SD IAP 758

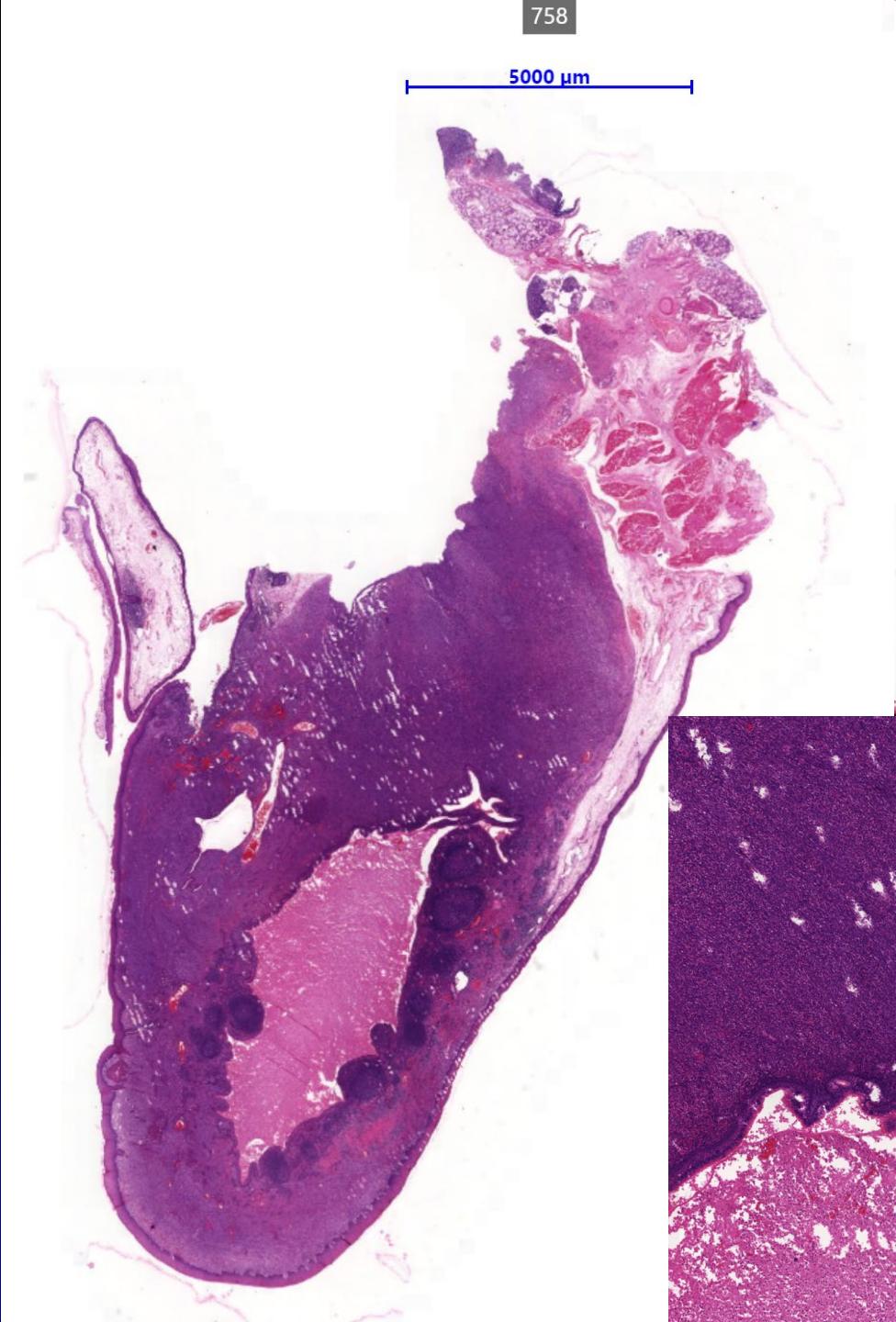
Peter SZÉPE

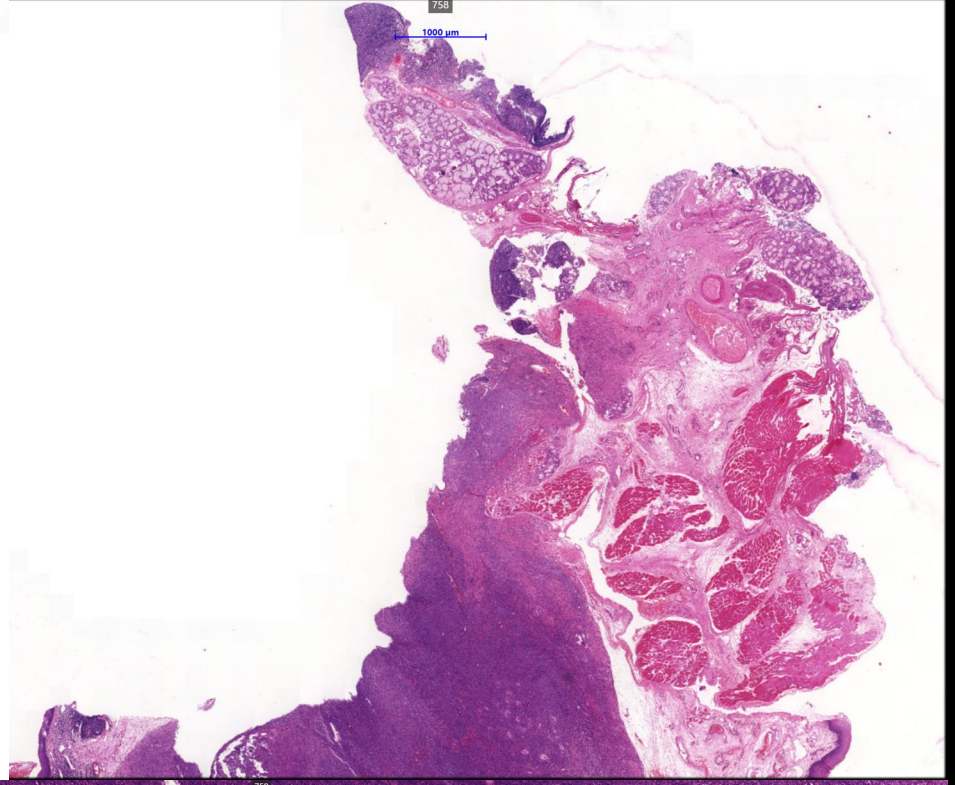
ÚPA JLF UK a UN Martin

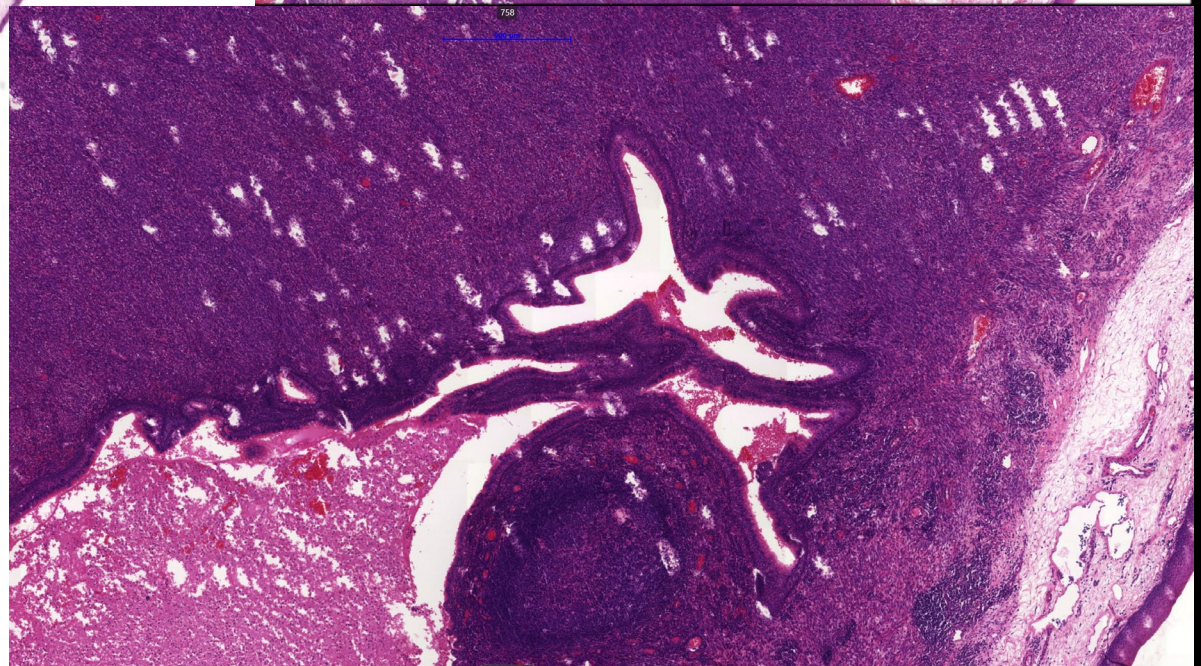
Letný bioptický seminár SD IAP, Senec, 17.-18.9 2021

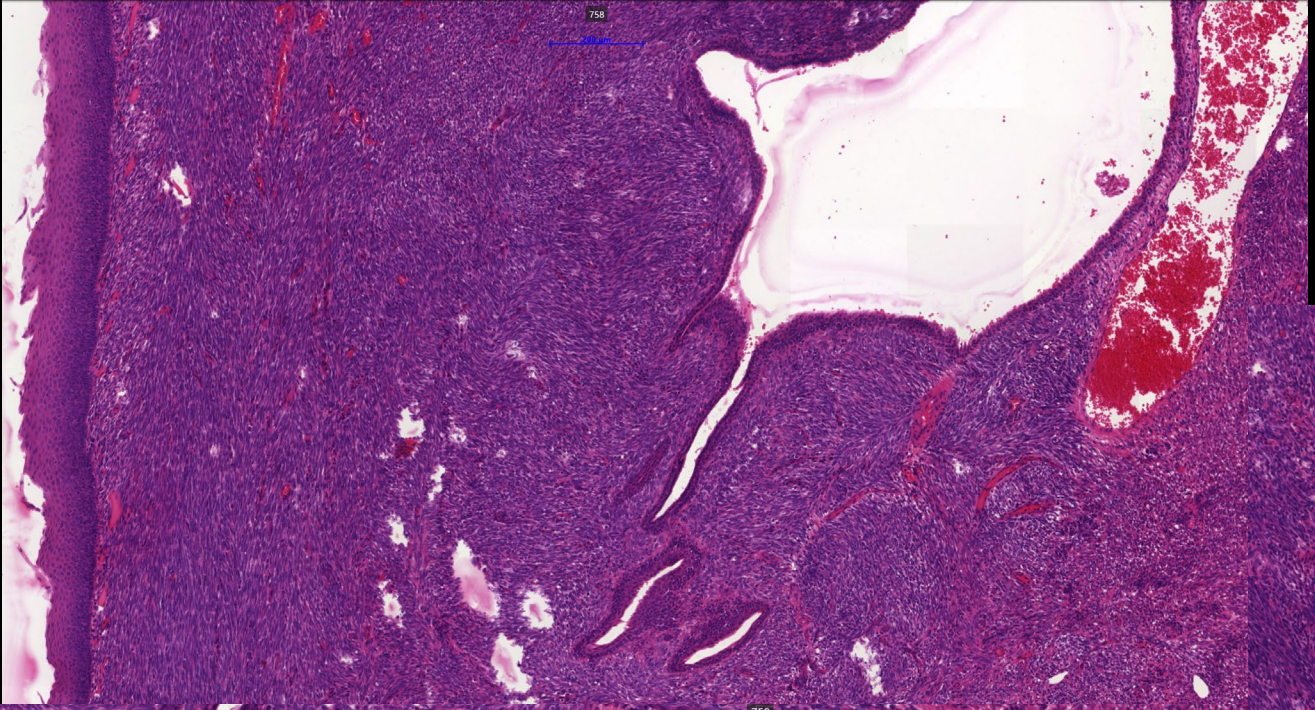
Klinické údaje

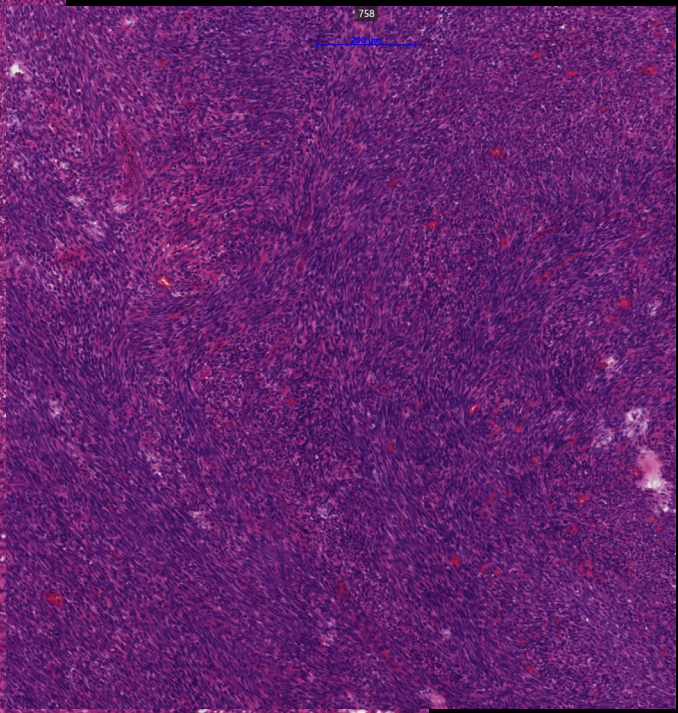
- 18-ročný muž, excízia z podnebného oblúka
- **Klinická dg.:** Susp. fibróm zadného podnebného oblúka vľavo, akcesórna tonzila ?
- Konzultácia z PAO ÚVN, RK

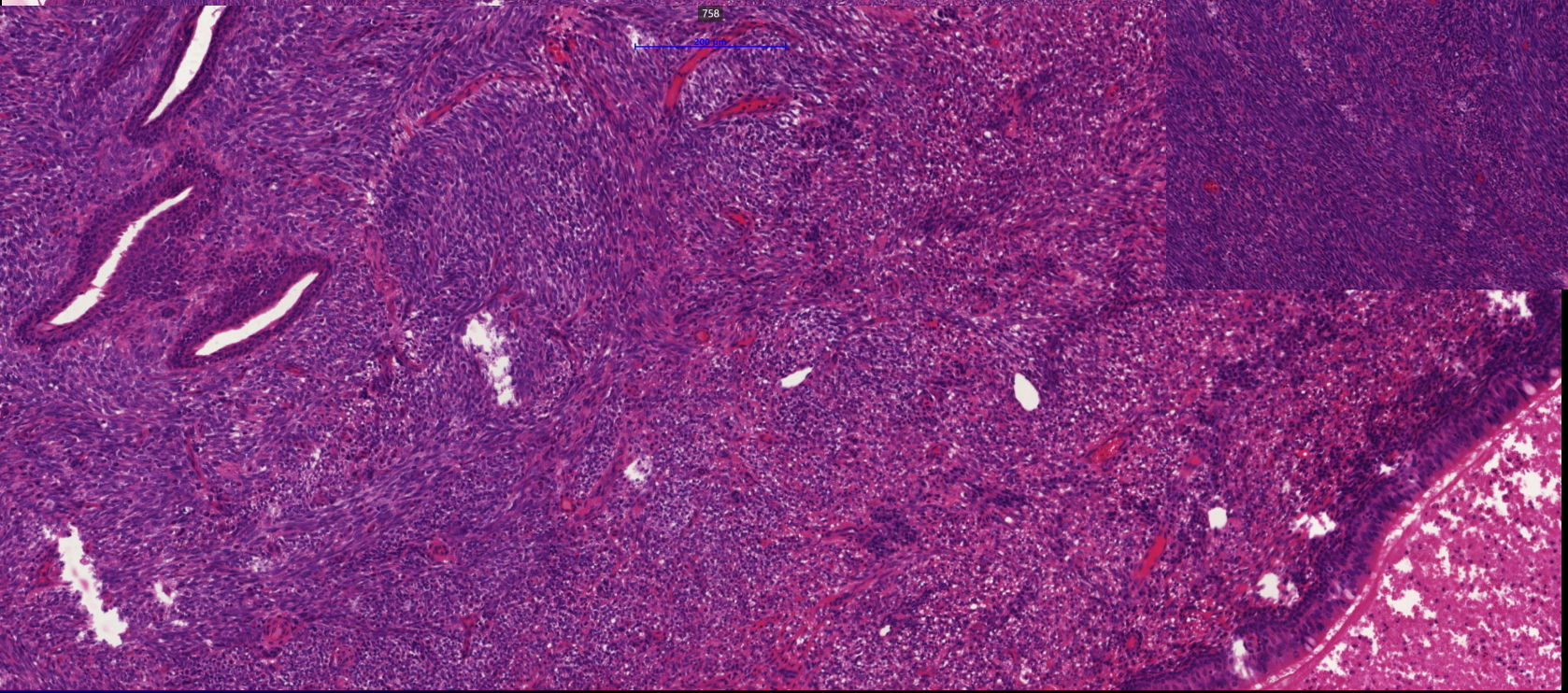

758
5000 µm

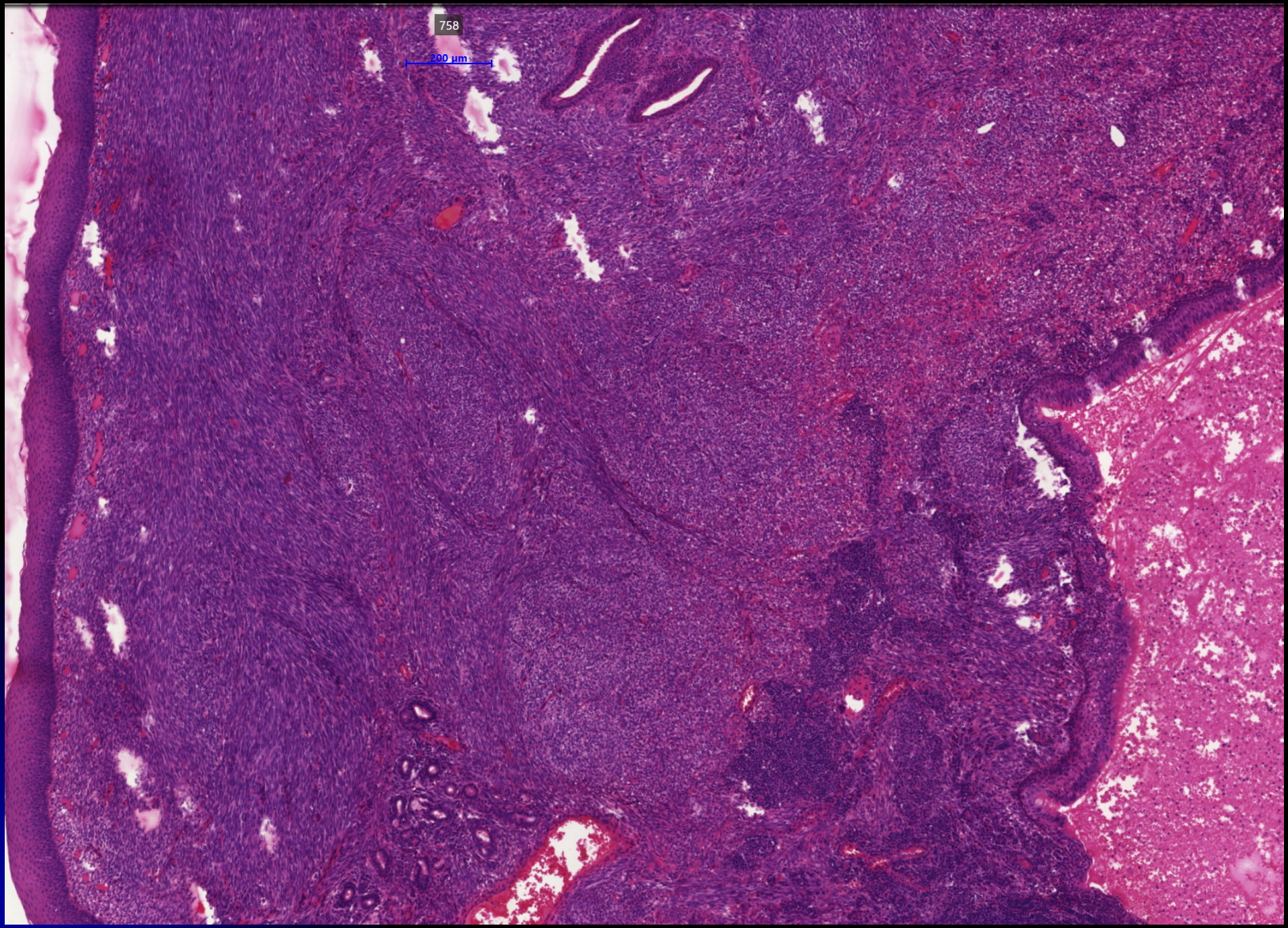

758
1000 µm

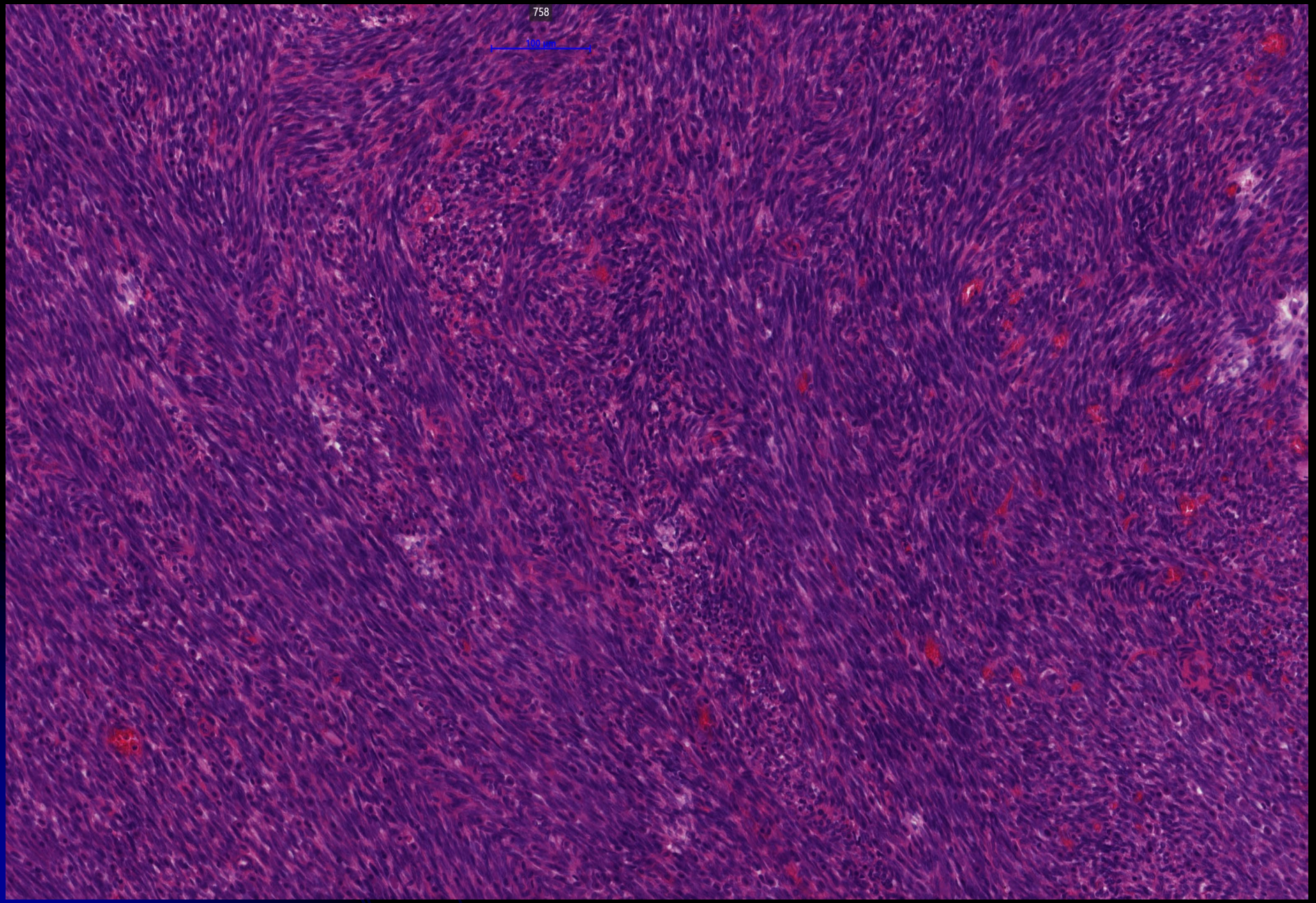

758

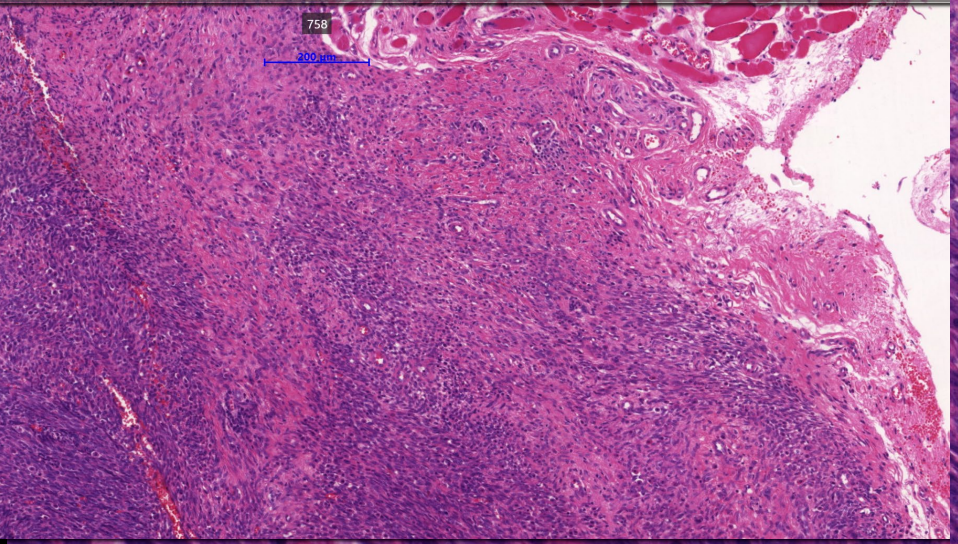

758

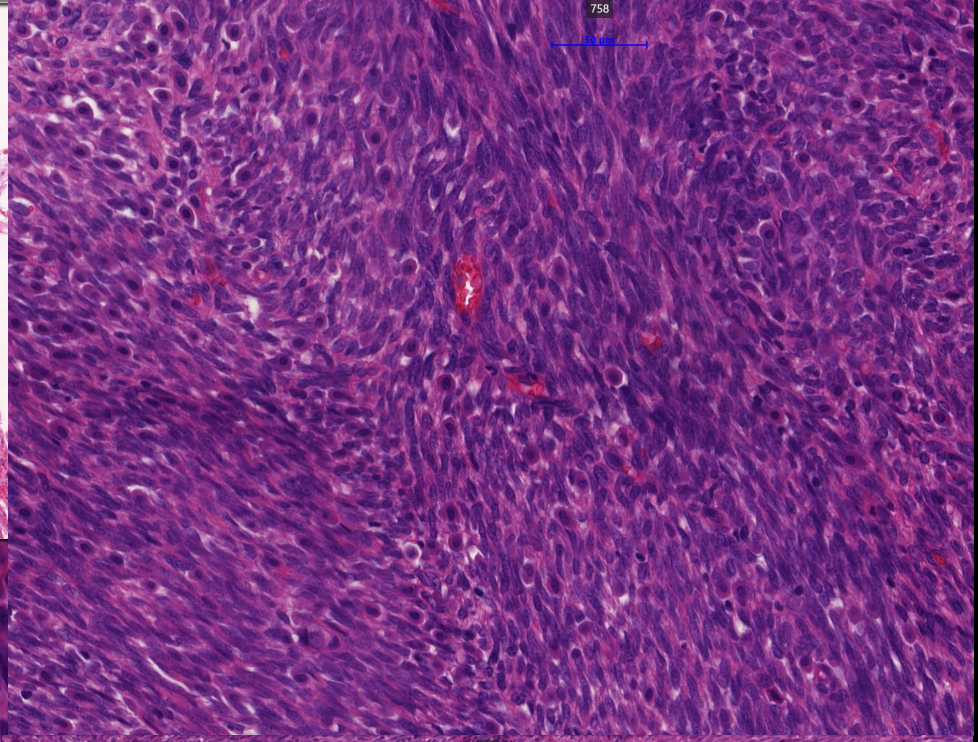

758

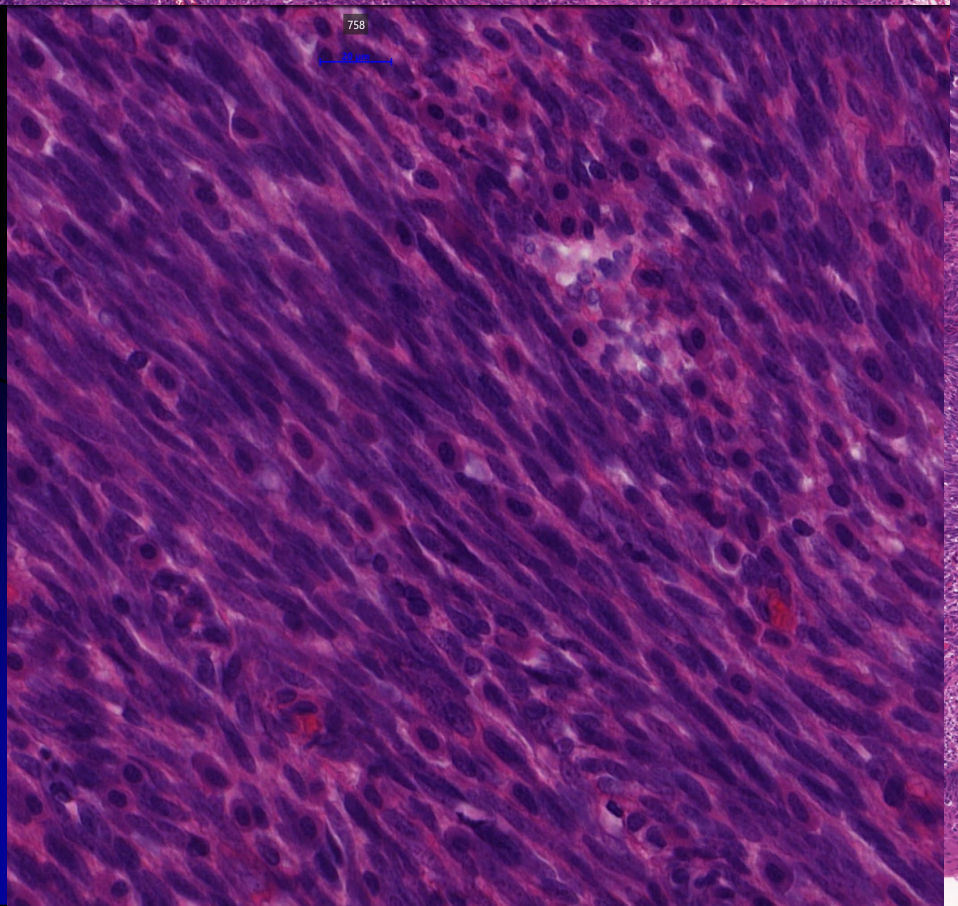

758

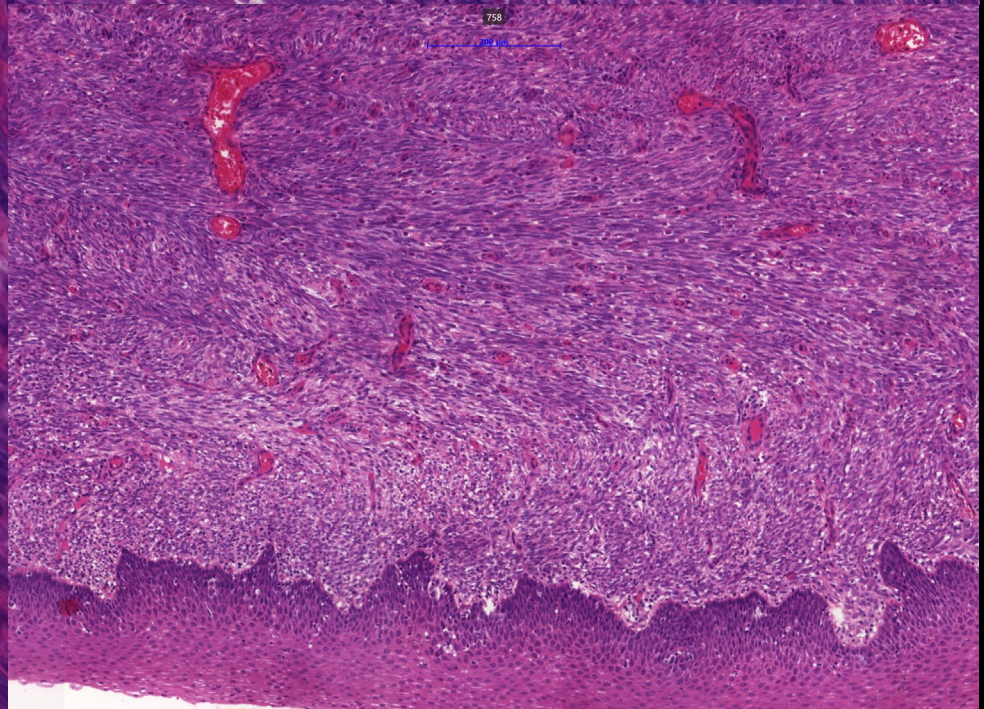

758
200 µm

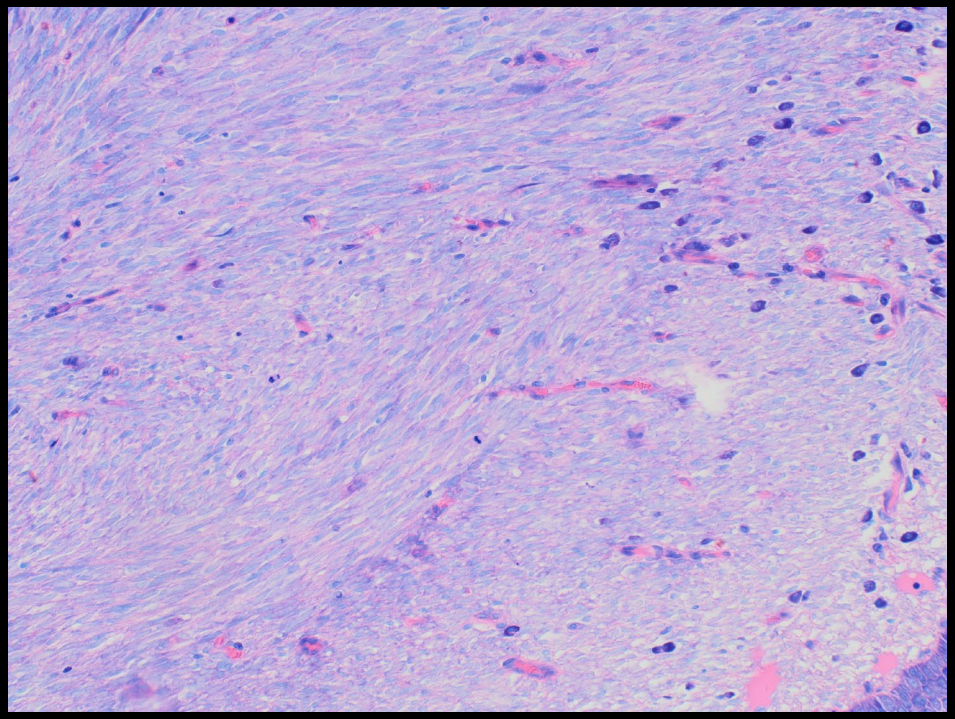
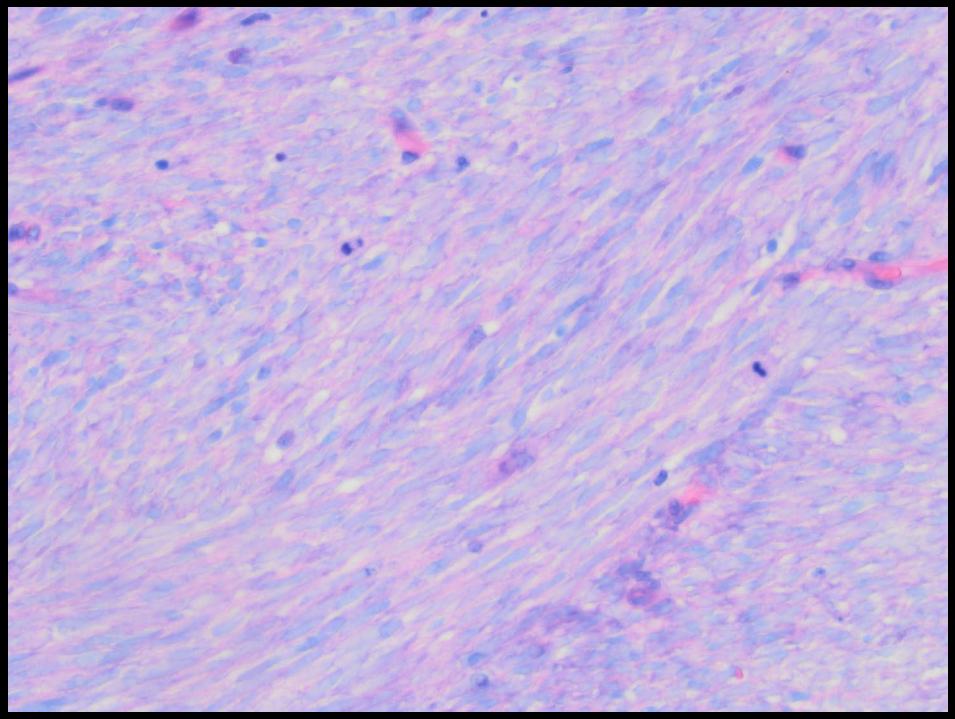
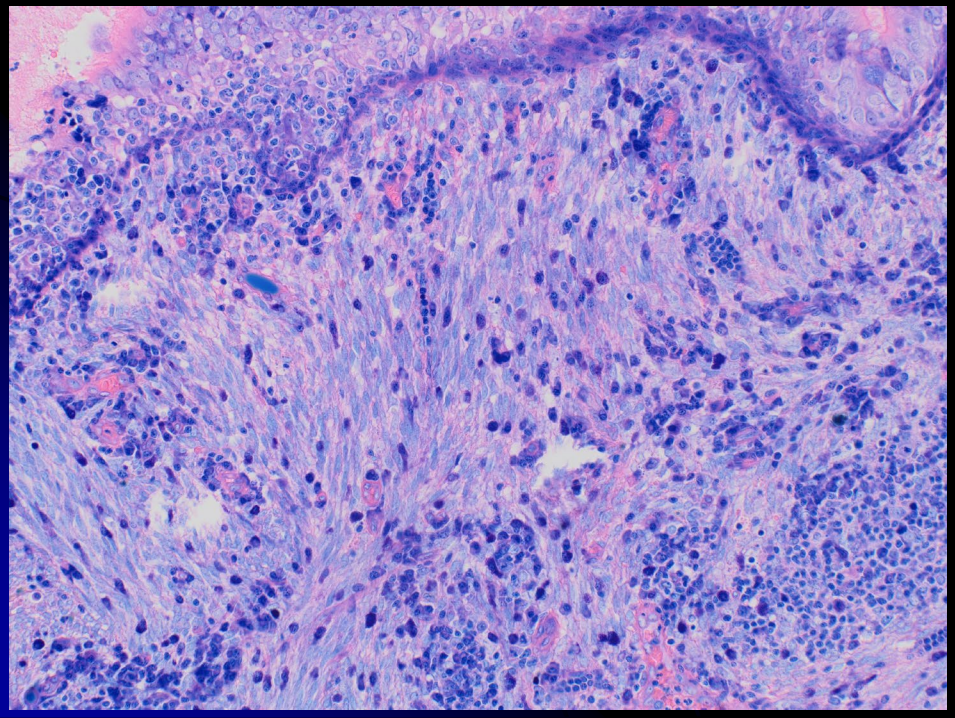
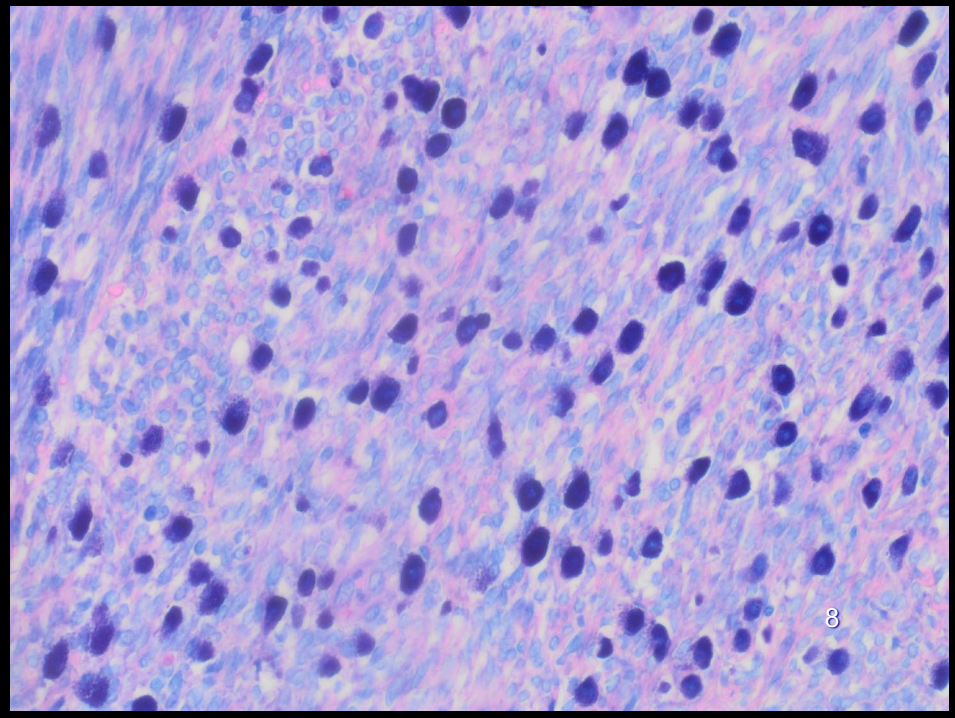

758
100 µm

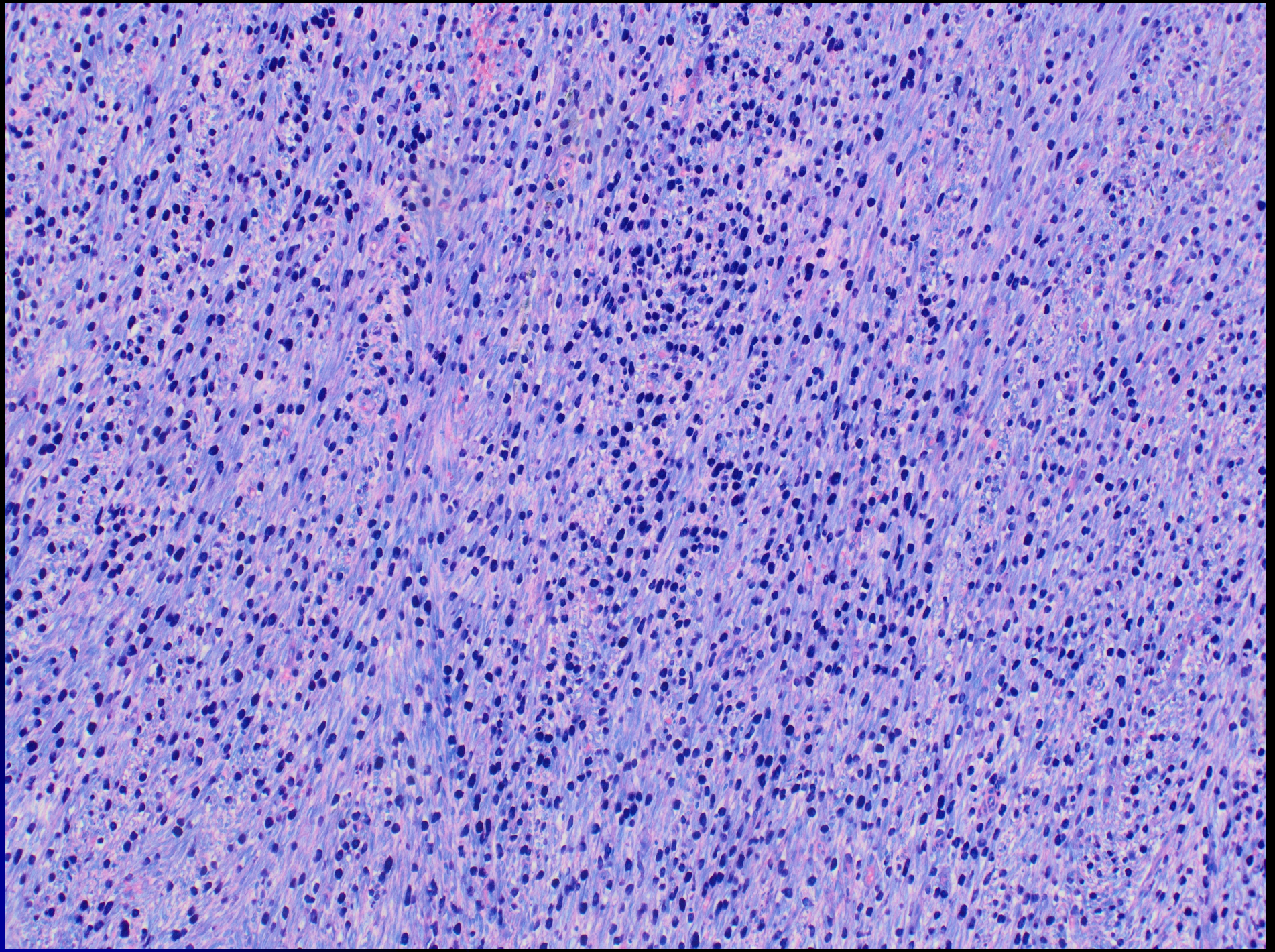

758
200 µm


758
50 µm


758
50 µm


758
200 µm

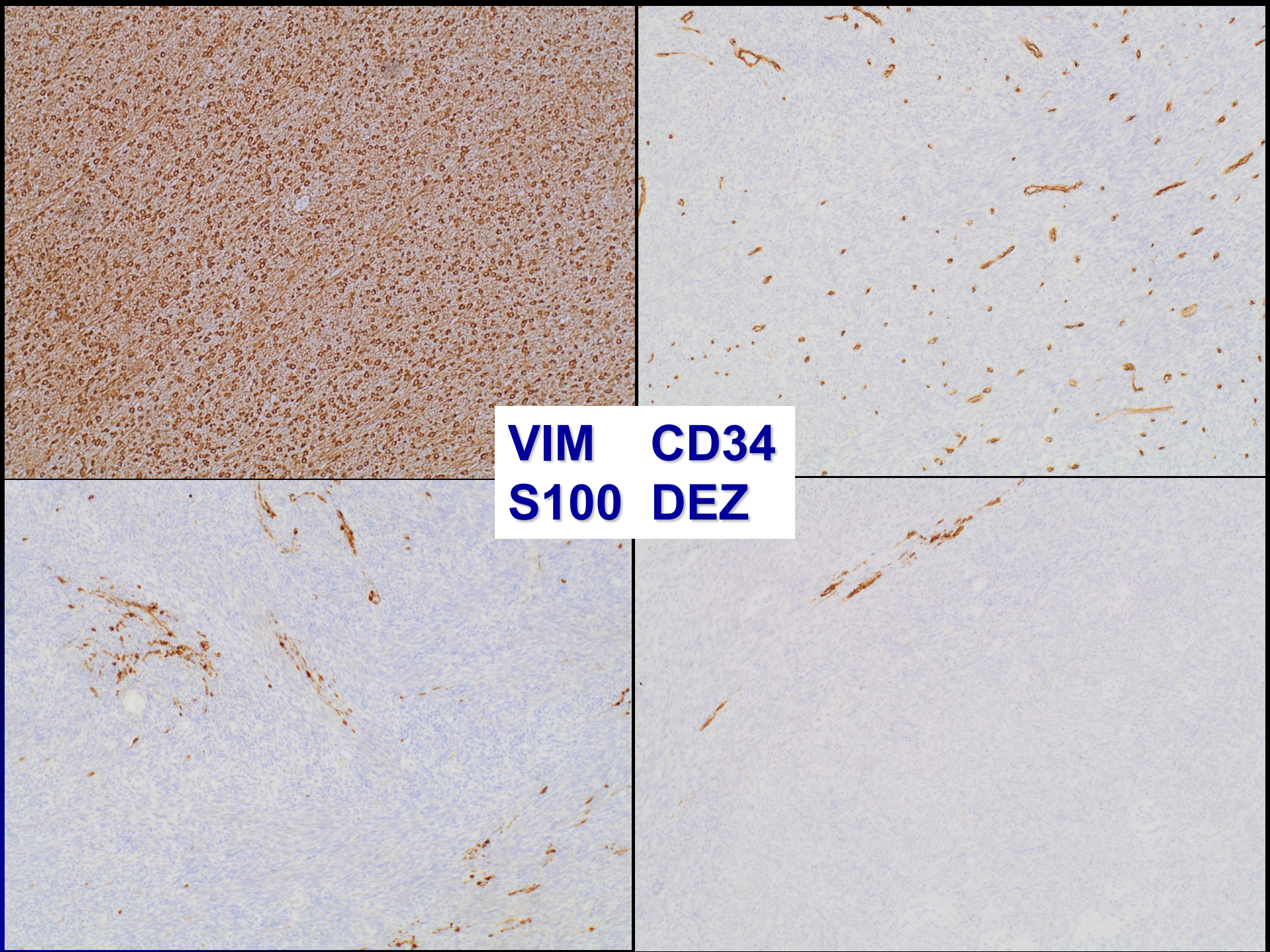





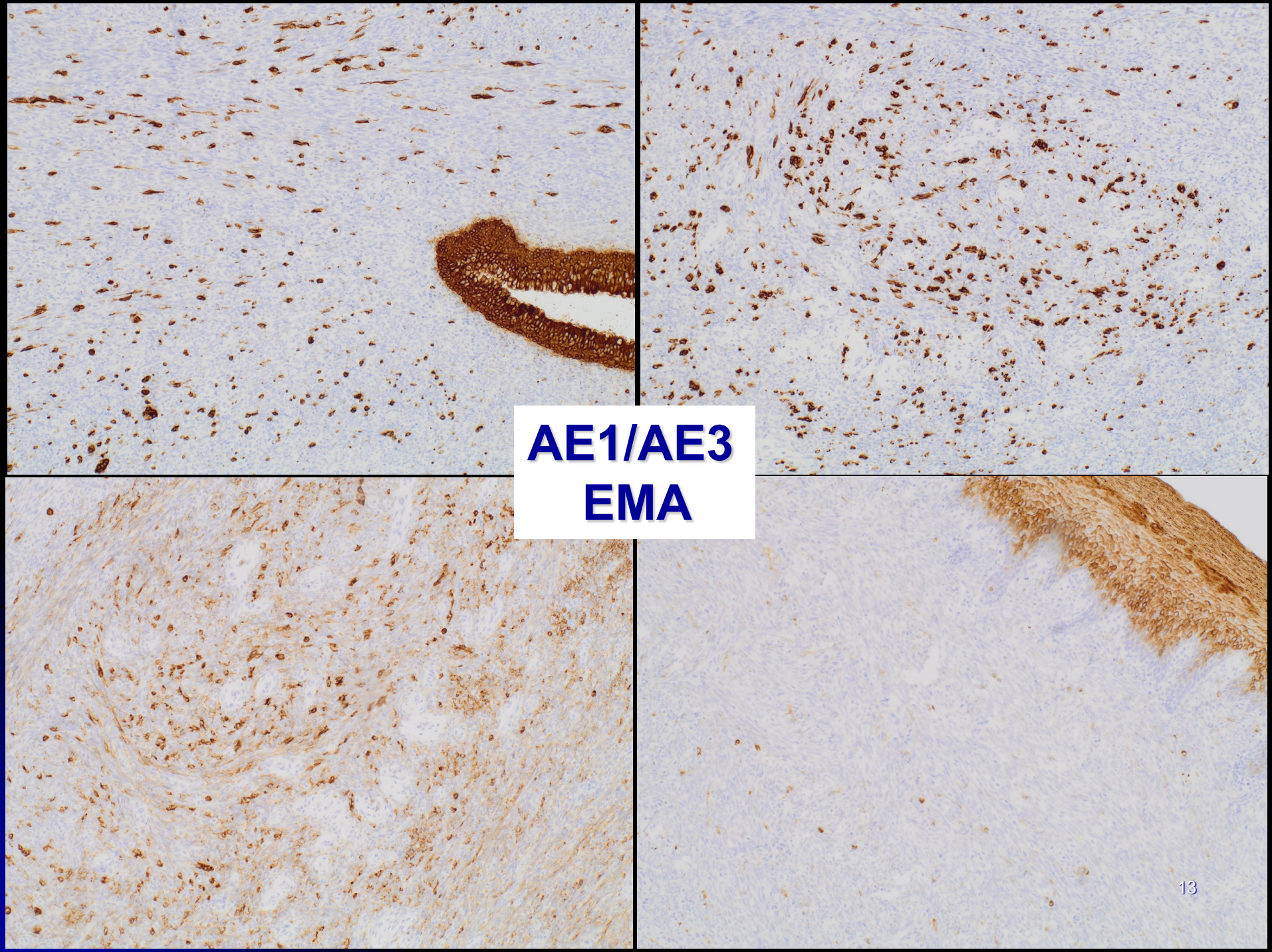


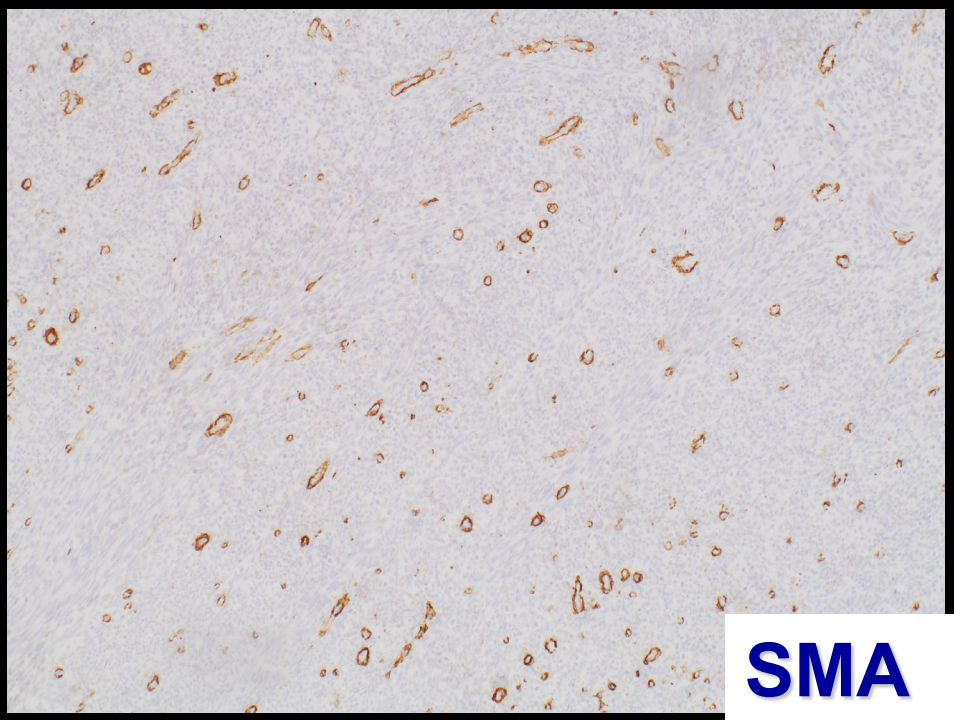
Sumarizácia morfológie

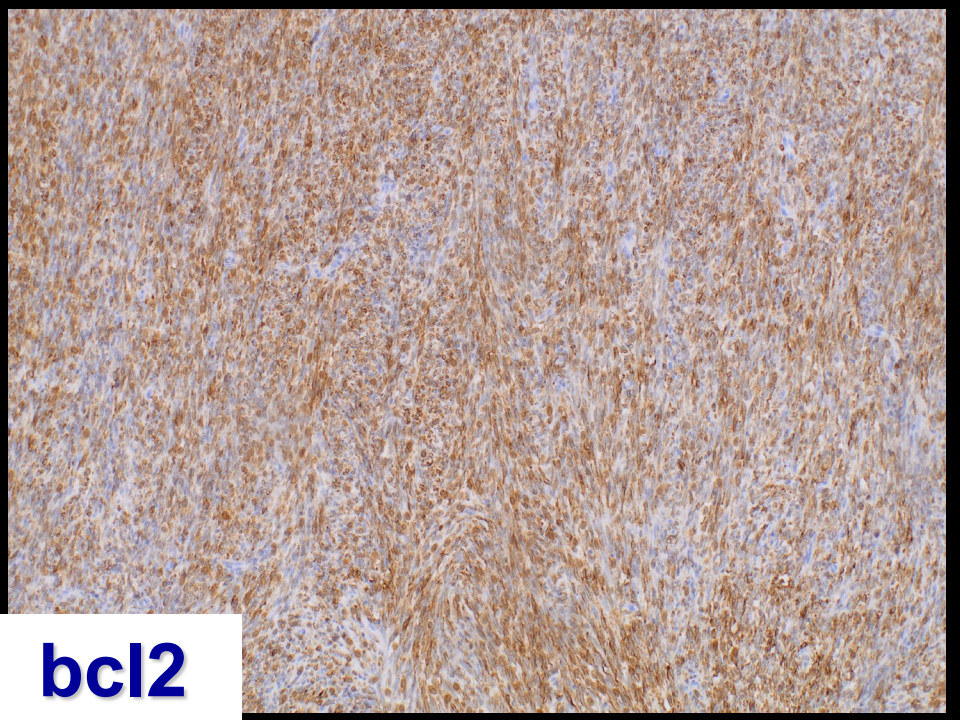
- polypoidný fragment lymfoepitelového tkaniva s centrálnou cystickou dutinou
- intaktný dlaždicový epitel
- histocytologicky blandný nádor, celkovo veľkosti 15x7 mm
- vretenovité bunky bez nápadnejších atypií
- infiltratívny rast
- nízka mitotická (MAI do 5 mf/10HPF) a proliferačná aktivita
- výrazná prímes granulovaných mastocytov

Diagnóza


VIM
CD34
S100
DEZ

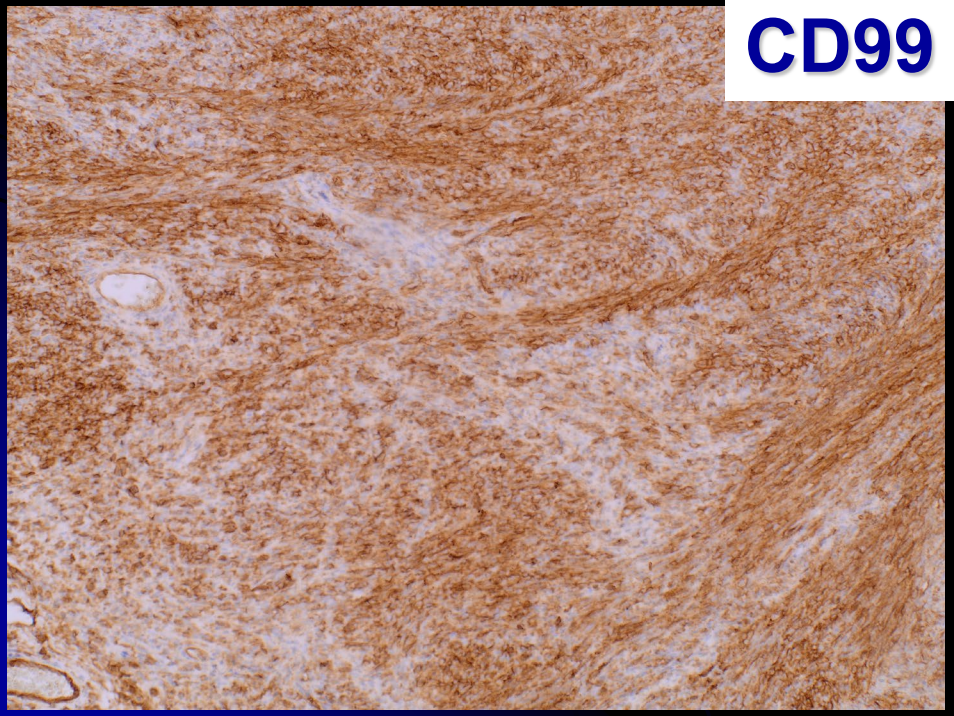

AE1/AE3
EMA

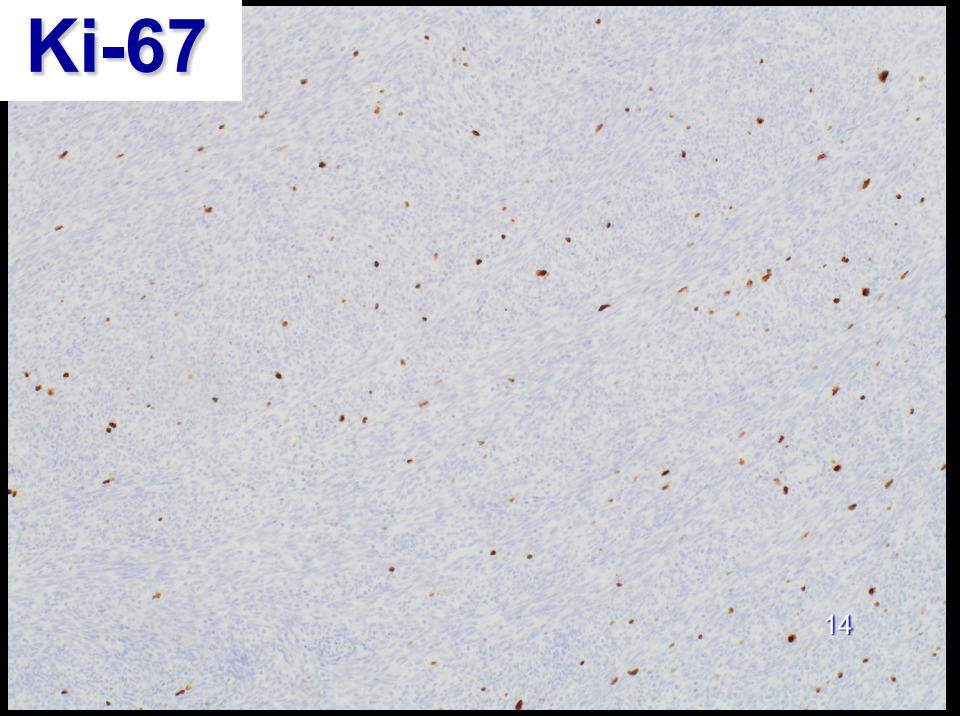


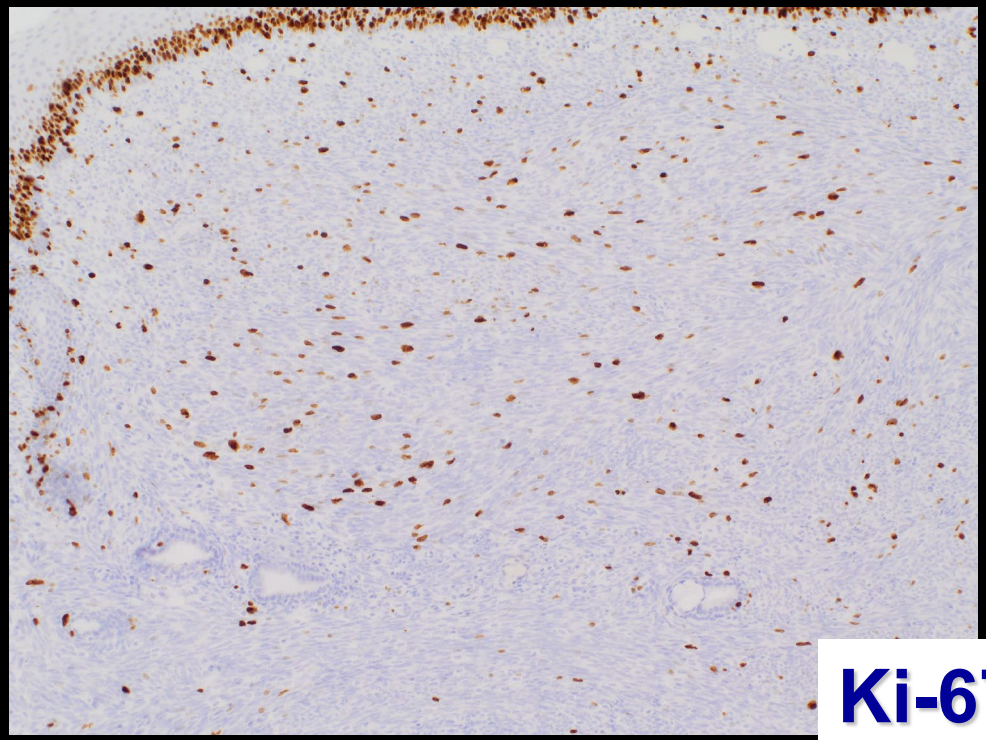


SMA bcl2

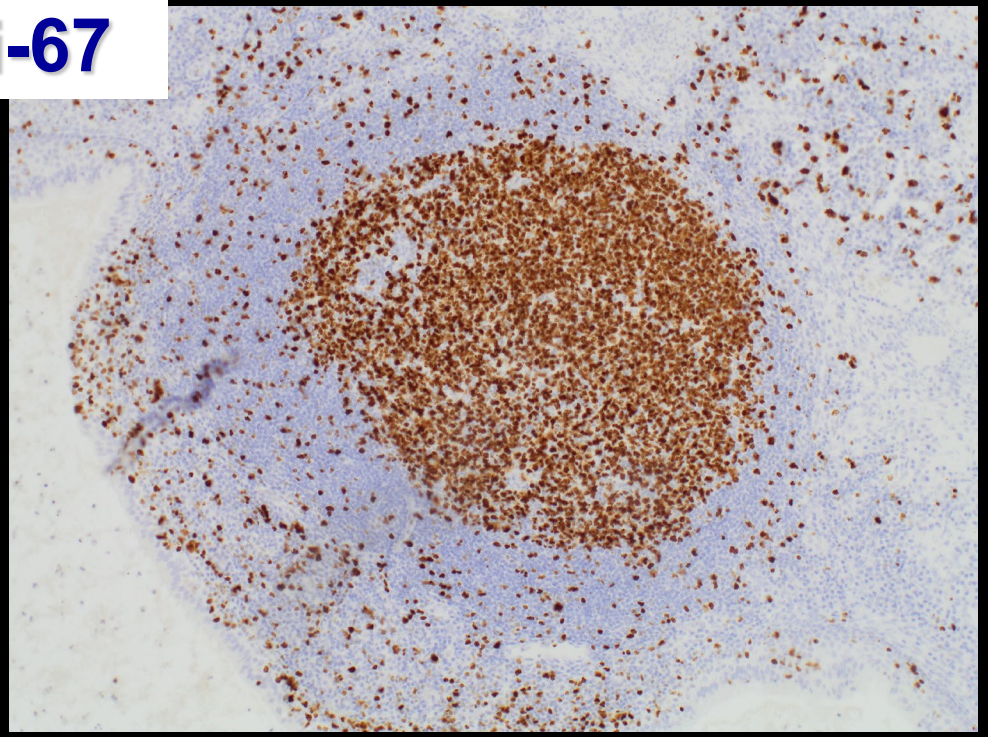
CD99 Ki-67







Ki-67



Sumarizácia imunoprofilu

- **Pozitivita:** vimentín, CD99, bcl-2
  EMA/CK fokálne

- Negativita: CD34, SMA, dezmín, SOX10,
  S-100 mono

FISH analýza

Metódou FISH dokázaný zlom/translokácia SYT (SS18) génu v cca 50% nádorových buniek (Poseidon SYT (18q11) Break probe, Kreatech N1).

RNDr. L.Fröhlichová, PhD.
RNDr. K.Fiedlerová
PAO UN LP, Košice

DIAGNÓZA

MONOFÁZICKÝ SYNOVIÁLNY SARKÓM TONZILÁRNEHO OBLÚKA

DIFERENCIÁLNA DIAGNÓZA

- spektrum fasciitíd
- SFT
- Fibrosarkóm
- MPNST
- RMS vretenobunkový

Primárny SS hlavy a krku

Review > Ear Nose Throat J. 2010 Jun;89(6):280-3.

Synovial sarcoma of the head and neck: a review of its diagnosis and management and a report of a rare case of orbital involvement

Shashidhar Kusuma [1], David J Skarupa, Kim A Ely, Anthony J Cmelak, Brian B Burkey

Affiliations – collapse

Affiliation

1 Vanderbilt Bill Wilkerson Center for Otolaryngology and Communication Sciences, Nashville, TN, USA. kusumas@ccf.org

PMID: 20556741

Abstract

Synovial sarcoma is typically an aggressive malignant tumor of the soft tissues, usually in the extremities, that affects young adults. Tumors of the head and neck are rare. Reported head and neck sites have included the hypopharynx (the most common site), the oropharynx, the larynx, and the soft tissues of the neck; only 4 cases of orbital involvement have been previously reported. We describe a case of synovial sarcoma of the medial canthus, which we discovered during a review of a tumor registry. The patient, an 18-year-old woman, underwent conservative excision and postoperative radiation therapy. Long-term follow-up detected no evidence of recurrence. Because there is no established, consistent approach to the treatment of synovial sarcoma of the head and neck, we also present a consensus management plan based on our review of the literature.

Primárny SS tonzily

Head and Neck Pathol (2010) 4:257–260
DOI 10.1007/s12105-010-0190-6

CASE REPORT

Molecular and Clinicopathological Findings in a Tonsillar Synovial Sarcoma. A Case Study and Review of the Literature

U. Vogel · M. Wehrmann · W. Eichhorn · B. Bültmann · M. Stiegler · W. Wagner

Received: 1 May 2010 / Accepted: 28 June 2010 / Published online: 13 July 2010

Abstract Synovial sarcoma (SS), 3–5% of which occurs in the head and neck region, has generally been regarded as high grade sarcoma. Recent analysis of clinical, morphological, and molecular characteristics of SS, however, identified low and high risk group of patients, resulting in important implications for the treatment of patients diagnosed with SS. We describe the case of a 31-year-old male who presented with biphasic SS with poorly differentiated areas (clinical stage IIA) in a palatine tonsil, an extremely rare site of SS. Molecular analyses revealed typical t(X;18) translocation of the SYT gene and a SYT/SSX1 fusion type. The tumor was surgically resected with free margins. Adjuvant radiotherapy or chemotherapy was not considered indicated. To date, the patient has remained free of tumor for 4 years after surgery. Literature review reveals that primary tonsillar HNSS has previously been documented only in three patients. In all of these patients the tumor was histologically biphasic; however only one published case and the case presented here showed areas of poor differentiation. We discuss the relevance of the presented findings with regard to prognostic and therapeutic considerations in SS in the head and neck region.

Keywords Synovial sarcoma · Tonsil · Tonsillar

U. Vogel · M. Wehrmann · W. Eichhorn · B. Bültmann
Institute of Pathology, University of Tübingen, Liebermeisterstr.8, 72076 Tübingen, Germany

M. Stiegler · W. Wagner (✉)
Department of Otolaryngology, Head and Neck Surgery, University of Tübingen, Elfriede-Aulhorn-Str.5, 72076 Tübingen, Germany
e-mail: w.wagner@med.uni-tuebingen.de

Introduction

Synovial sarcoma (SS) is defined as mesenchymal spin cell tumor that exhibits variable epithelial differentiati including gland formation, and specific chromosor translocation [1]. It may occur at any site in the bo however only 3–5% of SS arise in the head and neck reg [1–3]. We describe the clinical, pathological, and mol ular pathological findings in a biphasic, focal poorly d ferentiated SS in the left palatine tonsil (T-PDSS), a r site of this tumor [3–5]. We also discuss the relevance our findings with regard to prognostic and therapeu considerations in SS, particularly in the head and ne region (HNSS).

Case Report, Methods and Results

A previously healthy 31-year-old male presented with a 3-months history of left-sided sore throat, accompanied by increasing dysphagia. Pre-operative computed tomography (CT) disclosed a 4.2 × 2.3 × 2.6 cm encapsulated tumor in the left palatine tonsil. No signs of tumor necrosis or calcification, nor suspicious regional lymph nodes were seen on CT. The patient underwent transoral surgical resection. Intra-operative evaluation of frozen sections revealed an encapsulated malignant mesenchymal tumor covered by small remnants of pre-existing normal tonsillar tissue (Fig. 1a). Post-operative histological and immunohistological investigation showed biphasic SS with large areas of spindle cells that were positive for vimentin, CD99, bcl2, and calponin (all Dako, Hamburg, Germany) intermingled with glandular structures positive for cytokeratin (AEA1/3), EMA, CD99 and bcl2 (all Dako, Hamburg, Germany) (Fig. 1a–c). Thirty percentage of the

Diagnostic Pathology
BioMed Central

Oral presentation

Open Access

Synovial sarcoma of the left tonsil in a 31-year-old patient: report of a rare case

U Vogel*, M Wehrmann and B Bültmann

Address: Institut für Pathologie, Universität Tübingen, Germany
* Corresponding author

from 35te Tagung der Pathologen am Oberrhein/35th Meeting of Pathologists of the Upper Rhine Region (PATOR)
The Institute of Pathology, University Hospital Freiburg, Germany. 1 July 2006

Published: 14 March 2007

Diagnostic Pathology 2007, **2**(Suppl 1):S17 doi:10.1186/1746-1596-2-S1-S17

Aims
Synovial sarcoma (SS) is a mesenchymal spindle cell tumor with variable epithelial differentiation and a specific chromosomal translocation t(X;18)(p11.q11). Despite the name, SS is unrelated to synovium and may occur at any site of the body, mostly in the deep soft tissue of extremities. Around 5% arise in the head and neck region. Although the WHO textbook already describes the tonsils as an unusual site for the occurrence of SS, only two case reports could be found in the literature. Because of this rarity, we dare to present the following case.

Case report
A hitherto healthy 31-year-old male Turkish patient was admitted to hospital due to a left-sided sore throat accompanied by increasing dysphagia, which developed within 3–4 months. Preoperative computer tomography disclosed a 4.2 × 2.6 × 2.3 cm encapsulated tumor in the left tonsillar region expanding to the hypopharynx and the epiglottis.

Methods
Intraoperatively performed quick frozen sections detected an encapsulated malignant "small, round and blue" tumor, which was removed completely. Histologically, a malignant undifferentiated spindle-cell shaped component prevailed that stained immunohistochemically for CD99, bcl2, CD10 and calponin. At multiple sites, an additional pancytokeratin (AE1/AE3)- and EMA-positive epithelial differentiation of the tumor was detected, partly with glandular differentiation. Nuclear ki-67 expression was present in about 50% of the spindle cells and in about 15% of the epithelial compartment. The presence of the SYT(SS18) rearrangement indicative for the t(X;18) translocation and characteristic for synovial sarcoma was demonstrated by fluorescence in situ hybridization.

Results
Based on the histological, immunohistological and molecular pathological findings, the tumor was classified as biphasic SS with poorly differentiated areas, qualifying for grade III.

Conclusion
The diagnosis of SS should be kept in mind even in such unusual sites as the tonsils.

Case Report
Middle East Journal of Cancer 2010; 1(3): 141-146

Synovial Sarcoma of the Palatine Tonsil: Report of Two Cases and Review of the Literature

Bijan Khademi*, Hajar Bahranifard*, Mohammad Mohammadianpanah**♦, Mohammad Javad Ashraf***, Negar Azarpira***, Mehdi Dehghani****

*Cancer Research Center and Department of Otolaryngology, Shiraz University of Medical Sciences, Shiraz, Iran
**Department of Radiation Oncology, Shiraz University of Medical Sciences, Shiraz, Iran
***Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran
****Department of Hematology Oncology, Shiraz University of Medical Sciences, Shiraz, Iran

Abstract

Here, we describe young men with synovial sarcoma in the palatine tonsil, who presented with a 3-4 month history of progressive sore throat, tonsillar ulcerative mass and bleeding. Clinical and radiological examinations revealed that the tumors arose from the palatine tonsil and extended to the parapharyngeal space. Both tumors were too advanced to remove completely; therefore, they underwent surgical debulking during tonsillectomy and partial pharyngectomy. Histopathological and immunohistochemical studies confirmed the diagnosis of synovial sarcoma of the palatine tonsil. Despite postoperative radiotherapy and systemic chemotherapy, they relapsed 18 and 22 months later. The first patient died from unresectable local recurrent disease three years after primary diagnosis, and the second patient is alive after 36 months, but suffers from unresectable locoregional recurrent disease and is receiving palliative chemotherapy and supportive care.

Keywords: Synovial sarcoma, Palatine tonsil, Tonsillectomy, Radiotherapy, Chemotherapy

♦Corresponding Author:
Mohammad Mohammadianpanah, MD, Cancer Research Center, Department of Radiation Oncology, Nemazee Hospital, Shiraz University of Medical Sciences, 71936-13311 Shiraz, Iran
Tel/Fax: +98-711-6474320

Introduction

Synovial sarcomas account for 6% to 9% of all adult soft tissue sarcomas. These malignant soft tissue neoplasms primarily arise from the extremities during young adulthood.[1] (8%) and retroperitoneal/abdominal region (7%), which are the most frequent non-extremity primary sites of disease.[1] The head and neck region is a rare primary site for this neoplasm and only 3-5% of all

Primárny SS tonzily a orofaryngu

Head and Neck Pathol (2013) 7:400–403
DOI 10.1007/s12105-013-0440-5

CASE REPORT

Primary Monophasic Synovial Sarcoma of the Tonsil: Immunohistochemical and Molecular Study of a Case and Review of the Literature

Danny Soria-Céspedes · Aldo Iván Galván-Linares · Cuauhtemoc Oros-Ovalle · Francisco Gaitan-Gaona · Carlos Ortiz-Hidalgo

Received: 7 February 2013 / Accepted: 1 April 2013 / Published online: 7 April 2013

Abstract Synovial sarcoma (SS) arises primarily in the lower extremities with a predilection for sites in proximity to large joints, such as the knee. It rarely occurs in the head and neck region, and the tonsil is an unusual site for the tumor, with only eight previously published cases in this anatomical site. We present a case of a primary monophasic SS arising in the right tonsil in a 63-year-old male. His medical history was noncontributory. Immunohistochemistry showed that cytokeratin OSCAR, EMA, Bcl-2, vimentin, PGP 9.5, and TLE1 were diffusely positive. A molecular analysis using RT-PCR indicated that the patient was positive for the *SYT/SSX1* fusion transcript. A diagnosis of monophasic synovial sarcoma of the tonsil was made.

Keywords Synovial sarcoma · Tonsil · TLE1 · *SYT/SSX1*

Introduction

Synovial sarcoma (SS) is a rare high-grade soft tissue tumor of unknown histogenesis that primarily arises from the deep soft tissues of the extremities, and it accounts for 6–10 % of all soft tissue tumors [1, 2]. Only 3–5 % arise in the head and neck region, and SS arising in the tonsil is a rare finding, with only eight well-documented cases reported in the English language literature [1–6]. We describe the histopathological findings of a primary synovial sarcoma of the palatine tonsil arising in a 63-year-old male, with immunohistochemical expression of TLE1 and molecular detection of the SYT-SSX1 fusion gene transcript using reverse transcription-polymerase chain reaction (RT-PCR).

Case Report

A 63-year-old man presented with a 3-month history of progressive dysphagia and a growing tumor in the right side of the oropharynx. The remainder of his medical history was non-contributory. A physical examination revealed right tonsillar hypertrophy and a lobulated tonsillar surface. There were no cervical lymphadenomegalies. A right tonsillectomy was performed with the patient under general anesthesia.

Grossly, the right tonsil measured 5.7 × 4 × 3 cm with a smooth lobulated surface and firm consistency. The cut surface of the tonsil was solid, homogeneous, and white-tan, without necrosis or hemorrhage (Fig. 1). The specimen was fixed in 10 % buffered formalin and embedded in paraffin. Histologic 4-μm sections were stained by hematoxylin and eosin. Immunohistochemistry was performed using the standard streptavidin–biotin complex method. The antibodies and methodology used in this study are summarized in Table 1.

A histopathological examination showed a monophasic synovial sarcoma. The tumor was composed of closely packed spindle cells with scant pale cytoplasm forming

D. Soria-Céspedes · C. Ortiz-Hidalgo (✉)
Department of Pathology, The American British Cowdray Medical Center, Sur 136#116. Col Las Américas, 01120 Mexico, DF, Mexico
e-mail: dannysoriaces@yahoo.com

A. I. Galván-Linares · C. Oros-Ovalle · F. Gaitan-Gaona
Department of Pathology, Hospital Central "Dr. Ignacio Morones Prieto", San Luis Potosí, Mexico

C. Ortiz-Hidalgo
Department of Tissue and Cell Biology, Universidad Panamericana, Mexico City, Mexico

Indian J Surg Oncol (March 2014) 5(1):75–77
DOI 10.1007/s13193-013-0285-0

CASE REPORT

Synovial Sarcoma Oropharynx - A Case Report and Review of Literature

Nagendra P. B. Kadapa · L. Sudarshan Reddy · Ranganatha Swamy · Kumuda · M. Vishnu Vardhan Reddy · L. M. Chandra Sekhara Rao

Received: 13 August 2013 / Accepted: 30 December 2013 / Published online: 15 January 2014

Introduction

Synovial sarcoma is a rare malignant tumor that derives from a mesenchymal precursor stem cell that is unrelated to mature synovial tissue. It is primarily a disease of young adults typically arising in the deep soft tissues of the extremities, most commonly, the lower thigh-knee region. Reports in the literature have underscored the occurrence of this tumor in unusual sites, many seemingly distant from synovium-lined spaces, particularly, the abdominal wall [1] and head and neck region. The vast majority of this latter group, mostly case reports, were confined to the cervical and parapharyngeal region [2–8]. In this article, we report a case of synovial sarcoma that originated in oropharynx which is extremely rare. This is the first of its kind at our institute.

Case Report

A 40-year-old man presented with a history of foreign body sensation in the throat mainly on the left side & progressively worsening dysphagia and nasal regurgitation. He did not have other symptoms relating to the ear, nose and throat. He had no history of a loss of weight or appetite. He had given up smoking 7 years earlier.

Examination revealed the presence of a pedunculated mucosal lesion projecting into oropharynx arising from just behind the posterior pillar of tonsil on left side. No cervical lymphadenopathy was noted. Findings on the remainder of the ENT examination were normal. A lateral neck x-ray demonstrated an abnormal protrusion of soft tissue from the lateral wall of oropharynx on left side.

The patient underwent a pharyngoscopy showing well demarcated globular mass of 4×3 cm arising from left lateral pharyngeal wall just behind posterior pillar of tonsil. Computed Tomography scan revealed a lobulated homogenously enhancing soft tissue density mass lesion in left lateral pharyngeal wall extending from tip of epiglottis to hyoid bone. The mass was removed as an excisional biopsy by transoral endoscopic assisted approach under general anaesthesia (Figs. 1 and 2).

Histo-pathologic examination showed biphasic tumour with slit like spaces lined by *epithelial cells and Spindle cells.* Epethelial cells were cuboidal and pale with vesicular nuclei and *spindle cells* were in fascicles and bundles, closely approximated to epithelial and glandular spaces (Fig. 3) consistent with *synovial sarcoma/carcinosarcoma.* Excision of lesion was deemed complete. Immunohistochemistry highlighted spindle cells strongly expresses for *vimentin,* focal weak cytoplasmic positive for *bcl-2, pancytokeratin* positive in epithelial cells and *CD99 was* negative consistent with *Synovial sarcoma*ma/*Carcino sarcoma.* It was subjected to polymerase chain reaction technique for t(X; 18) (p11.2; q11.2) SYT-SSX analysis. Results turned positive for *SSX1* and *SYT genes* and confirmed as biphasic synovial sarcoma [9].

Patient received adjuvant radiotherapy *66 Gy in 33 fractions.* Case was followed up for 3 years at regular intervals for both local and metastatic work up and is free of any symptoms. Residual or recurrent mass ruled out by VLS.

N. P. B. Kadapa · L. S. Reddy · R. Swamy · Kumuda · M. V. V. Reddy · L. M. C. S. Rao
Department of Surgical Oncology, Basavatarakam Indo-American Cancer Hospital, Banjarahills, Hyderabad, Andhra Pradesh, India

L. S. Reddy (✉)
Osmania Medical College/Govt. ENT Hospital, Koti, Hyderabad 500095, Andhra Pradesh, India
e-mail: drlsudarshanreddy9@gmail.com

L. S. Reddy
e-mail: aoiapstatebranch@gmail.com

Primárny SS tonzily

Case Report doi: 10.5146/tjpath.2018.01449

Pediatric Tonsillar Synovial Sarcoma- Very Rare Localization: A Case Report and Review of the Literature

Koray YALÇIN[1], Gülen TÜYSÜZ[1], Mine GENÇ[2], İrem Hicran ÖZBUDAK[3], Alper Tunga DERİN[4], Kamil KARAALİ[5], Alphan KÜPESİZ[1], Elif GÜLER[1]

Department of [1]Pediatric Hematology and Oncology, [2]Radiation Oncology, [3]Pathology, [4]Nose Throat Ear Surgery, and [5]Radiology, Akdeniz University Medicine Faculty, ANTALYA, TURKEY

ABSTRACT

Tonsillar synovial sarcoma is an extremely rare entity and only 9 adult patients have been reported up to now. Here, we describe the first pediatric tonsillar synovial sarcoma of the literature in a patient who presented with a 2-month history of dysphagia and snoring. Clinical and radiological examinations showed that the tumor arose from the right palatine tonsil and narrowed the parapharyngeal space. An incisional biopsy from the palatine tonsil revealed the diagnosis of synovial sarcoma. The patient has underwent total tonsillectomy and received radiotherapy and chemotherapy because of the positive surgical margins. The patient is clinically in good condition and free of tumor 30 months after the initial diagnosis. We achieved a long-term complete remission with a combination of surgery, radiotherapy and chemotherapy in our case. Tonsillar synovial sarcoma should be kept in mind while dealing with tonsillar masses. We can conclude that a multidisciplinary approach is warranted while treating synovial sarcoma with this localization.

Key Words: Synovial Sarcoma, Tonsillar neoplasm, Pediatric tumor , Dysphagia, Snoring

INTRODUCTION

Synovial sarcoma (SS) is the most common soft tissue sarcoma after rhabdomyosarcoma in the pediatric population. The annual incidence rate is 0.5-07 /million in children and adolescents younger than 20 years of age (1,2). Synovial sarcoma primarily arises from deep soft tissues of the extremities, usually from the lower extremities followed by the upper extremities, trunk, and retroperitoneal/ abdominal region. Head and neck localization is rare with a percentage of only 3-10% of all SS cases (3,4). The most frequent areas in the head and neck region include the hypopharynx, followed by the parapharyngeal space and post pharyngeal area. Primary SS of the palatine tonsil is extremely rare and only 9 adult cases have been reported so far (5-11). As far as we are aware, we report the first pediatric case of tonsillar SS in the literature. Because of the unusual localization, the management and treatment of tonsillar SS is based on case reports (5-11). It is known that both the treatment and prognosis of SS differs among children and adults (2, 12). We therefore believe this report may contribute to the existing literature with its rare location and treatment approach.

CASE REPORT

A 13-year-old boy presented with a 2-month history of dysphagia and snoring. His physical examination revealed an ulcerative, green colored mass in the right tonsil which narrowed the oropharynx. Contrast-enhanced magnetic resonance imaging (MRI) revealed a 50x47x45 mm solitary mass originating from the right lateral oropharynx, extending to the hypopharynx and narrowing the oropharyngeal lumen irregularly (Figure 1,2). The Positron Emission Tomography (PET) scan showed metabolic activity (SUV max: 9.3) in the right tonsillar area. There was no evidence of distant metastases or regional lymph node involvement in the PET scan. To make a diagnosis, an incisional biopsy was performed from the right tonsil and the histopathological examination of the specimen revealed the presence of a biphasic SS. Immunohistochemical examination for PanCK, CK19, CK18, CK7 revealed positive staining in the epithelial component and staining for vimentin revealed positive staining in the mesenchymal component. The Hematoxylin-Eosin staining of tumor is illustrated in Figure 3 and the CK7 immunopositivity of the epithelial cells is illustrated in Figure 4. Muscle and neural markers were negative. The Ki67 proliferation index was 50%. The diagnosis was confirmed by additional

(Turk Patoloji Derg 2020, 36:82-86)

Received : 02.10.2018 **Accepted :** 12.11.2018

Correspondence: Elif GÜLER
Department of Pediatric Hematology Oncology
Akdeniz University Medical Faculty, Antalya, TURKEY
E-mail: elifguler@akdeniz.edu.tr Phone: +90 242 249 60 00 / 6539

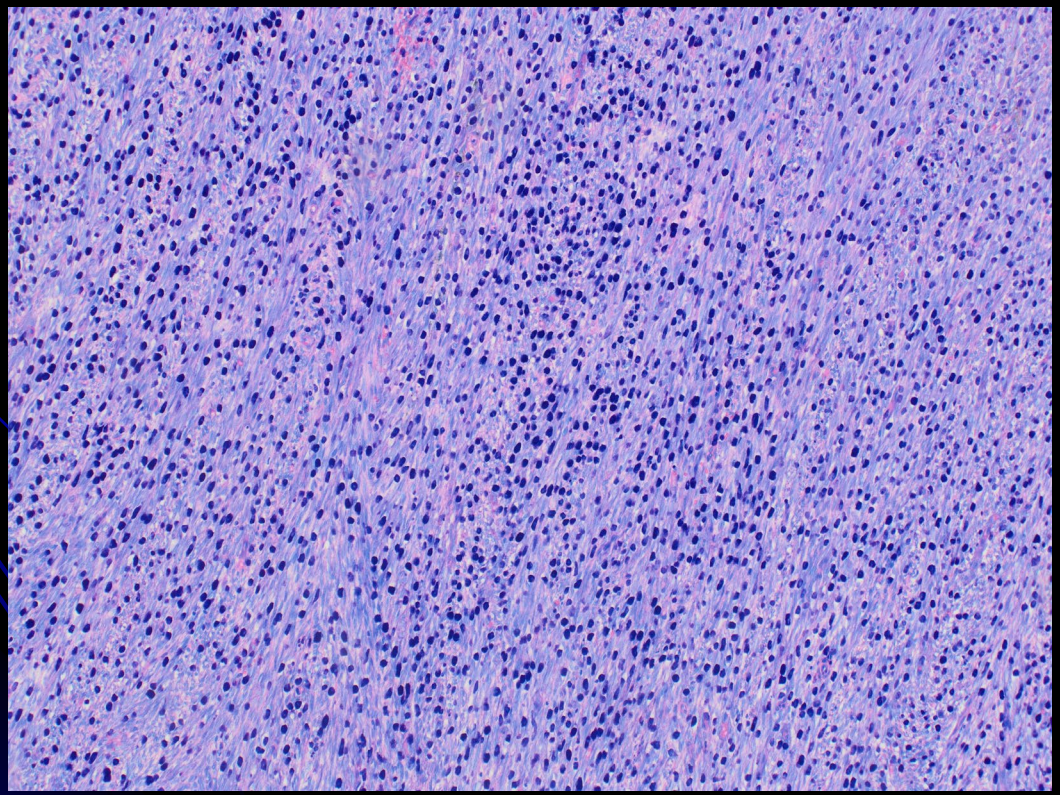
9 pacientov v literatúre, všetko muži nad 18 rokov

Primárny SS hlavy a krku

- extrémne zriedkavá lokalizácia (len cca 3-7 % všetkých SS)
- výskyt v každom veku, avšak viac ako 50% u adolescentov s rovnakým zastúpením pohlaví, ale v tonzile takmer výlučne u mužov
- malé tumory (do 1 cm) sú zriedkavé s excelentnou prognózou
- pozitivita TLE1 je dobrým, ale nešpecifickým markerom, môže byť aj v AFH, SFT, PNST, Ca prsníka, SETTLE, GIST, a i ných sarkómoch (LMS, RMA, chondro Sa, clear cell Sa, ESS...)

Mišo Pauer, 199?

...keď vidíš veľa mastocytov v mezenchmálnom vretenobunkovom tumore, mysli v prvom rade na synoviálny sarkóm...



Ďakujem za pozornosť