Hospital Universitario de Fuenlabrada

SaludMadrid

Comunidad de Madrid

XXV Congreso de la SEAP-IAP

Club de Patología de Partes Blandas

Juan C. Tardío
Servicio de Anatomía Patológica
Hospital Universitario de Fuenlabrada

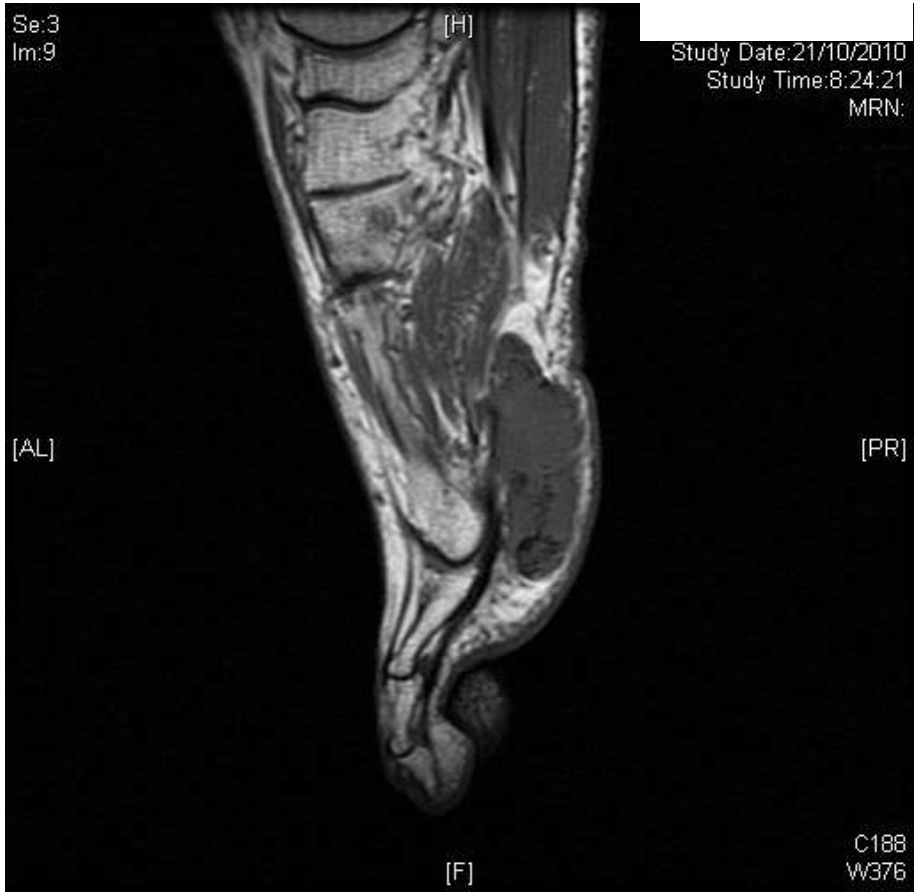
Antecedentes Personales

- **Mujer 52 años**
- **Osteomalacia vit D-resistente (8 años) y osteoporosis**
- **Doble fractura de rama isquiopubiana derecha y fractura de cadera izquierda**
- **Dolores óseos y dificultad para la deambulación**

Proceso Actual

- **Consulta en Dermatología por “ampolla hemorrágica” dolorosa en la planta del pie izquierdo**
- **Lesión tumoral exofítica de 1.5 cm sangrante al roce**
- **Dx clínico: Granuloma piógeno vs Melanoma**
- **Biopsia en cuña fragmentada**

Diagnóstico anatomopatológico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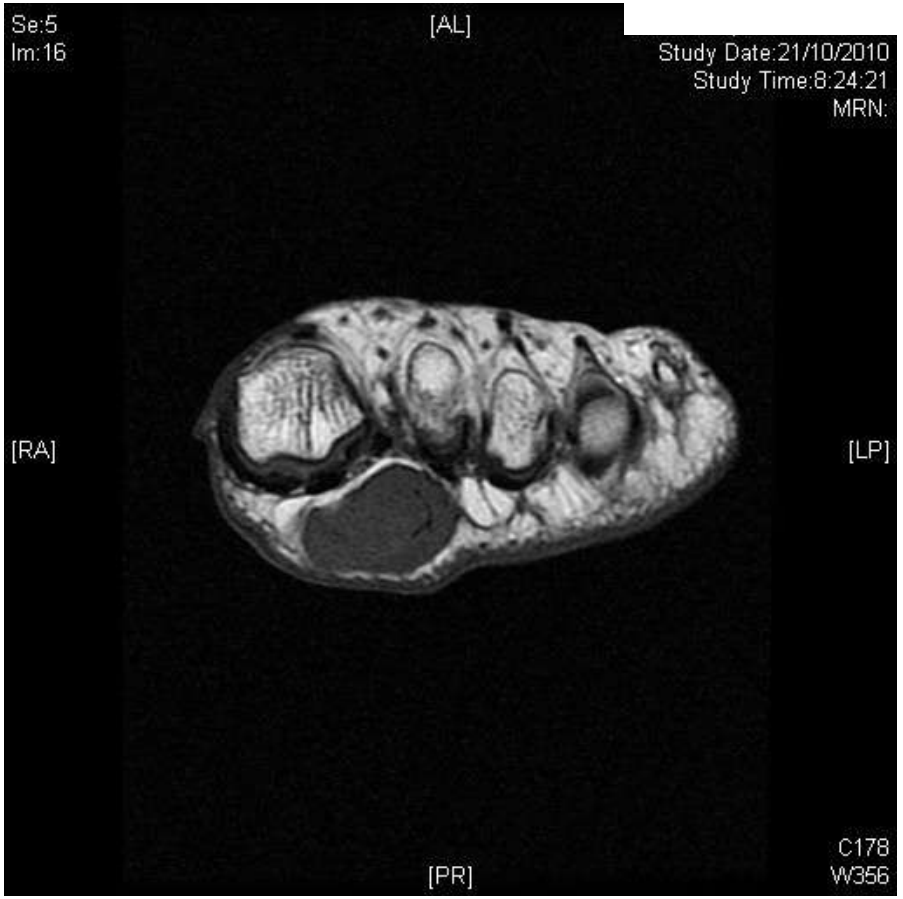
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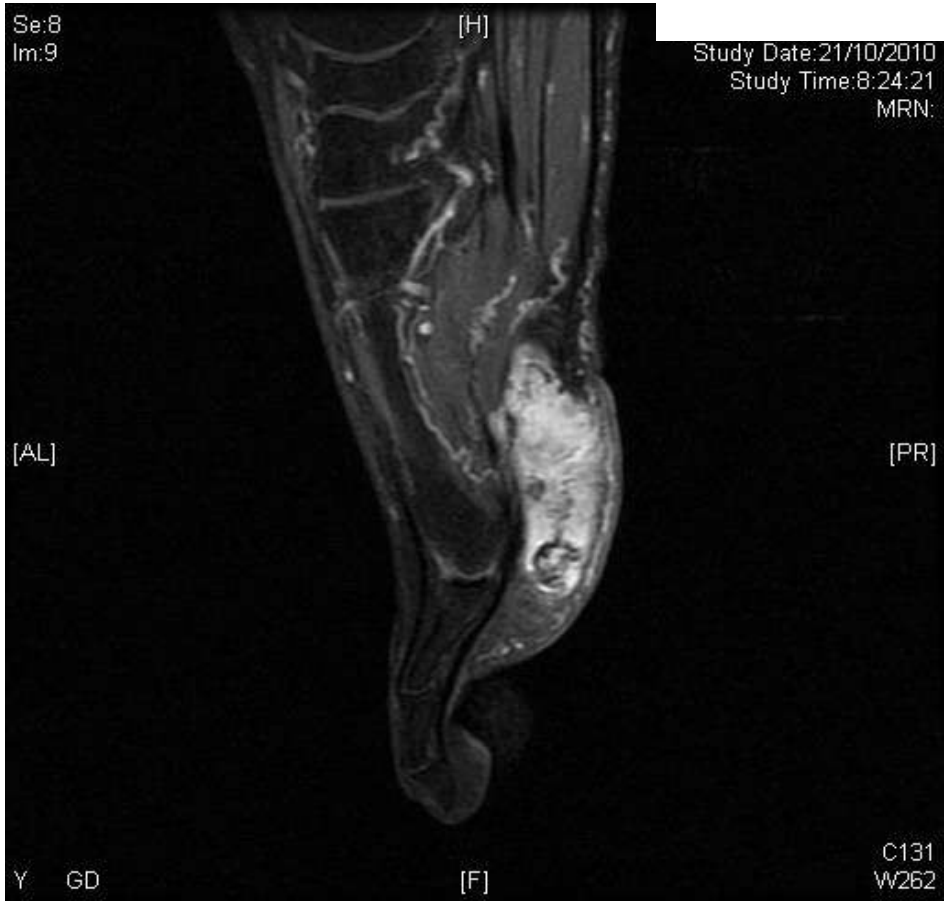
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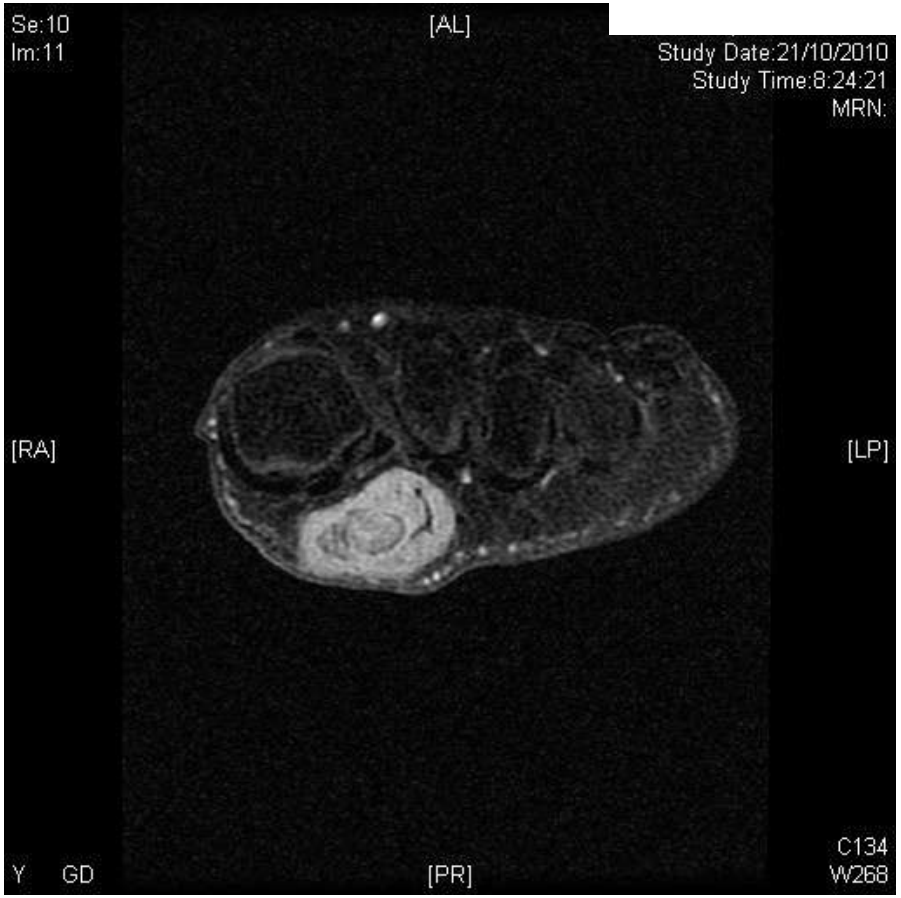
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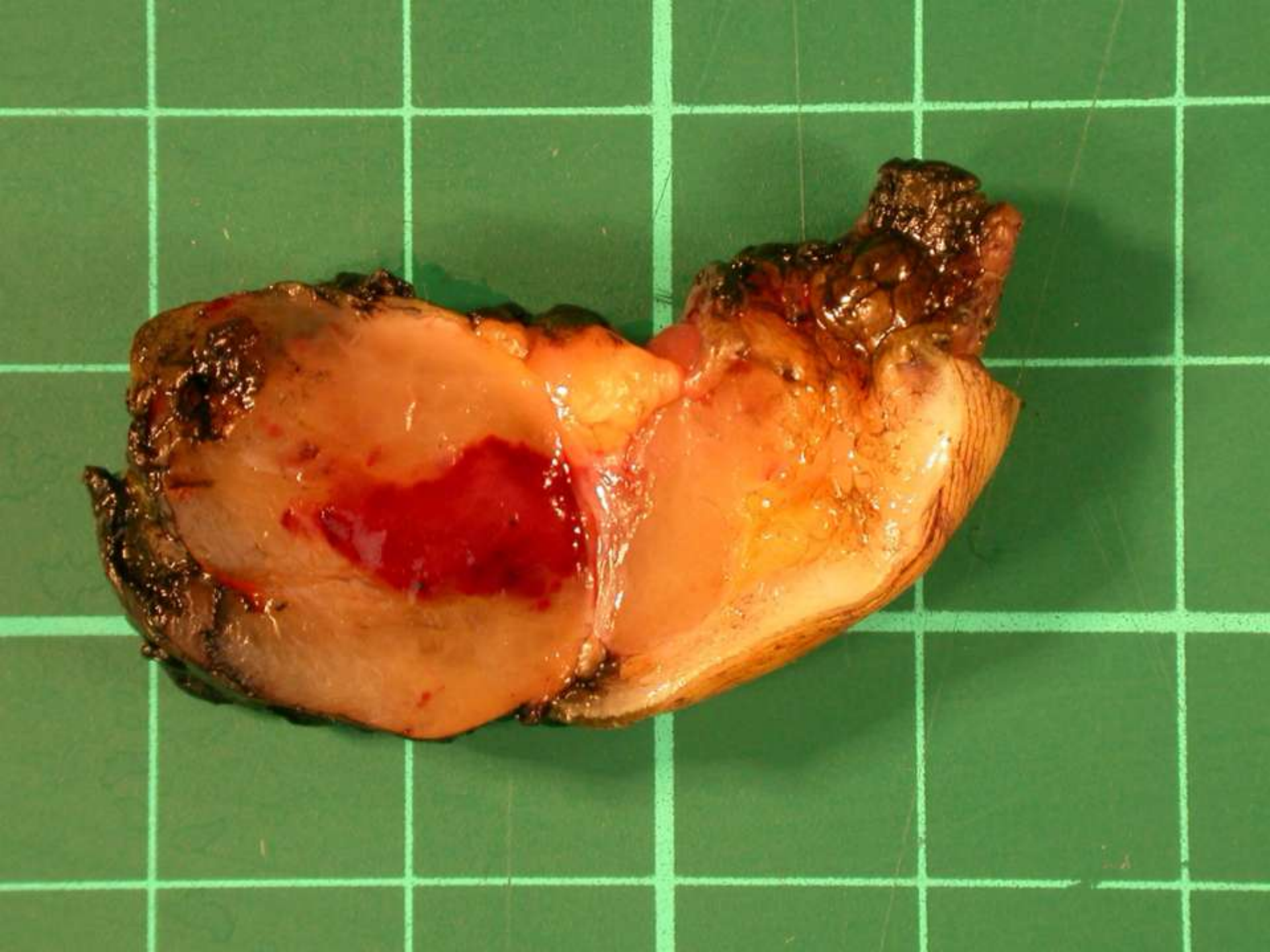
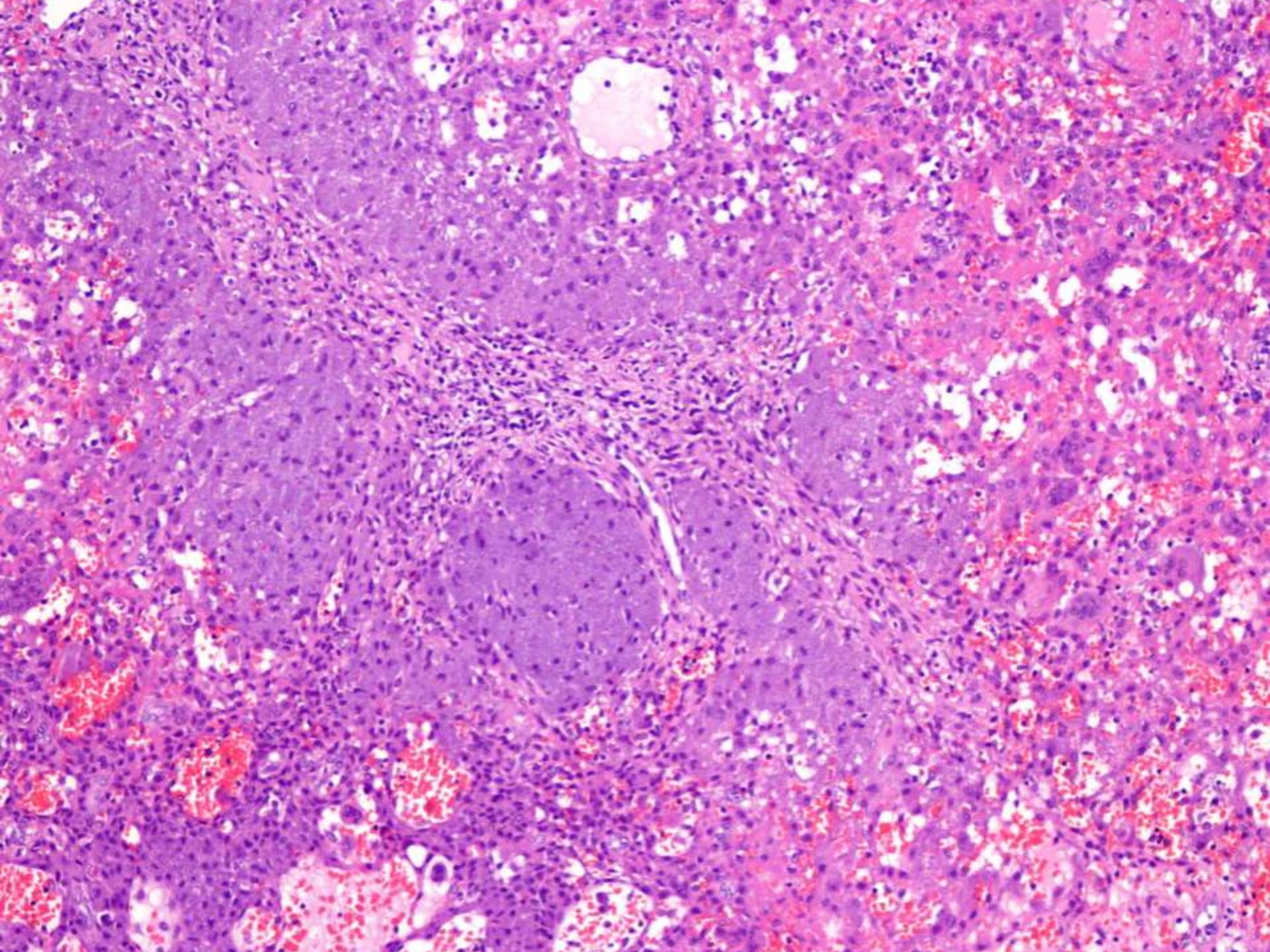
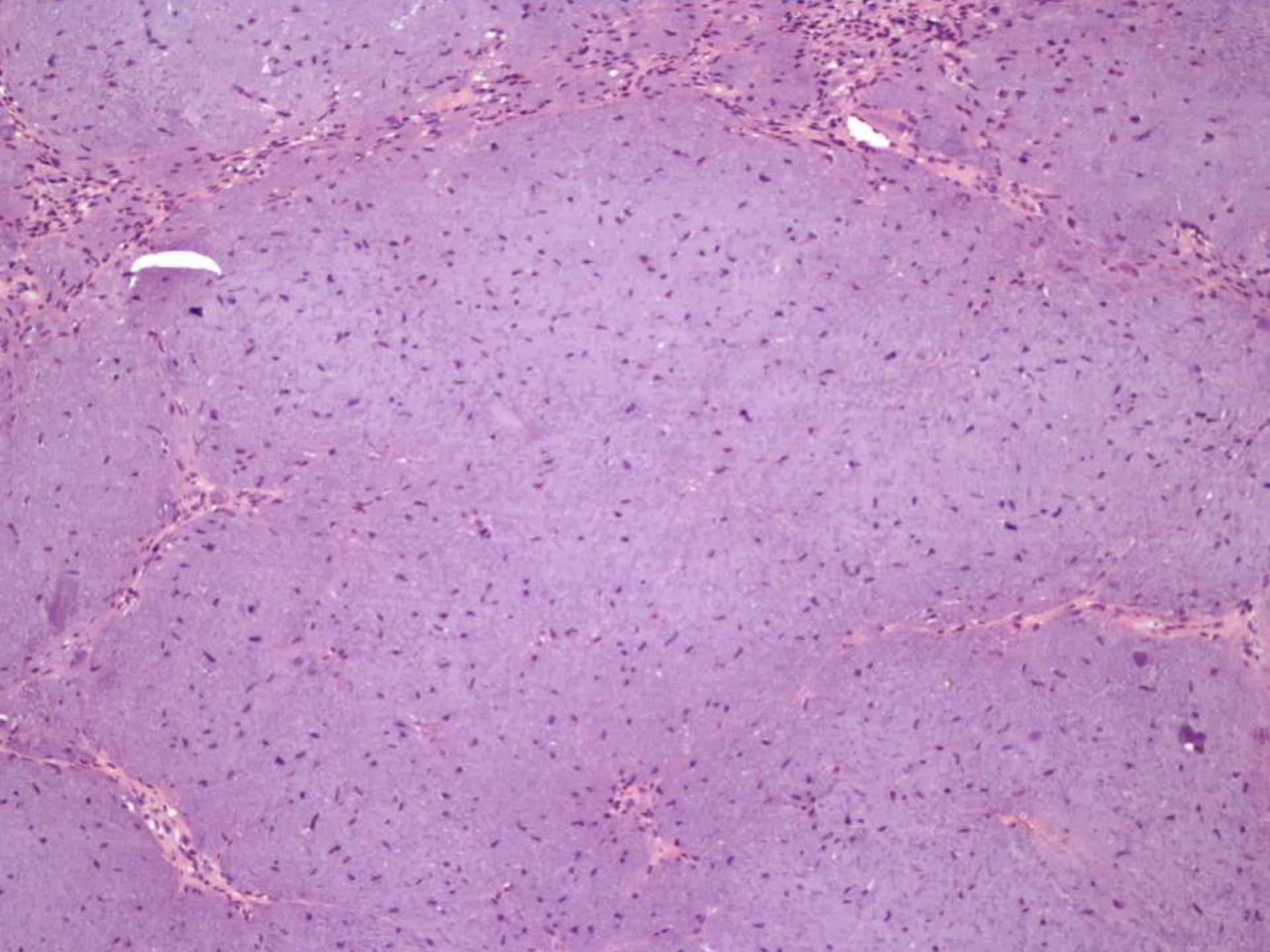
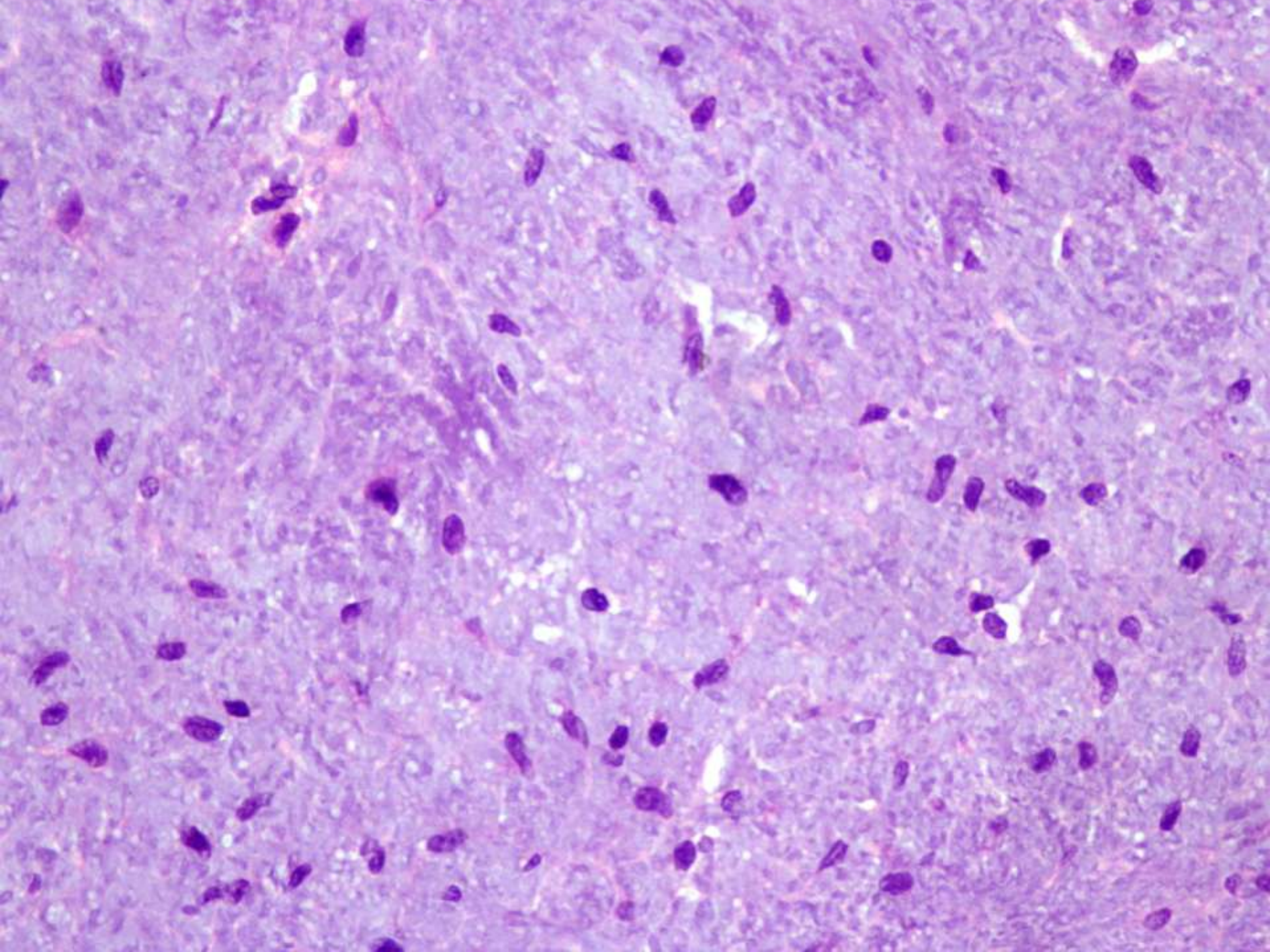
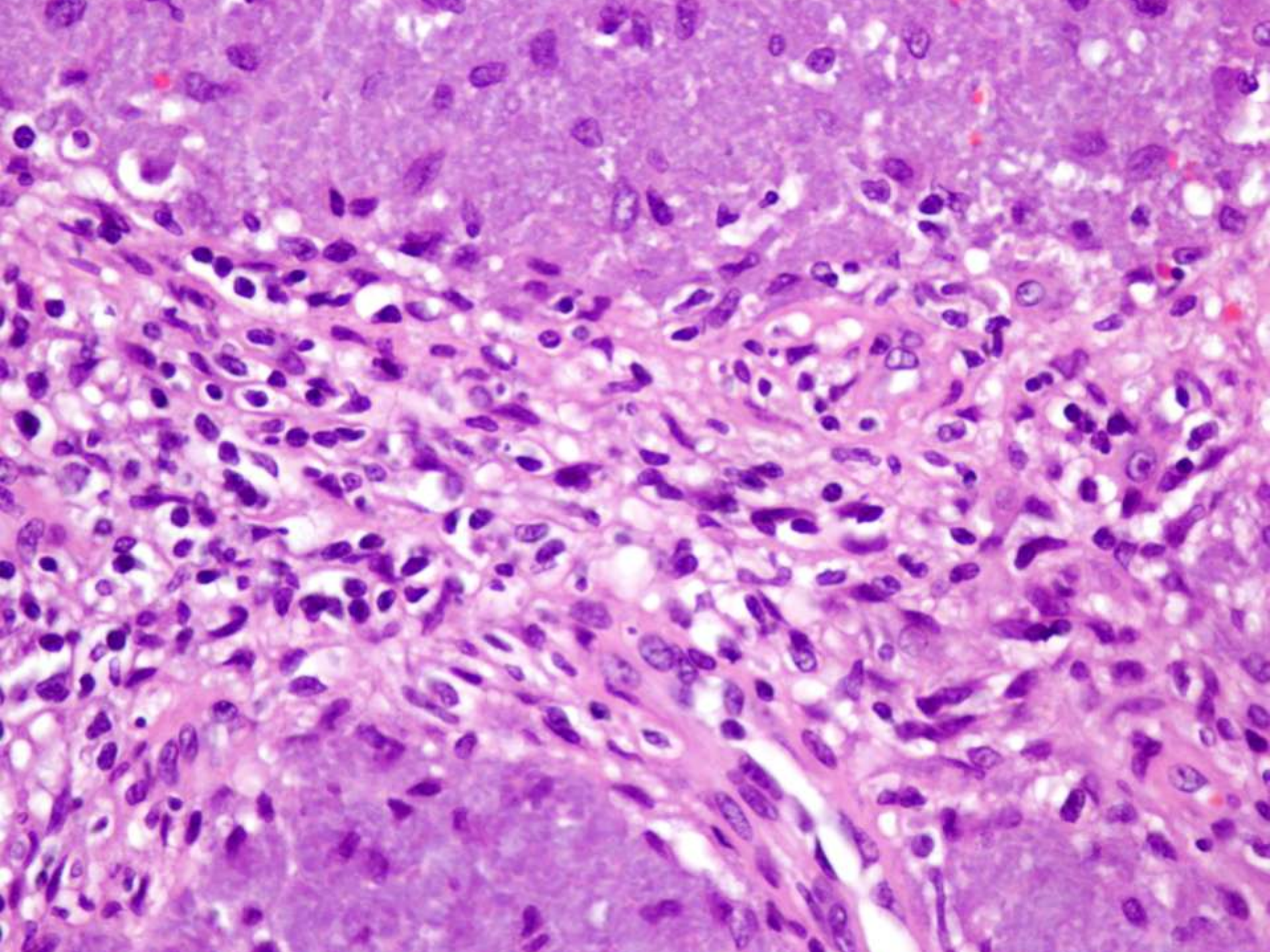
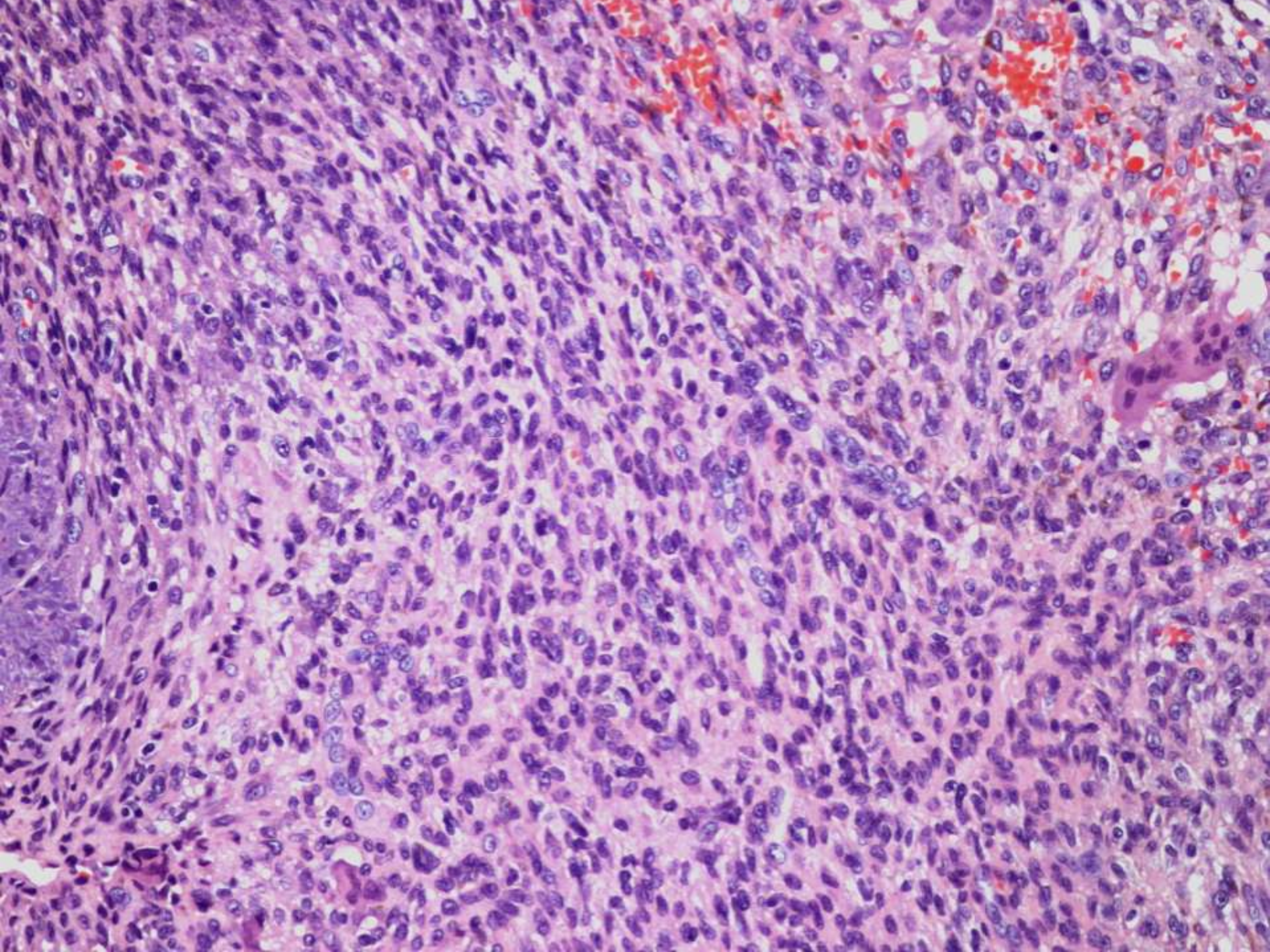


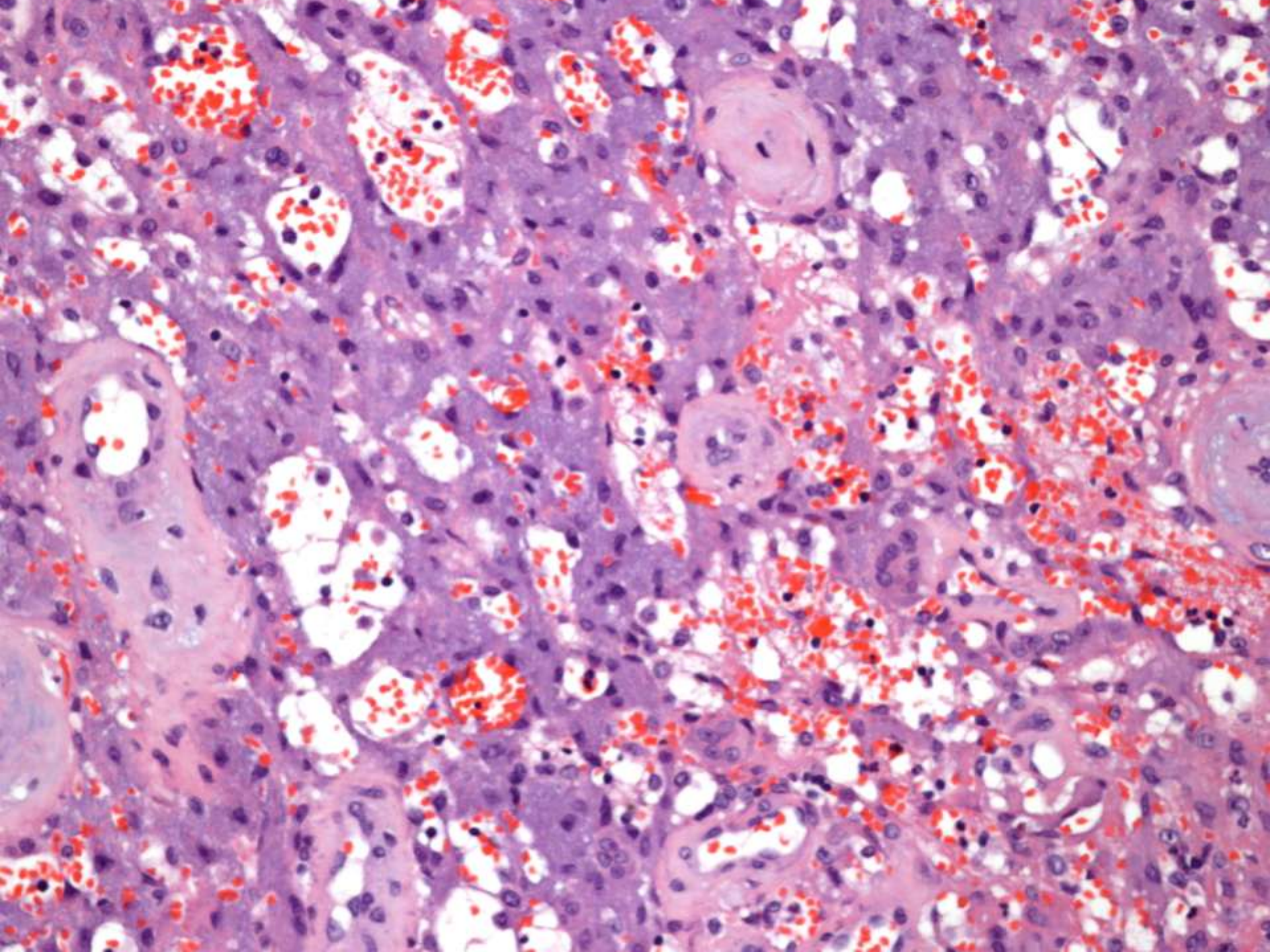


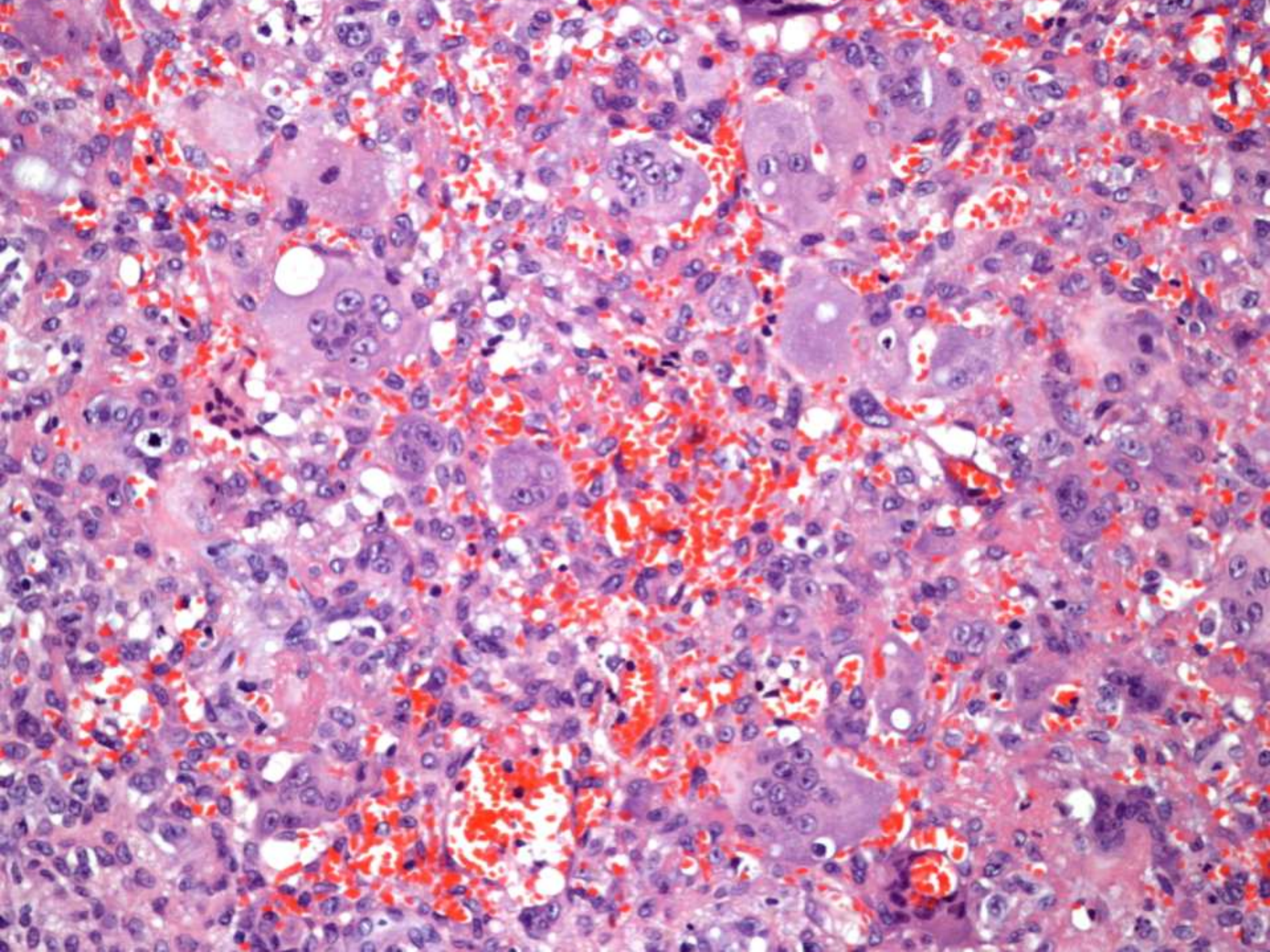


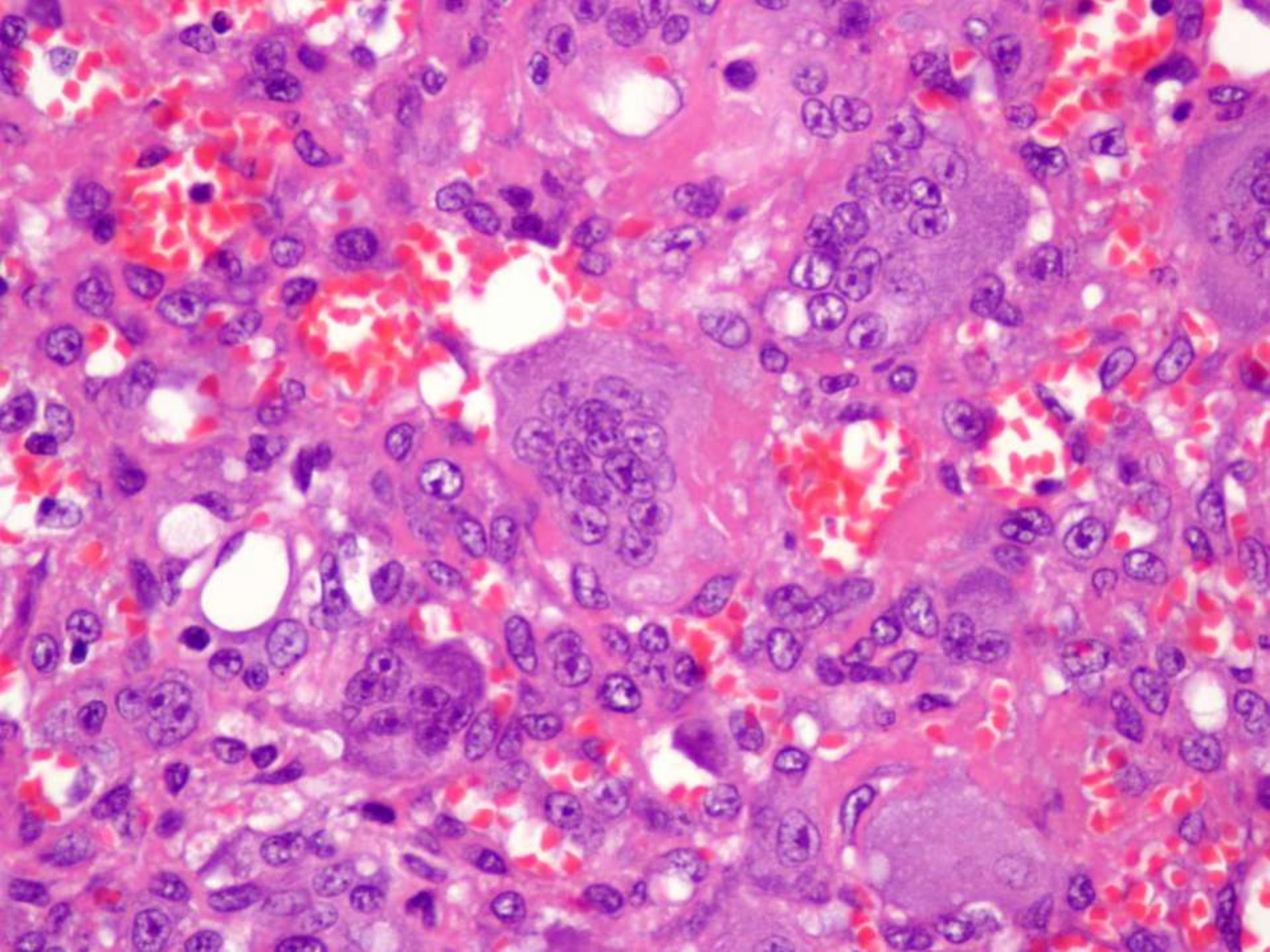


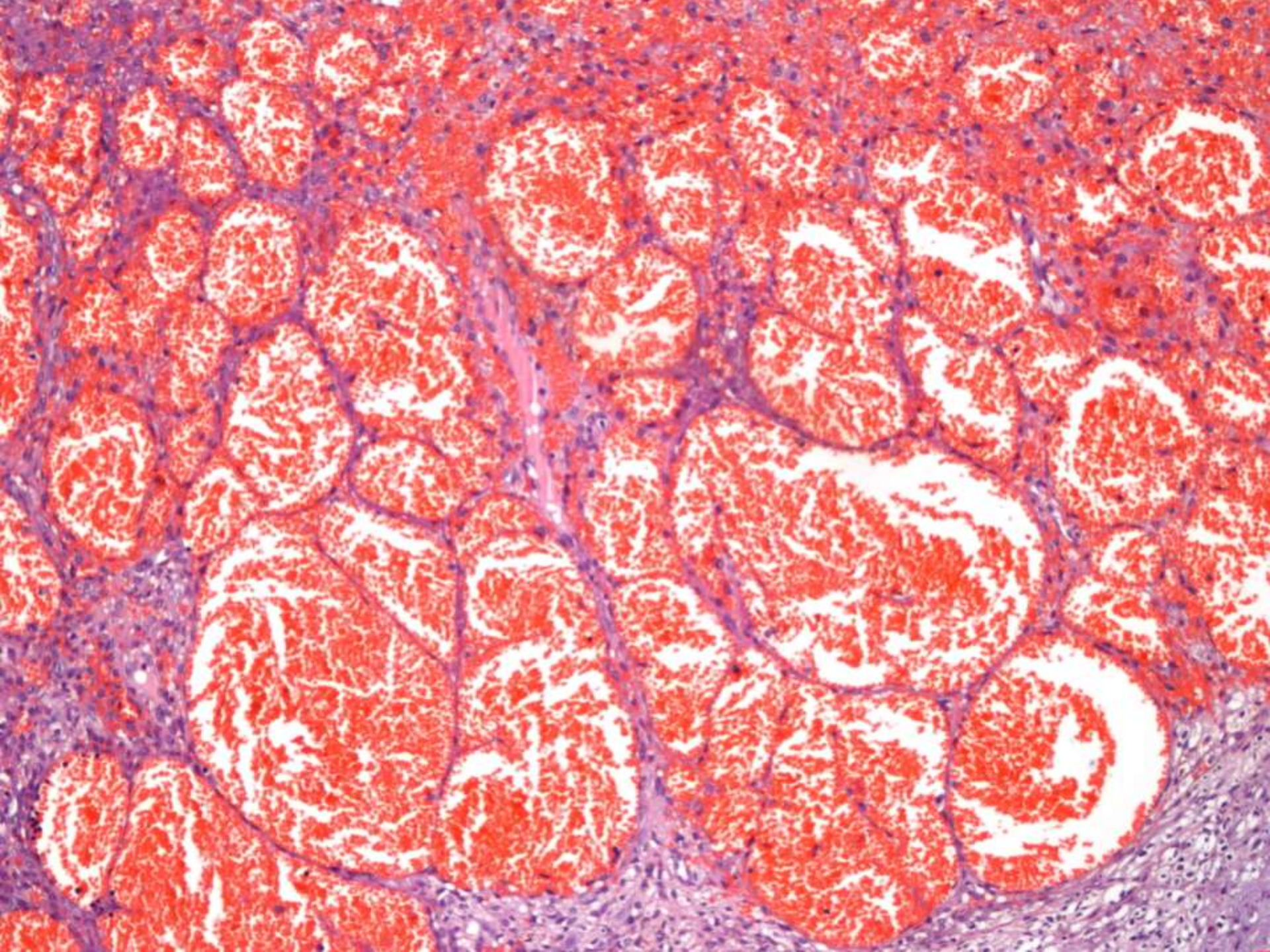


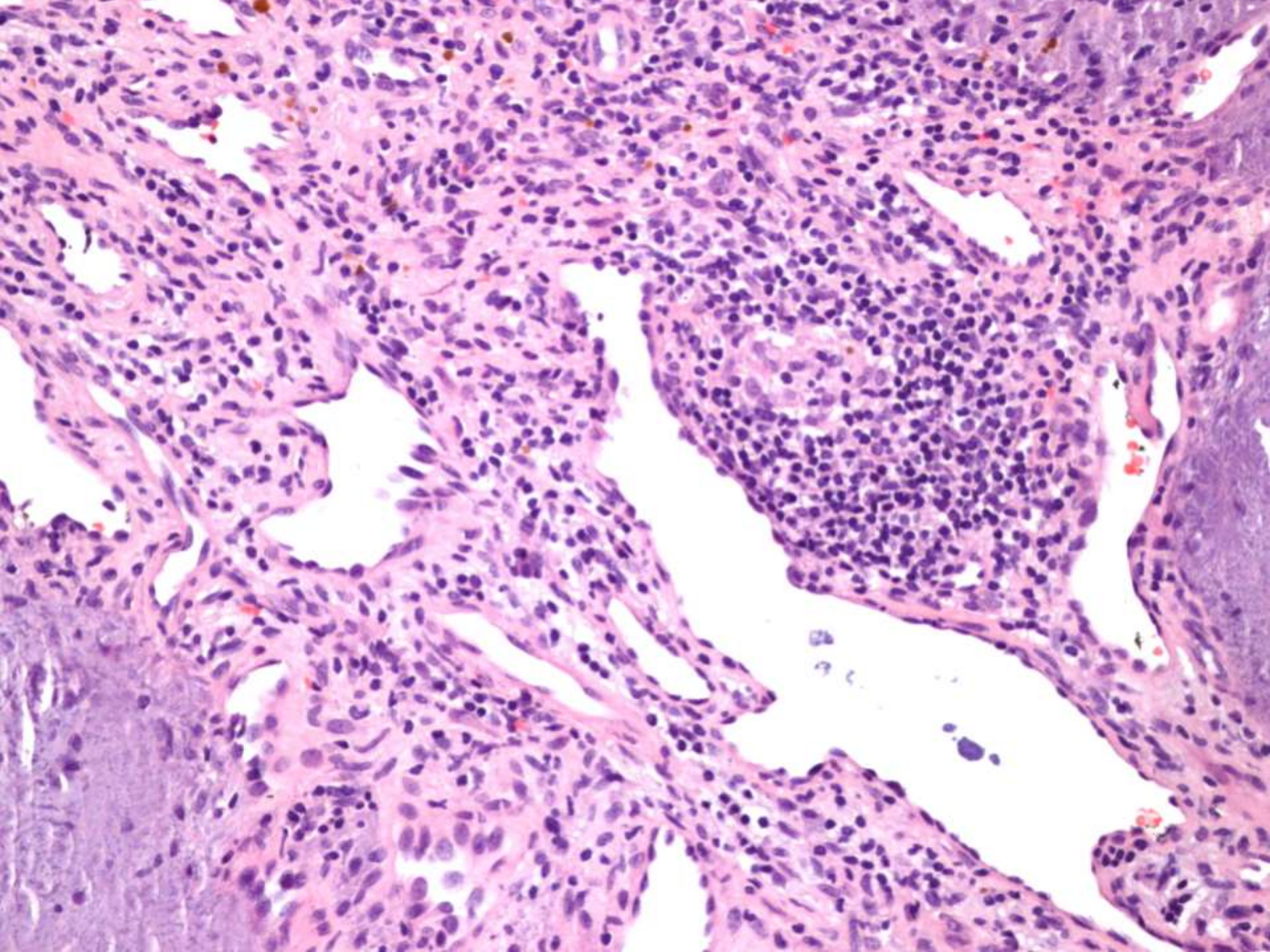


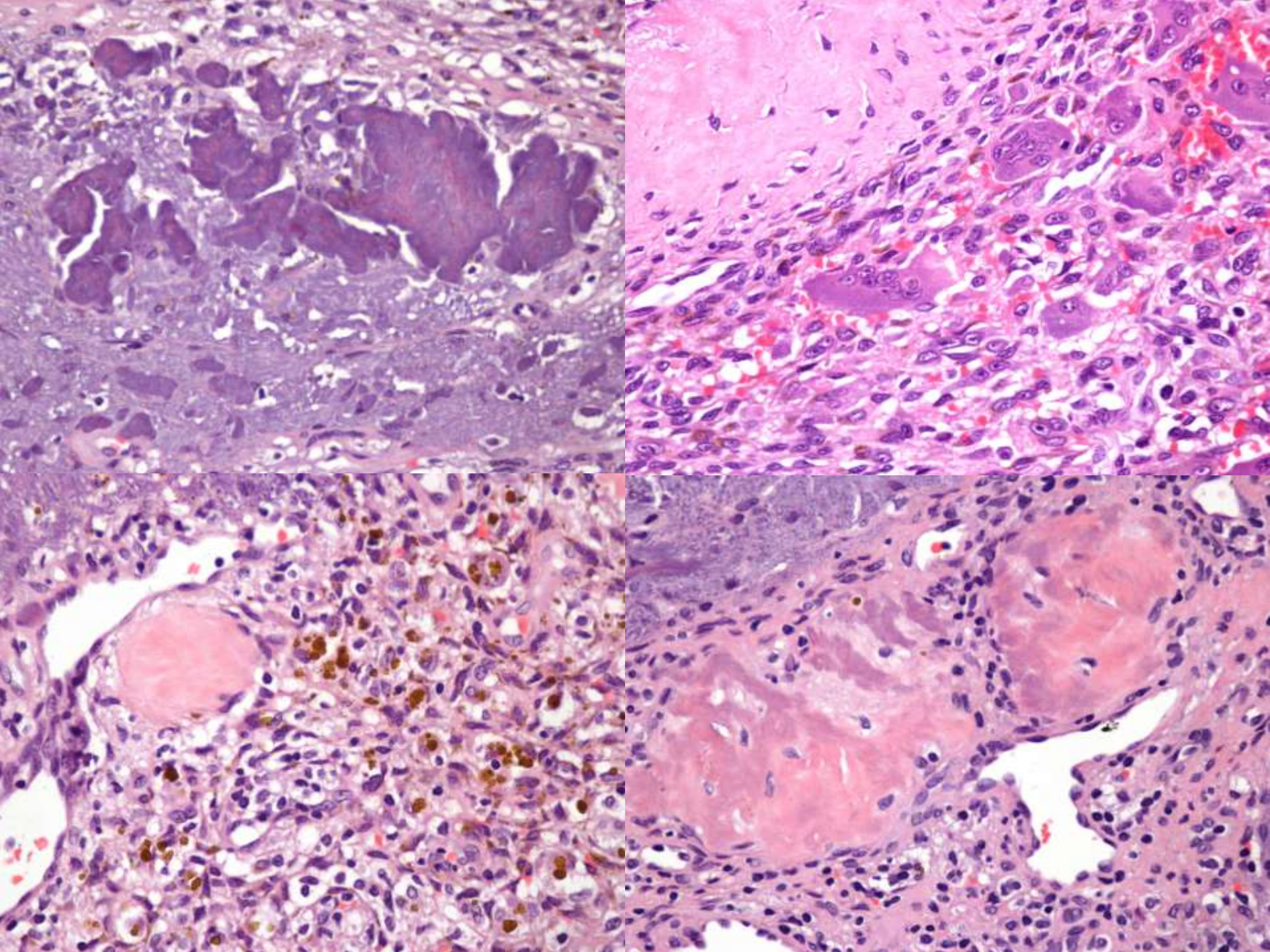


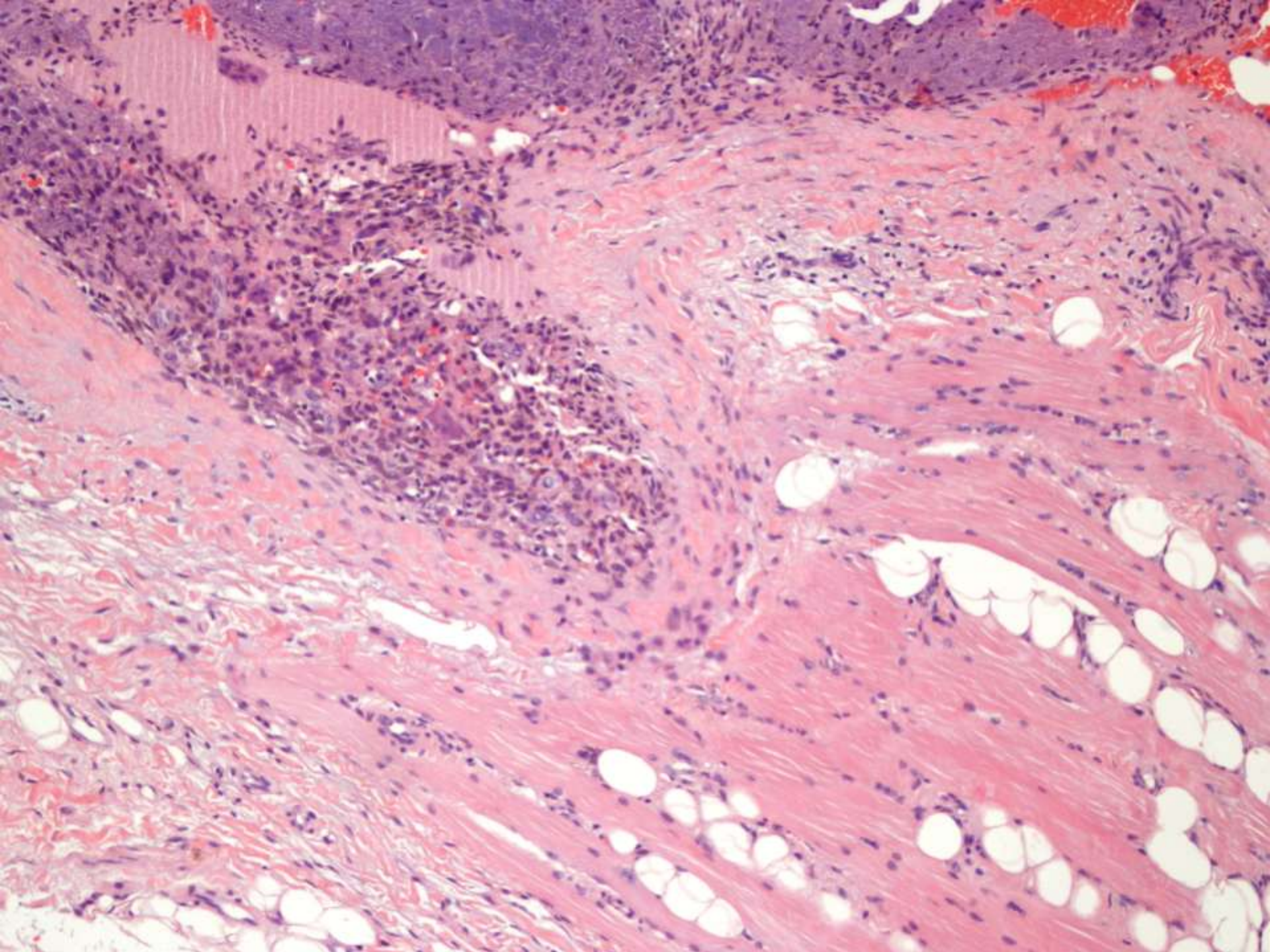












Diagnóstico

Tumor mesenquimal fosfatúrico variante mixta del tejido conectivo

TMF Seguimiento

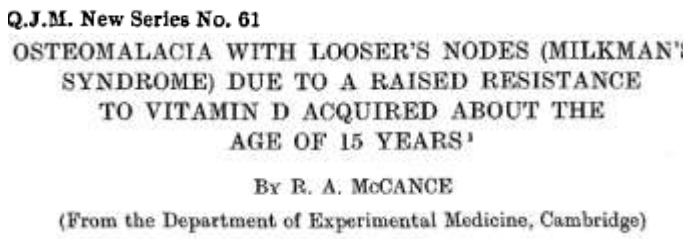
- **Antes de la cirugía:**
  Pemia: 1,9 mgr/dl (n: 2,5-4,5 mgr/dl)
- **Un mes después de la cirugía:**
  Pemia: 5,1 mgr/dl (n: 2,5-4,5 mgr/dl)
- **Tres meses después de la cirugía:**
  Pemia: 4,0 mgr/dl (n: 2,5-4,5 mgr/dl)
  Puria normal
  Fosfatasa alcalina normal
  Clínicamente bien dentro de la limitación
  RM: sin signos de persistencia/recidiva

Osteomalacia Oncogénica

- **Desmineralización ósea sistémica causada por una neoplasia**
- **Dolor y fracturas óseas**
- **Hiperfosfaturia**
- **Hipofosfatemia**
- **Descenso de 1,25-dihidroxivit D3 sérica**
- **Aumento de fosfatasa alcalina sérica**
- **Resistencia a vit D**

150 casos publicados

- Tumores óseos y partes blandas
- Neurofibromatosis-2
- Síndrome de McCune-Albright
- Displasia fibrosa poliostótica
- Síndrome del nevus epidérmico
- Carcinomas

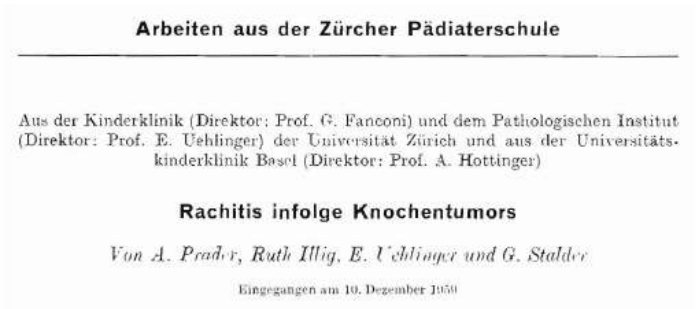


Q.J.M. New Series No. 61

OSTEOMALACIA WITH LOOSER'S NODES (MILKMAN'S SYNDROME) DUE TO A RAISED RESISTANCE TO VITAMIN D ACQUIRED ABOUT THE AGE OF 15 YEARS[1]

By R. A. McCANCE

(From the Department of Experimental Medicine, Cambridge)



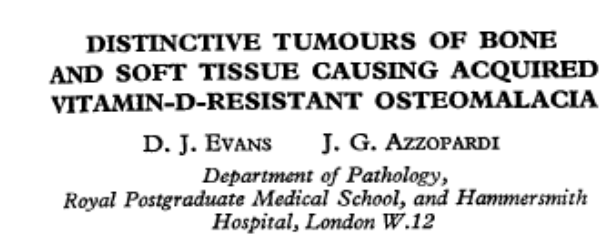
Arbeiten aus der Zürcher Pädiaterschule

Aus der Kinderklinik (Direktor: Prof. G. Fanconi) und dem Pathologischen Institut (Direktor: Prof. E. Uehlinger) der Universität Zürich und aus der Universitätskinderklinik Basel (Direktor: Prof. A. Hottinger)

Rachitis infolge Knochentumors

Von A. Prader, Ruth Illig, E. Uehlinger und G. Stalder

Eingegangen am 10. Dezember 1950

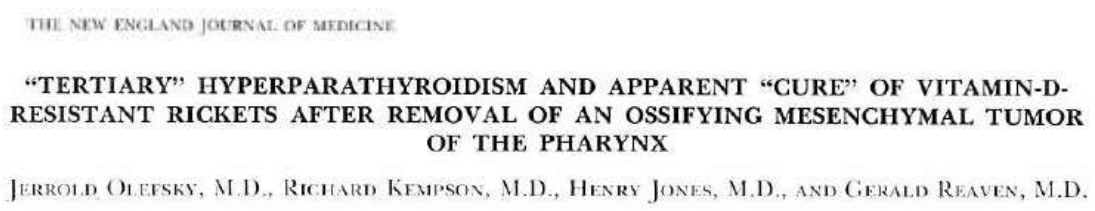


THE LANCET, FEBRUARY 12, 1972

DISTINCTIVE TUMOURS OF BONE AND SOFT TISSUE CAUSING ACQUIRED VITAMIN-D-RESISTANT OSTEOMALACIA

D. J. Evans J. G. Azzopardi

Department of Pathology, Royal Postgraduate Medical School, and Hammersmith Hospital, London W.12



THE NEW ENGLAND JOURNAL OF MEDICINE

"TERTIARY" HYPERPARATHYROIDISM AND APPARENT "CURE" OF VITAMIN-D-RESISTANT RICKETS AFTER REMOVAL OF AN OSSIFYING MESENCHYMAL TUMOR OF THE PHARYNX

Jerrold Olefsky, M.D., Richard Kempson, M.D., Henry Jones, M.D., and Gerald Reaven, M.D.

Phosphaturic Mesenchymal Tumors

A Polymorphous Group Causing Osteomalacia or Rickets

NOEL WEIDNER, MD,* AND DANIEL SANTA CRUZ, MD†

Reported are the pathologic features of 17 mesenchymal tumors documented as causing osteomalacia or rickets. Although these tumors were histologically polymorphous, they were classifiable into four morphological groups. In the first group there were ten unique tumors showing mixed connective tissue features and containing variably prominent vascular and/or osteoclast-like giant-cell components. Tumors of this group also displayed focal microcystic changes, osseous metaplasia, and/or poorly developed cartilaginous areas. The cartilaginous areas sometimes showed considerable dystrophic calcification. With one exception, all tumors of this group occurred in soft tissue and demonstrated benign clinical behavior. The single malignant tumor originated in bone, recurred locally, and metastasized to lung. The tumors comprising the remaining three groups (six tumors) occurred in bone, demonstrated benign clinical behavior, and were grouped according to their close resemblance to tumors known to occur in bone, that is osteoblastoma-like (four tumors), nonossifying fibroma-like (two tumors), and ossifying fibroma-like (one tumor).

***Cancer* 59:1442–1454, 1987.**

Tumor mesenquimal fosfatúrico

- Variante mixta del tejido conectivo
- Osteoblastoma-like
- Fibroma no osificante-like
- Fibroma osificante-like

Tumor mesenquimal fosfatúrico variante mixta del tejido conectivo

- **Células mesenquimales pequeñas, redondas o fusiformes, en sábanas pobremente definidas**
- **Células osteoclasto-like asociadas a hemorragia**
- **Prominente vascularización con patrón hemangiopericitoide focal**
- **Estroma pseudocartilaginoso con calcificaciones y focos de osteoide y hueso**
- **Áreas microquísticas**

Most Osteomalacia-associated Mesenchymal Tumors Are a Single Histopathologic Entity

An Analysis of 32 Cases and a Comprehensive Review of the Literature

Andrew L. Folpe, MD, Julie C. Fanburg-Smith, MD,† Steven D. Billings, MD,‡ Michele Bisceglia, MD,§ Franco Bertoni, MD,¶ Justin Y. Cho, BS,‖ Michael J. Econs, MD,** Carrie Y. Inwards, MD,†† Suzanne M. Jan de Beur, MD,‖ Thomas Mentzel, MD,‡‡ Elizabeth Montgomery, MD,§§ Michal Michal, MD,¶¶ Markku Miettinen, MD,† Stacey E. Mills, MD, [a] John D. Reith, MD,‖‖ John X. O'Connell, MD,*** Andrew E. Rosenberg, MD,††† Brian P. Rubin, MD, PhD,‡‡‡ Donald E. Sweet, MD,§§§ Tuyethoa N. Vinh, MD,§§§ Lester E. Wold, MD,†† Brett M. Wehrli, MD,¶¶¶ Kenneth E. White, PhD,** Richard J. Zaino, MD,‖‖ and Sharon W. Weiss, MD**

Tumores:

- **18 de partes blandas**
- **9 óseos**
- **2 de senos paranasales**
- **3 sin osteomalacia**

Diagnóstico:

- **24 TMFMTC (incluyendo los 3 sin osteomalacia osteogénica)**
- **3 TMFMTC malignos**
- **1 hemangiopericitoma con cells osteoclasto-like**
- **1 encondroma atípico**
- **1 osteosarcoma esclerosante**
- **2 tumores hemangiopericitoma-like paranasales**

Revisan 109 casos de osteomalacia osteogénica publicados:

- **75 (69%) TMFMTC o variantes**
- **7 (6%): HPC-like paranasal (2), osteosarcoma (2), HPC, hemangioma óseo, fibroma no osificante**
- **27 (25%): datos insuficientes**

Osteomalacia Oncogénica

Expresión de FGF-23 en TMF

Cloning and characterization of FGF23 as a causative factor of tumor-induced osteomalacia

Takashi Shimada*, Satoru Mizutani†, Takanori Muto*, Takashi Yoneya*, Rieko Hino*, Shu Takeda‡§, Yasuhiro Takeuchi‡, Toshiro Fujita‡, Seiji Fukumoto¶, and Takeyoshi Yamashita*‖

*Pharmaceutical Research Laboratory, Nephrology, Kirin Brewery Co. Ltd., 3 Miyahara, Takasaki, Gunma 370-1295, Japan; †Central Laboratories for Key Technology, Kirin Brewery Co. Ltd., 1-13-5 Fukuura, Kanazawa-ku, Yokohama, Kanagawa 236-0004, Japan; and ‡Division of Endocrinology, Department of Medicine, University of Tokyo School of Medicine, and ¶Department of Laboratory Medicine, University of Tokyo Branch Hospital, 3-28-6 Mejirodai, Bunkyo-ku, Tokyo 112-8688, Japan

European Journal of Endocrinology (2003) **148** 269–276 ISSN 0804-4643

EXPERIMENTAL STUDY

Immunohistochemical detection of FGF-23 protein in tumors that cause oncogenic osteomalacia

Tobias Larsson[1,3], Richard Zahradnik[2], Jeffrey Lavigne[2], Östen Ljunggren[1], Harald Jüppner[3] and Kenneth B Jonsson[1,3]

[1] *Department of Medical Sciences, University Hospital, Uppsala, Sweden,* [2] *Immutopics Inc., 929 Calle Negocio, Suite A, San Clemente, California 92673, USA and* [3] *Endocrine Unit, Massachusetts General Hospital and Harvard Medical School, Boston, Massachusetts 02114, USA*

(Correspondence should be addressed to K B Jonsson, Department of Medical Sciences, University Hospital, SE-751 85 Uppsala, Sweden; Email: Kenneth.Jonsson@medsci.uu.se)

RT-PCR Analysis for FGF23 Using Paraffin Sections in the Diagnosis of Phosphaturic Mesenchymal Tumors With and Without Known Tumor Induced Osteomalacia

Armita Bahrami, MD, Sharon W. Weiss, MD,† Elizabeth Montgomery, MD,‡ Andrew E. Horvai, MD, PhD,§ Long Jin, MD,* Carrie Y. Inwards, MD,* and Andrew L. Folpe, MD**

Abstract: Phosphaturic mesenchymal tumors of the mixed connective tissue type (PMTMCT) are extremely rare, histologically distinctive neoplasms, which cause tumor-induced osteomalacia (TIO) in most cases through the elaboration of a

Key Words: tumor-induced osteomalacia, FGF23, reverse transcription polymerase chain reaction, phosphaturic mesenchymal tumor

(*Am J Surg Pathol* 2009;33:1348–1354)

European Journal of Endocrinology (2008) **158** 431–437 ISSN 0804-4643

CASE REPORT

Tumor producing fibroblast growth factor 23 localized by two-staged venous sampling

Gerben van Boekel[1], Janneke Ruinemans-Koerts[2], Frank Joosten[3], Paul Dijkhuizen[4], Adriaan van Sorge[5] and Hans de Boer[1]

Departments of [1]*Internal Medicine,* [2]*Clinical Chemistry,* [3]*Radiology,* [4]*Gynecology and* [5]*Clinical Pharmacy, Ziekenhuis Rijnstate, Wagnerlaan 55, 6800 TA, Arnhem, The Netherlands*

(Correspondence should be addressed to H de Boer; Email: hdeboer@dijnis.nl)

Tumor Mesenquimal Fosfatúrico

- **Extremadamente infrecuente**
- **Adultos edad media (3 m – 73 a)**
- **Larga historia osteomalacia vit D resistente**
- **Localizados en partes blandas (47%), hueso (47%), senos paranasales (5%)**
- **La resección del tumor produce corrección de la hiperfosfaturia y de la hipofosfatemia y aumento de la mineralización ósea**
- **Histológicamente reconocibles aun en ausencia de osteomalacia**

TMF: Bibliografía

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