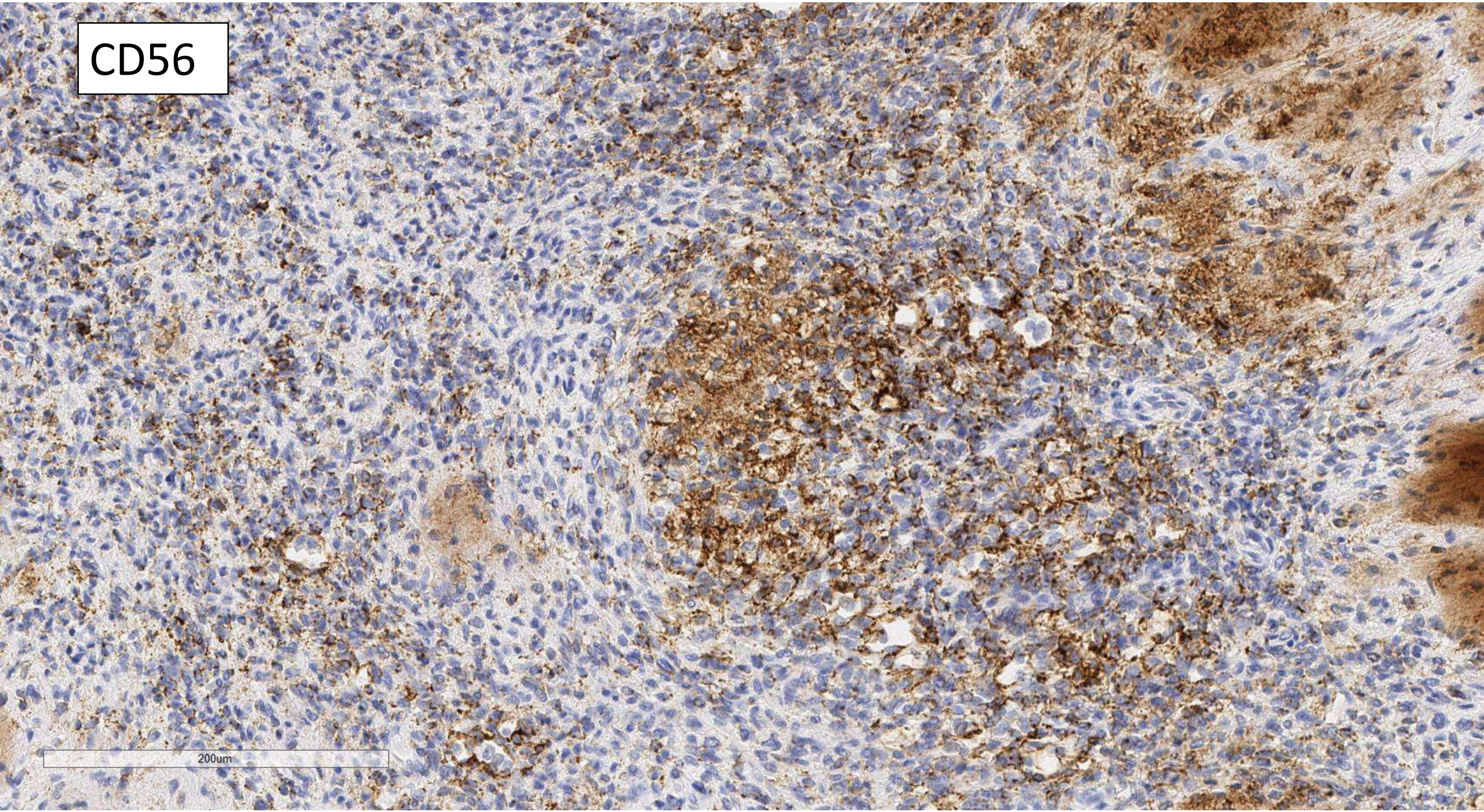


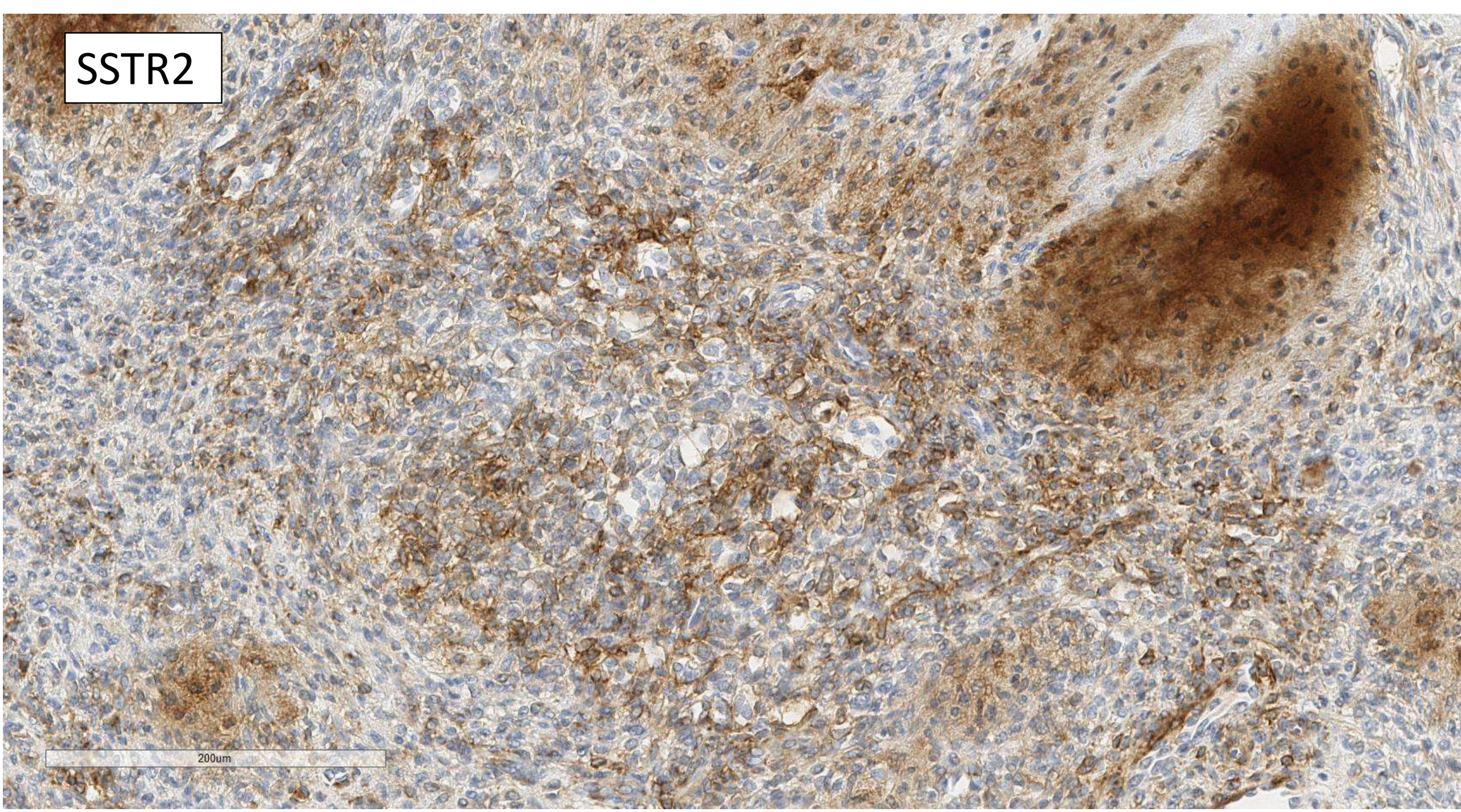

300um

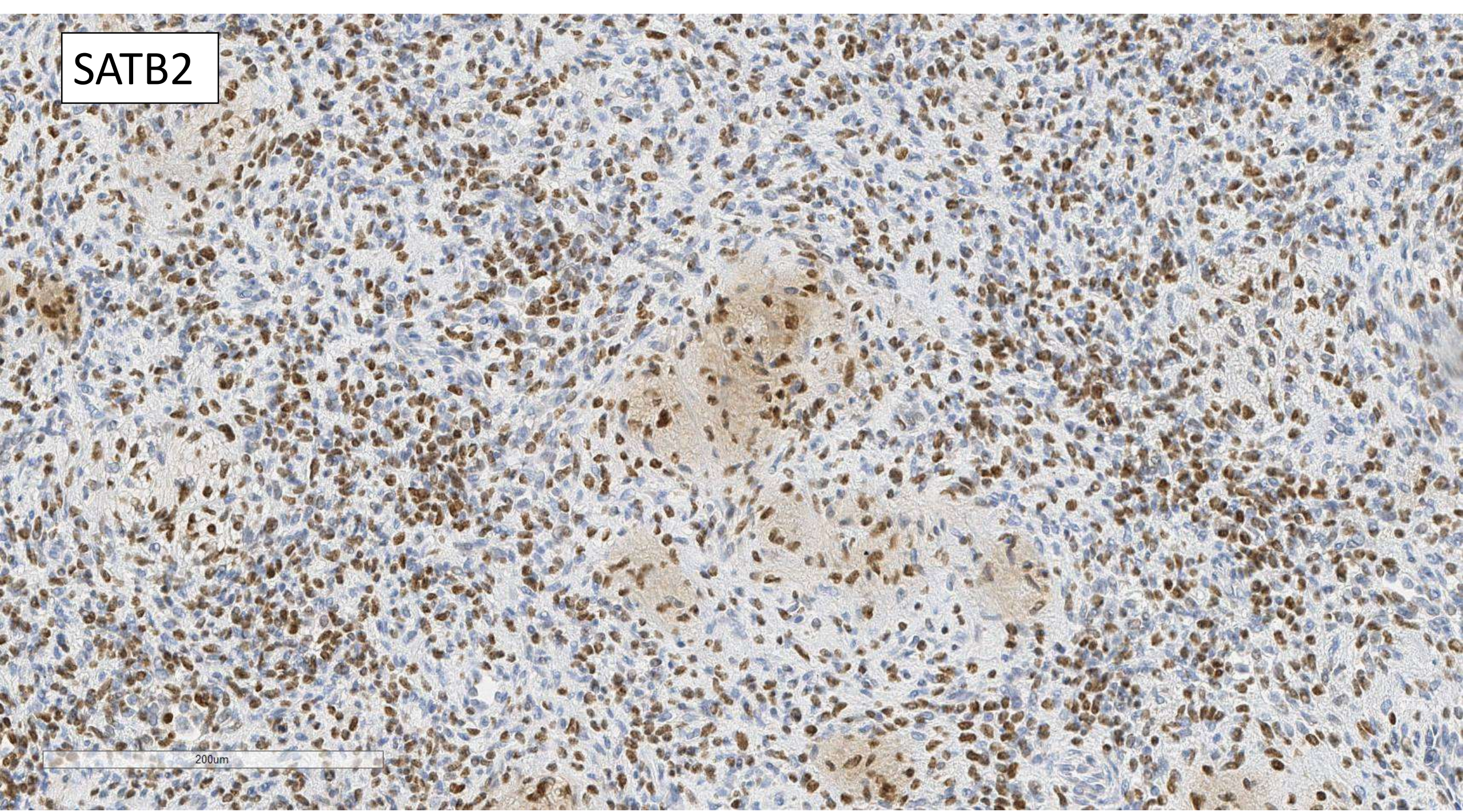

200um


CK
200um


STAT6
200um


CD56
200um


SSTR2
200um


SATB2
200um

případ č.1

Diagnóza

Fosfaturický mezenchymální tumor

případ č.1

Diskuze

Seminars in Diagnostic Pathology
Volume 36, Issue 4, July 2019, Pages 260-268

Review article

Phosphaturic mesenchymal tumors: A review and update

Andrew L. Folpe ✉

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› Am J Surg Pathol. 2017 Oct;41(10):1371-1380. doi: 10.1097/PAS.0000000000000890.

Phosphaturic Mesenchymal Tumors: Clinicopathologic, Immunohistochemical and Molecular Analysis of 22 Cases Expanding their Morphologic and Immunophenotypic Spectrum

Abbas Agaimy [1], Michael Michal, Simion Chiosea, Fredrik Petersson, Ladislav Hadravsky, Glenn Kristiansen, Raymund E Horch, Jan Schmolders, Arndt Hartmann, Florian Haller, Michal Michal

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PMID: 28614212 DOI: 10.1097/PAS.0000000000000890

případ č.1

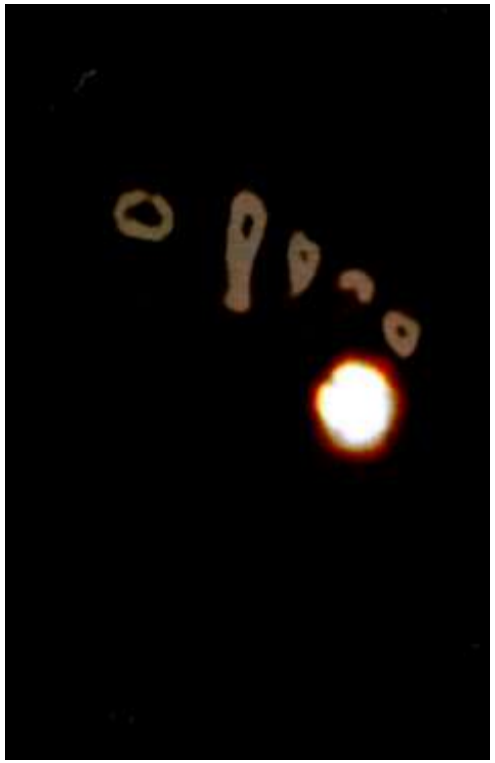
Diskuze

- 1957 Prader, 1972 Evans, Azzopardi, 1987 Weidner, Santa Cruz
- 450 případů
- střední věk, kdekoliv (1/2 případů na končetinách/akrálně)
- nespecific. příznaky (svalová bolest, progresivní slabost) – 3 roky k dg. TIO a 5 let k resekci tumoru
- „non-fosfaturická“ varianta PMT, maligní PMT
- TIO vzácně i u jiných tumorů (NF1, McCune Albright sy, SCLC, CRC, RCC, Ca prostaty, anaplastický Ca ŠŽ, HG serózní Ca tuby/ovária)

případ č.1

Diskuze

- PET/CT



- FGF23 (osteoblasty) - ↓ reabsorbce fosfátů v prox. tubulech (vzácně SFR4, FGF7)

případ č.1

Diskuze

TABLE 2. Immunohistochemical and Molecular Findings in Phosphaturic Mesenchymal Tumors (n = 22)

No.	SSTR2A	SATB2	ERG	CD56	S100	CD34	DOG1	STAT6	Beta-catenin	*FGFR1* FISH
1	+	+++	+	NA	NA	NA	NA	NA	–	–*
2	+	–	+	NA	NA	NA	NA	NA	–	–*
3	–	+++	–	NA	NA	NA	–	NA	NA	+
4	+	+++	+	NA	–	NA	–	NA	NA	+
5	+(F)	–	+	NA	–	NA	–	NA	NA	–
6	+	++	–	NA	NA	NA	NA	NA	–	NA
7	NA	NA	NA	NA	NA	NA	NA	NA	–	NA
8	+	+++	+(F)	NA	NA	NA	–	NA	NA	–*
9	NA	+	+	NA	NA	NA	–	NA	NA	NR
10	–	+++	+	NA	–	NA	NA	NA	NA	+
11	+	+++	+	+	–	+	–	–	–	–*
12	+	+++	+	+	–	–	–	–	NA	–*
13	+	+++	+	+	–	–	–	NA	–	–*
14	+	+++	+	+	–	–	–	–	–	–
15	+	+++	+	+	–	–	–	–	–	+
16	–	+	+	+(F)	–	+	–	–	–	NR
17	+(wk)	+++	+	+	–	–	–	–	–	+
18	+(wk)	+++	+(F)	NA	–	NA	–	NA	–	+
19	NR	++	+	+	–	–	–	NA	NA	+
20	–	++	+	+	NA	–	–	NA	–	NR
21	+	+++	+	+	–	–	–	NA	–	–
22	+(wk)	+++	+	+	–	–	–	–	–	+

*Limited assessability due to poor signal quality.

F indicates focal; NA, not available; NR, no results due to poor tissue preservation; wk, weak; –, negative; +, positive in < 25% tumor cells; ++, positive in 26–50% tumor cells; +++, positive in > 50% tumor cells.

- IHC: SATB2, SSTR2, CD56, ERG
- ISH: *FN1::FGFR1 (FGF1)* (FISH 60 – 70 %)

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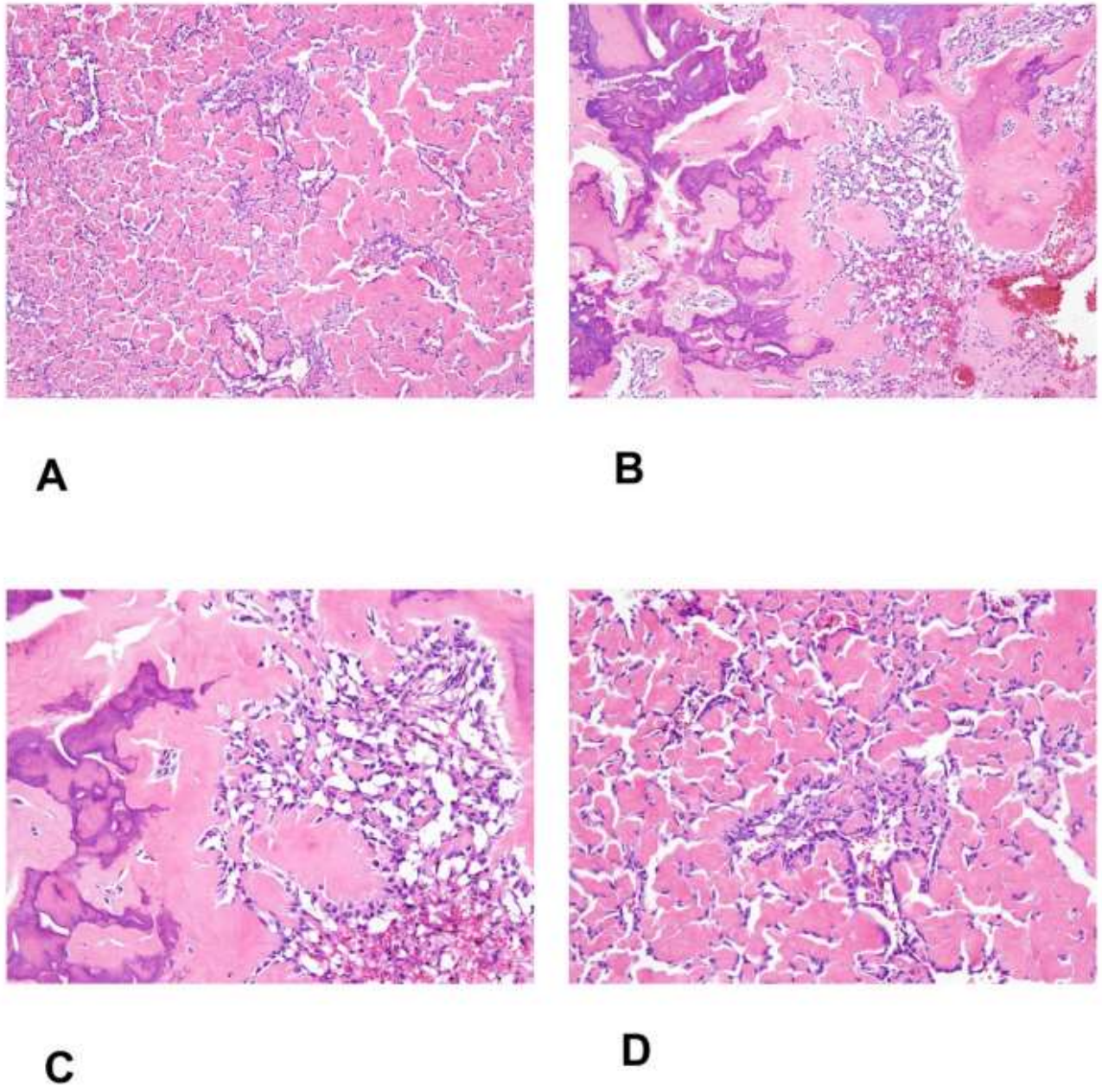


Fig. 6. "Osteoblastoma-like" phosphaturic mesenchymal tumor from the femur

Andrew L. Folpe, Phosphaturic mesenchymal tumors: A review and update, Seminars in Diagnostic Pathology, Volume 36, Issue 4, 2019, Pages 260-268, ISSN 0740-2570, https://doi.org/10.1053/j.semdp.2019.07.002.

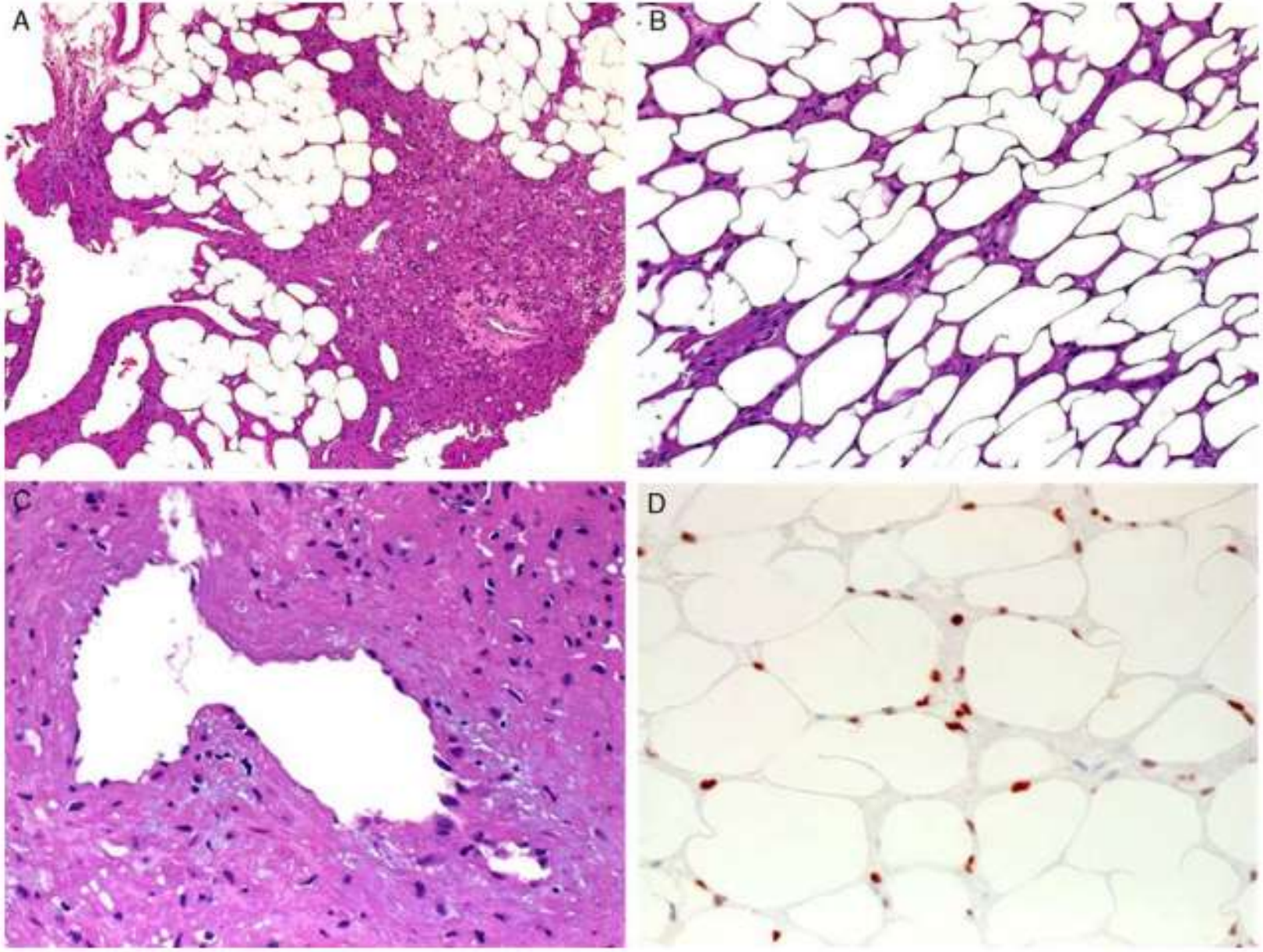
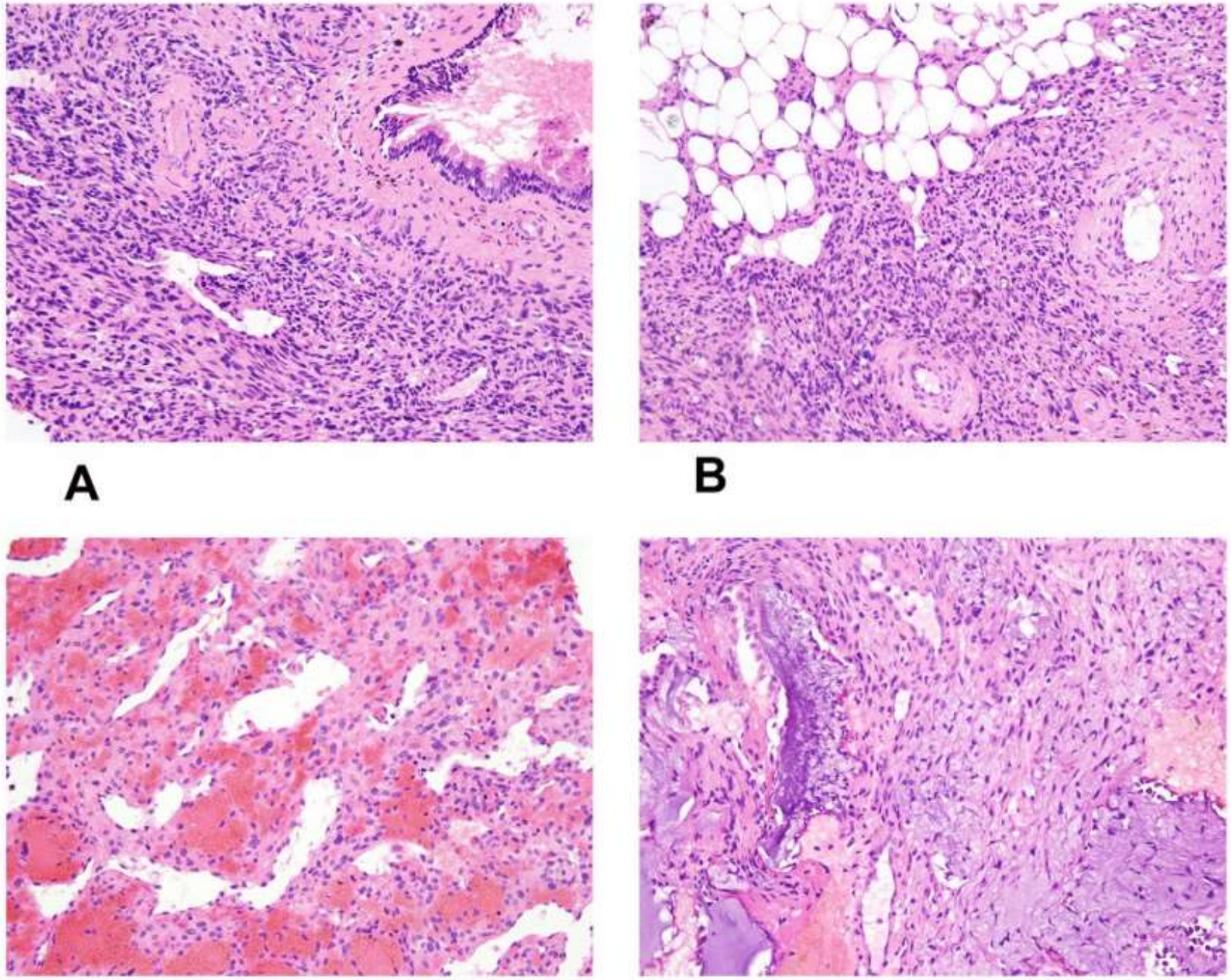


FIGURE 3. A, This angiomyolipoma-like variant PMT contained prominent fatty component imparting biphasic pattern, note prominent thick-walled vessels. B, Tumor cells were intermingled between adipocytic cells. C, thick-walled veins embedded within sclerotic matrix. D, The scattered neoplastic cells are highlighted by SATB2 immunostaining.

Agaimy A, Michal M, Chiosea S, Petersson F, Hadravsky L, Kristiansen G, Horch RE, Schmolders J, Hartmann A, Haller F, Michal M. Phosphaturic Mesenchymal Tumors: Clinicopathologic, Immunohistochemical and Molecular Analysis of 22 Cases Expanding their Morphologic and Immunophenotypic Spectrum. Am J Surg Pathol. 2017 Oct;41(10):1371-1380. doi: 10.1097/PAS.0000000000000890. PMID: 28614212.



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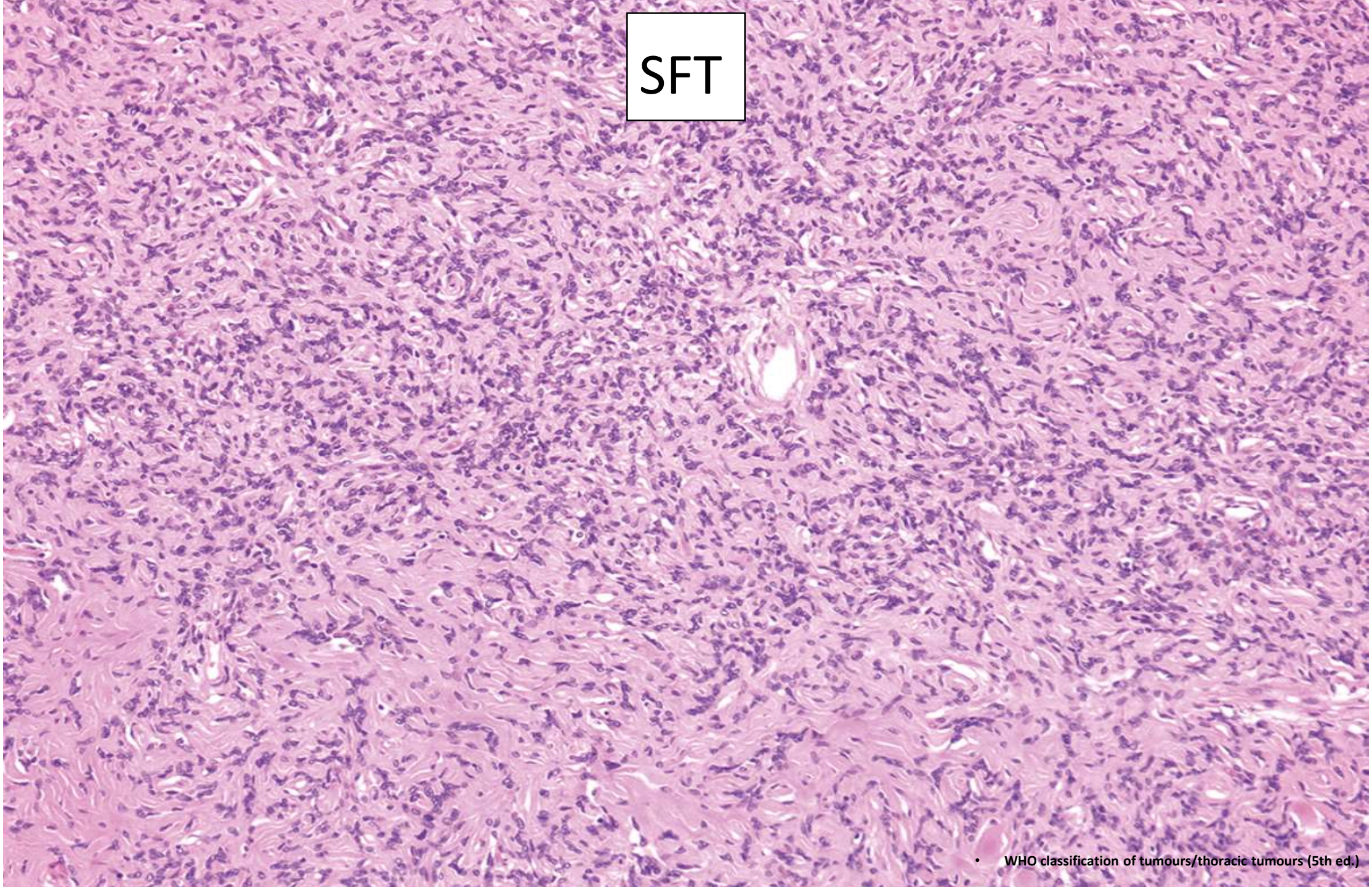
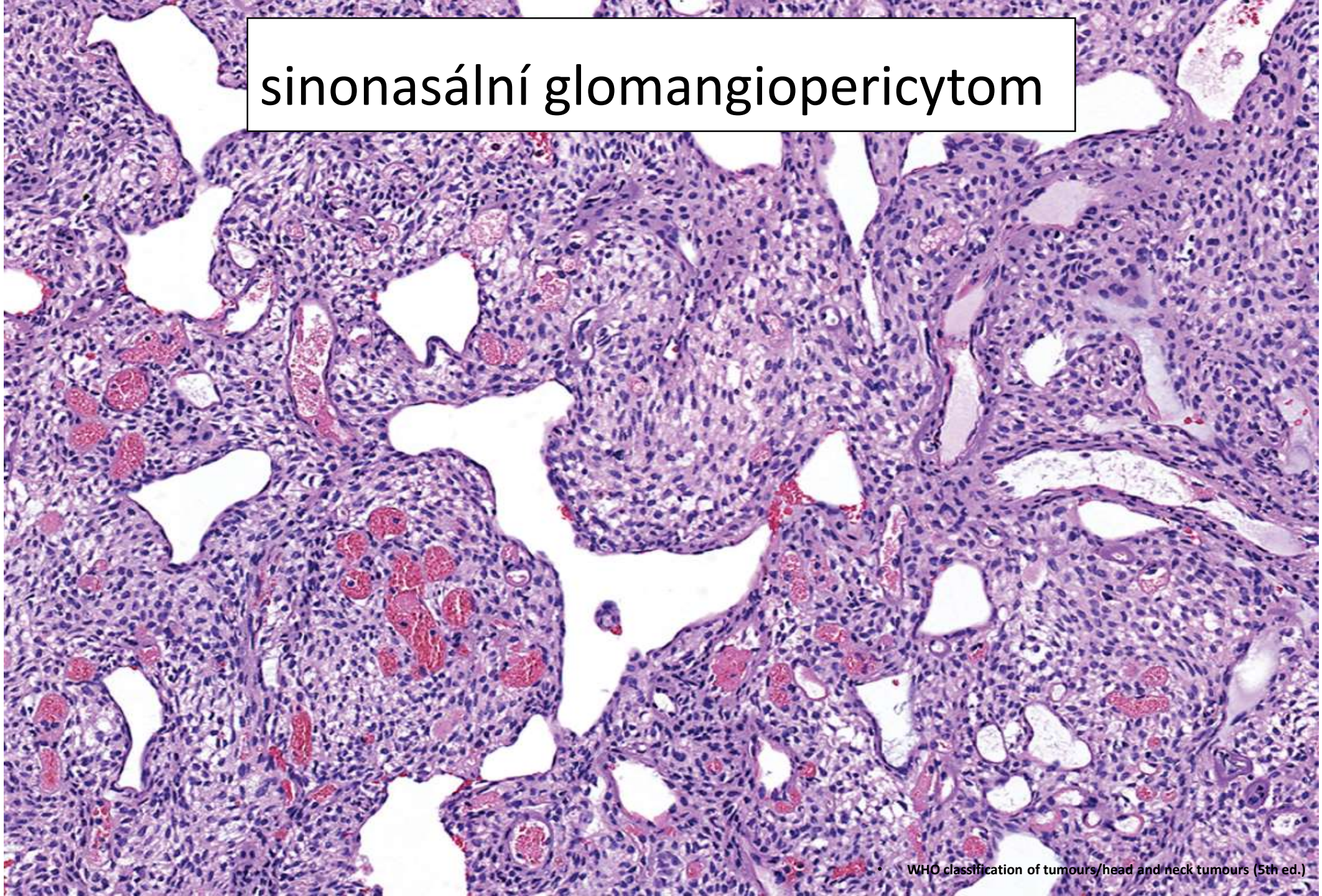
případ č.1

Dif Dg



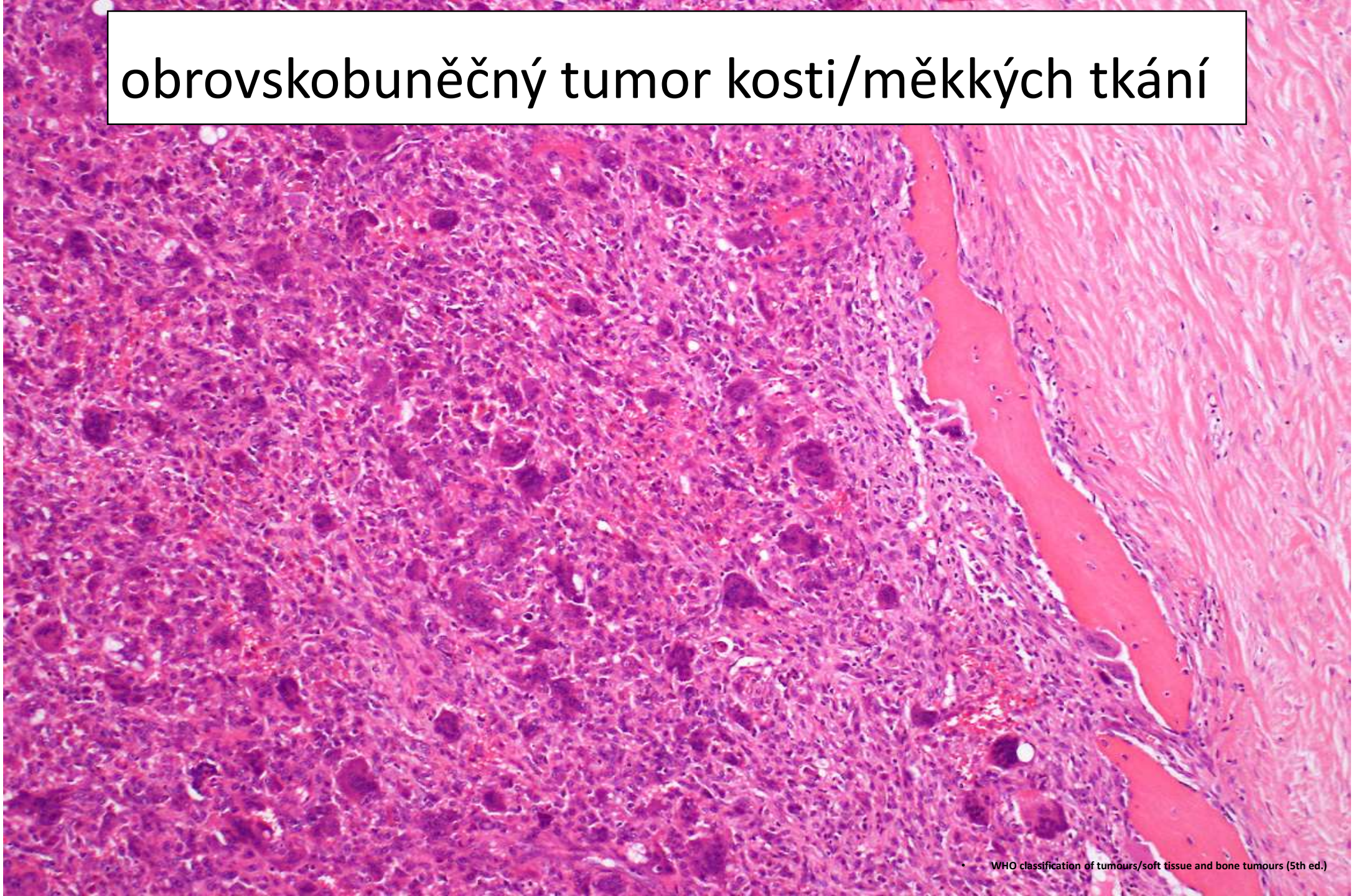
- WHO classification of tumours/thoracic tumours (5th ed.)

sinonasální glomangiopericytom



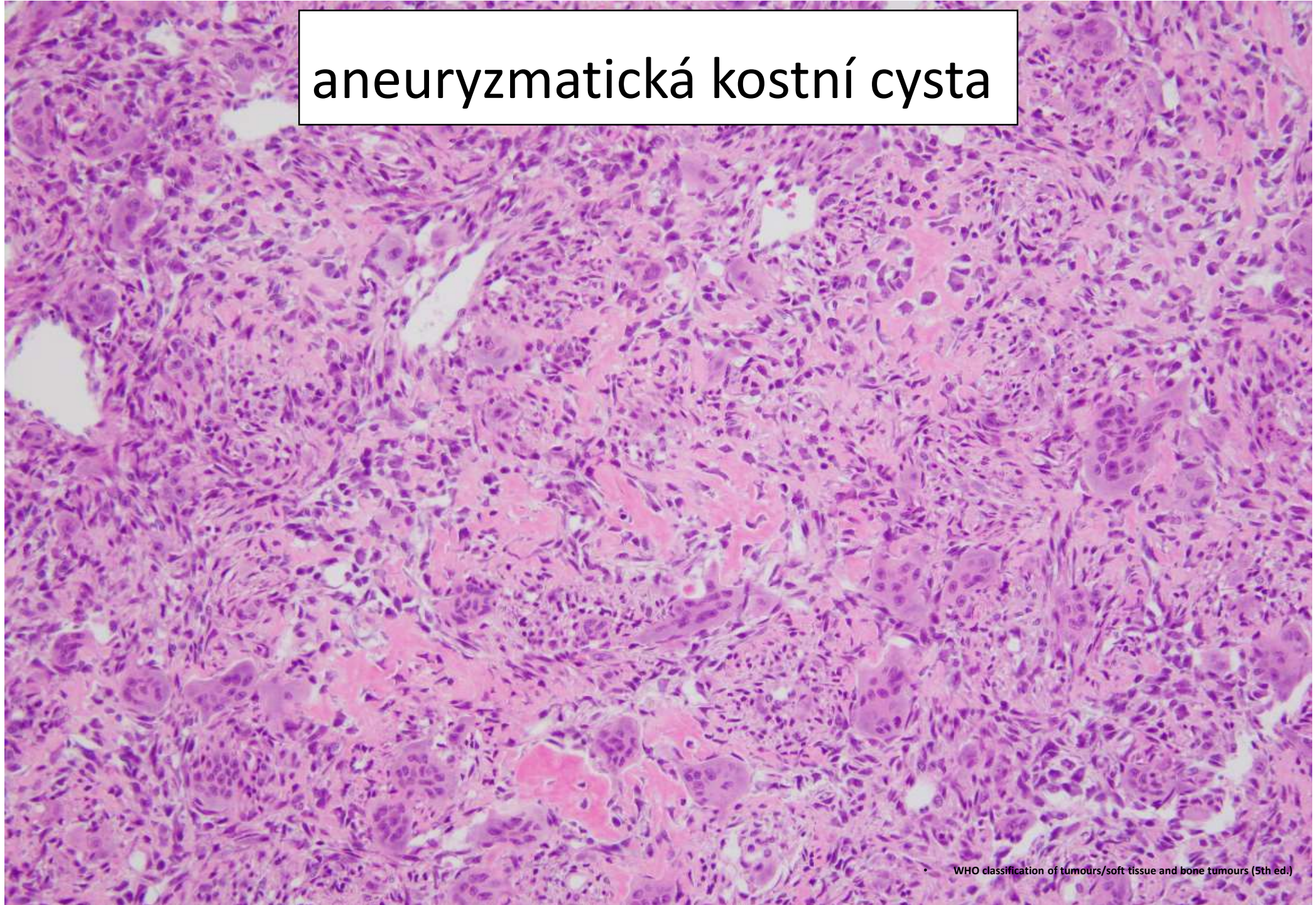
- WHO classification of tumours/head and neck tumours (5th ed.)

obrovskobuněčný tumor kosti/měkkých tkání



- WHO classification of tumours/soft tissue and bone tumours (5th ed.)

aneuryzmatická kostní cysta



- WHO classification of tumours/soft tissue and bone tumours (5th ed.)

případ č.1

Dif Dg

- sinonasální glomangiopericytom
- sinonasální angiofibrom
- obrovskobuněčný tenosinoviální tumor (chondroidní)
- neosifikující fibrom
- chondromyxoidní fibrom
- chondrom měkkých částí
- mezenchymální chondrosarkom
- osteosarkom
- osteoblastom
- vřetenobuněčný lipom
- hemangiom/vaskulární malformace
- AML, NET, GIST