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SARCOMA ALVEOLAR DE PARTES BLANDAS

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SéAP-IAP

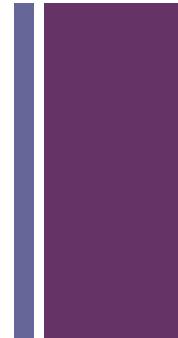


SEAP





CASO



- Una niña de 13 años acude a consulta por presentar un progresivo aumento de volumen, de unos 8 meses de evolución, en el brazo derecho
- La RMN reveló una tumoración de 3 x 2 cm localizada en el músculo bíceps derecho sin afectación de planos óseos
- El estudio de extensión descartó la presencia de enfermedad metastásica

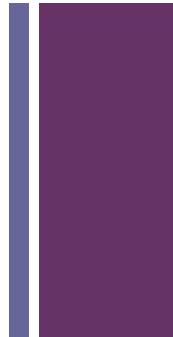
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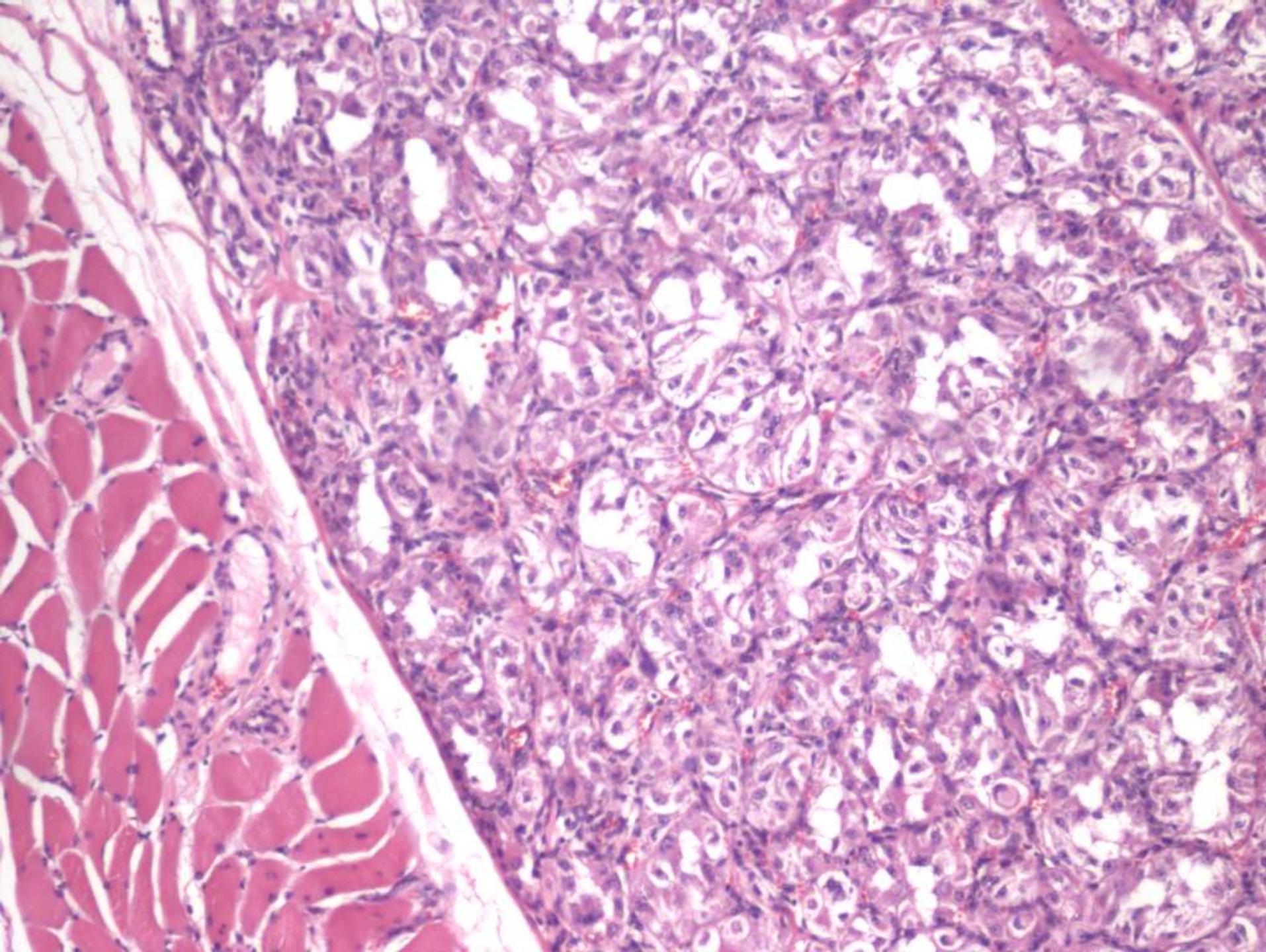
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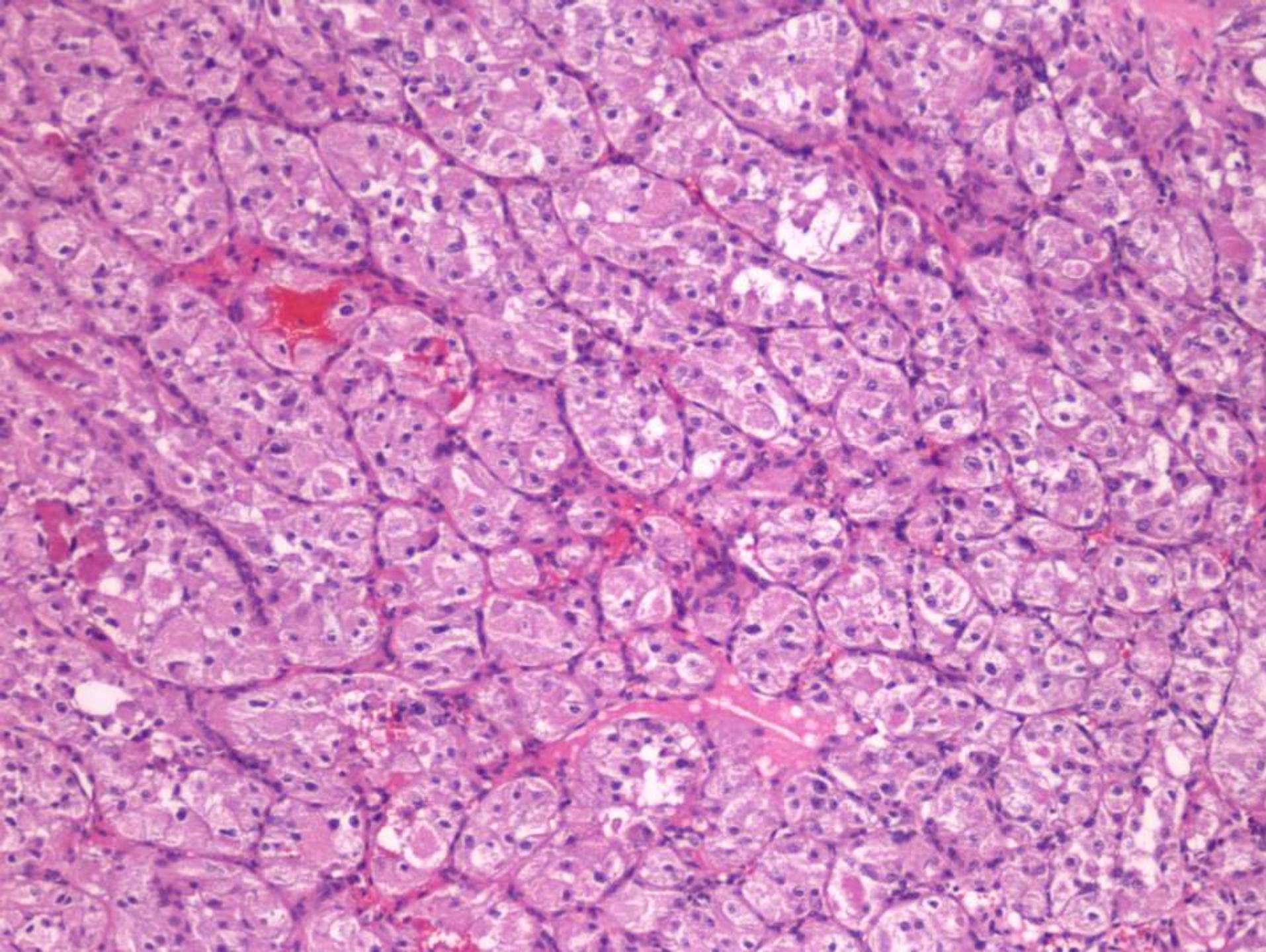


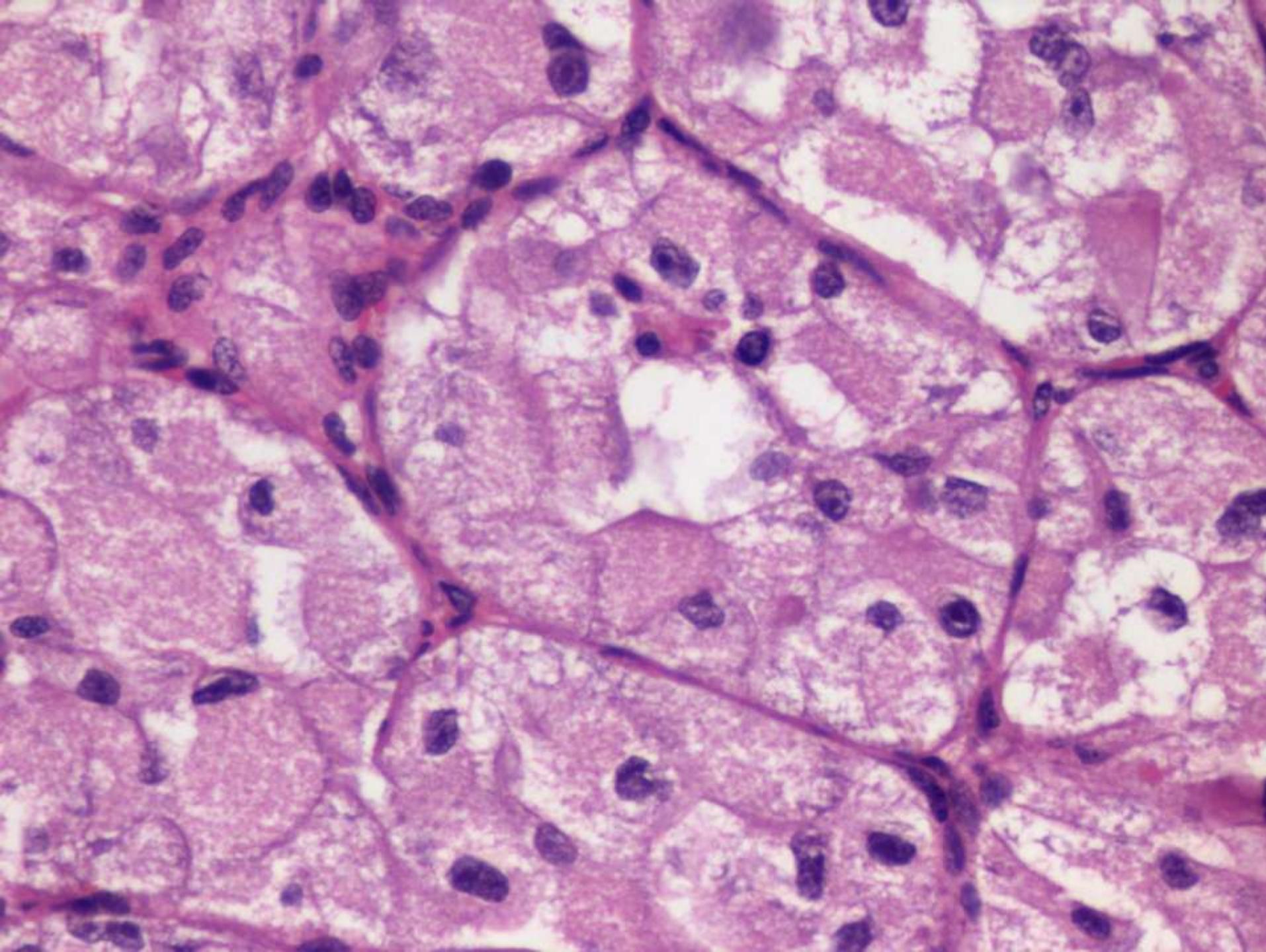
CASO

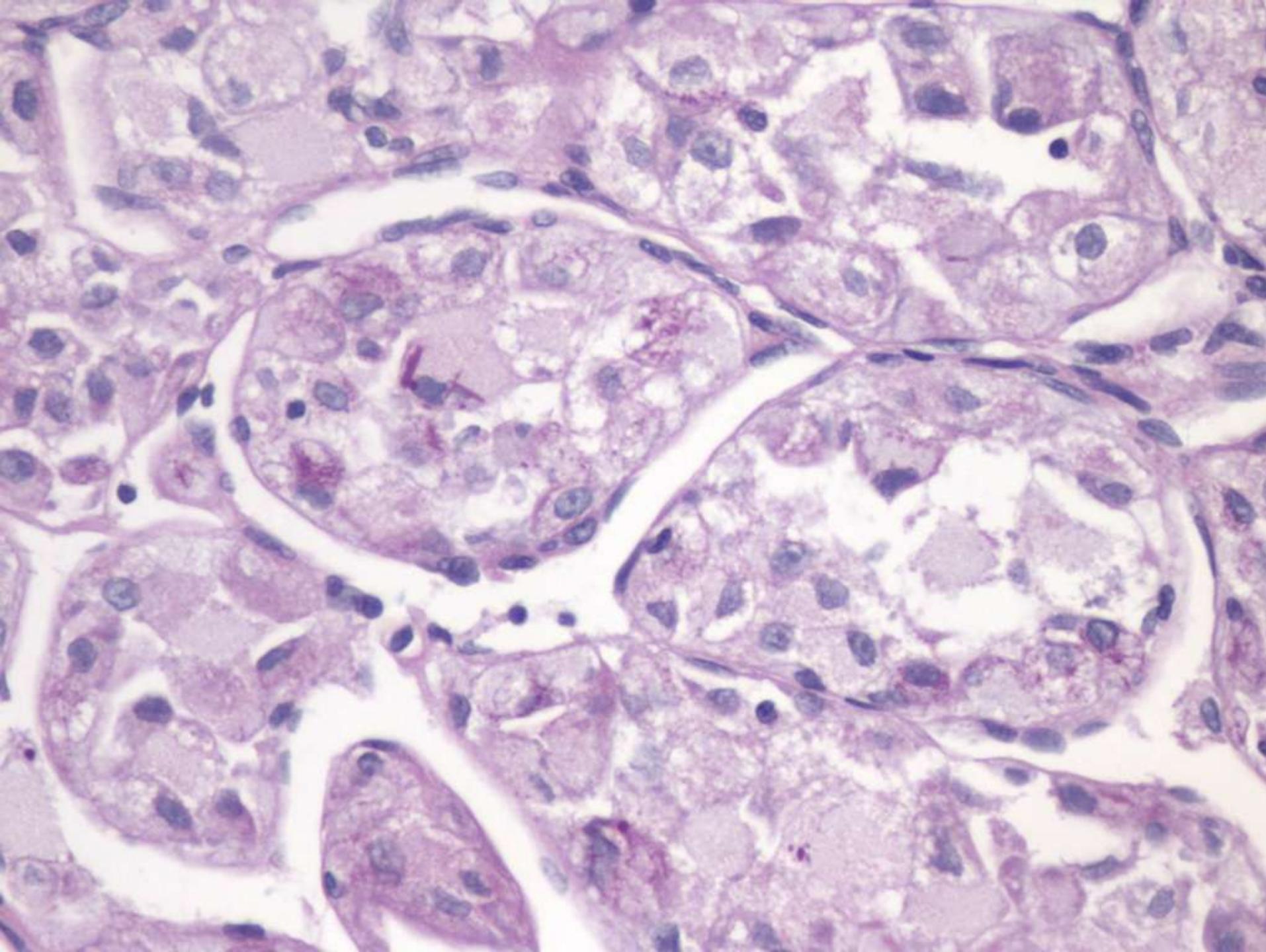


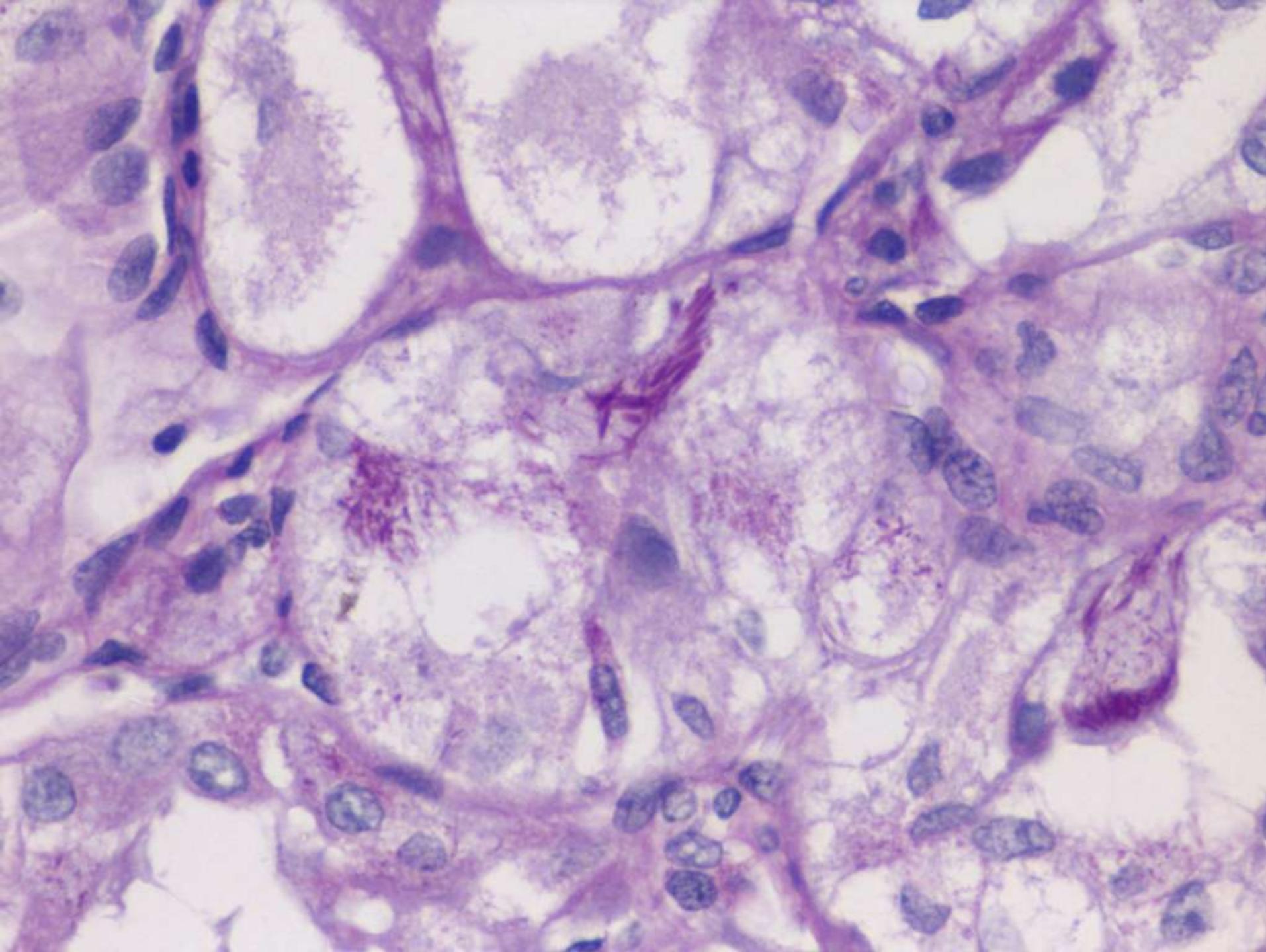
- Se realizó PAAF de la lesión con posterior extirpación quirúrgica con márgenes libres
- Los hallazgos citológicos fueron los típicos del Sarcoma Alveolar de Partes Blandas
- La histología confirmó el diagnóstico

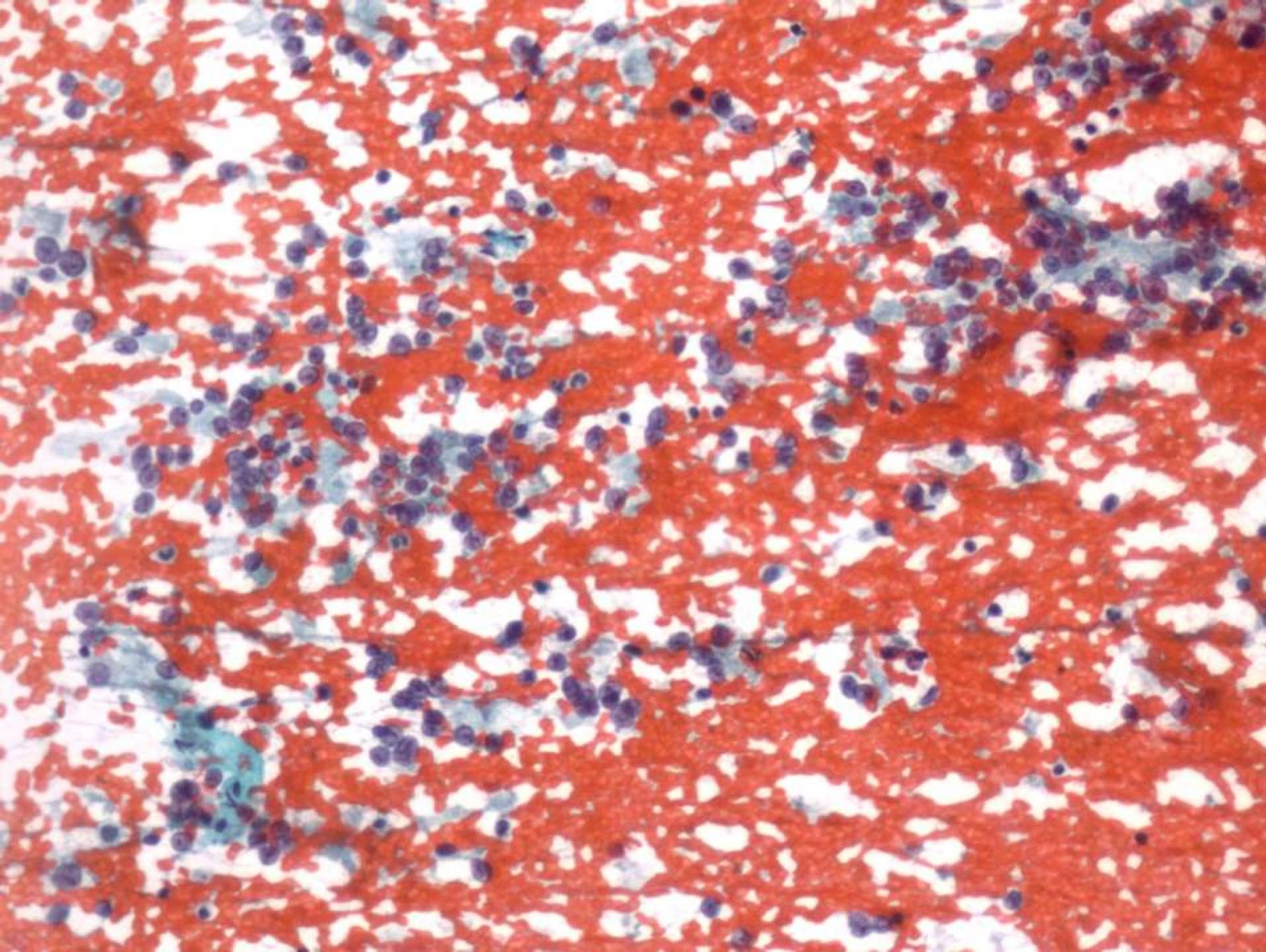


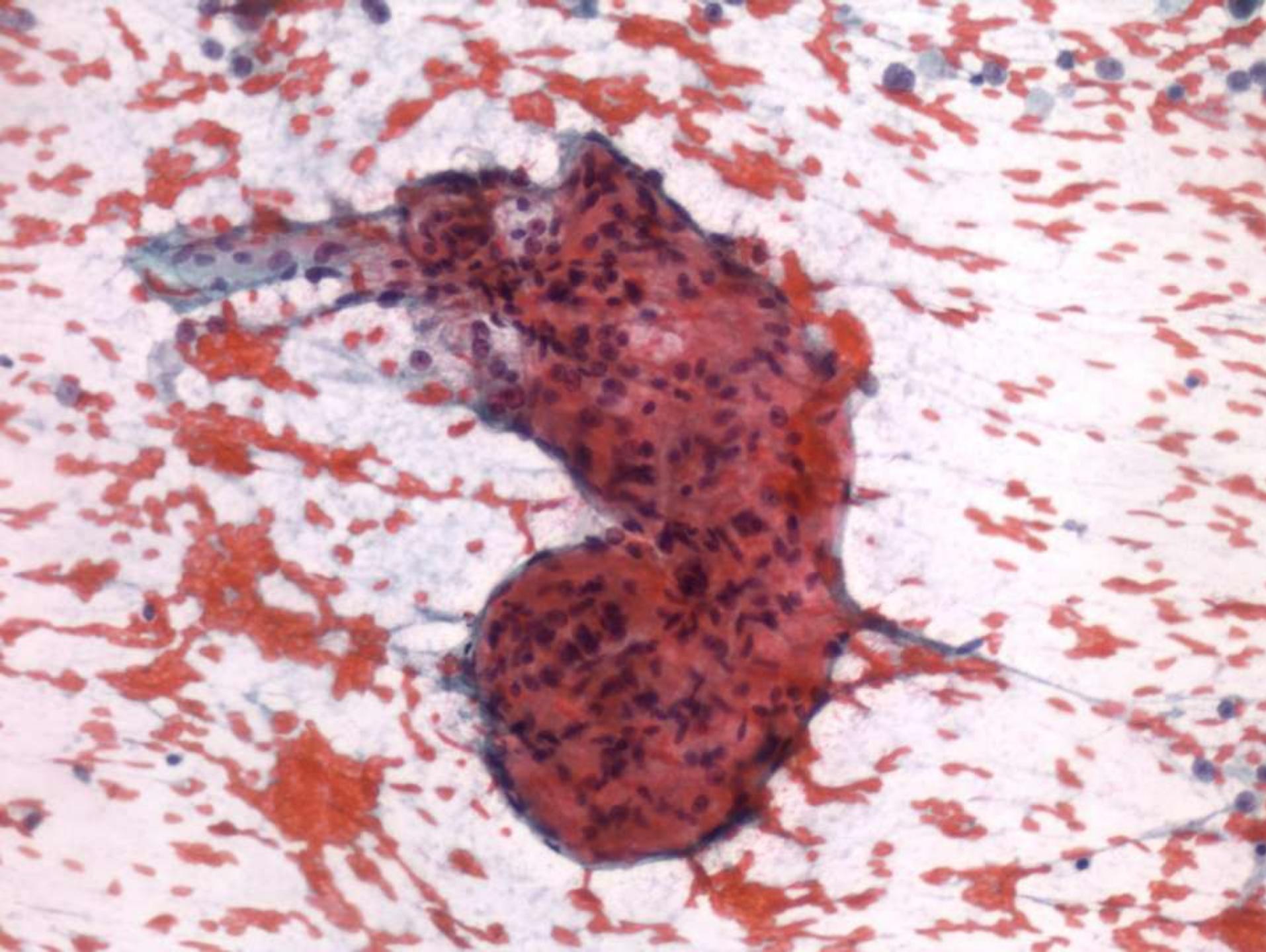


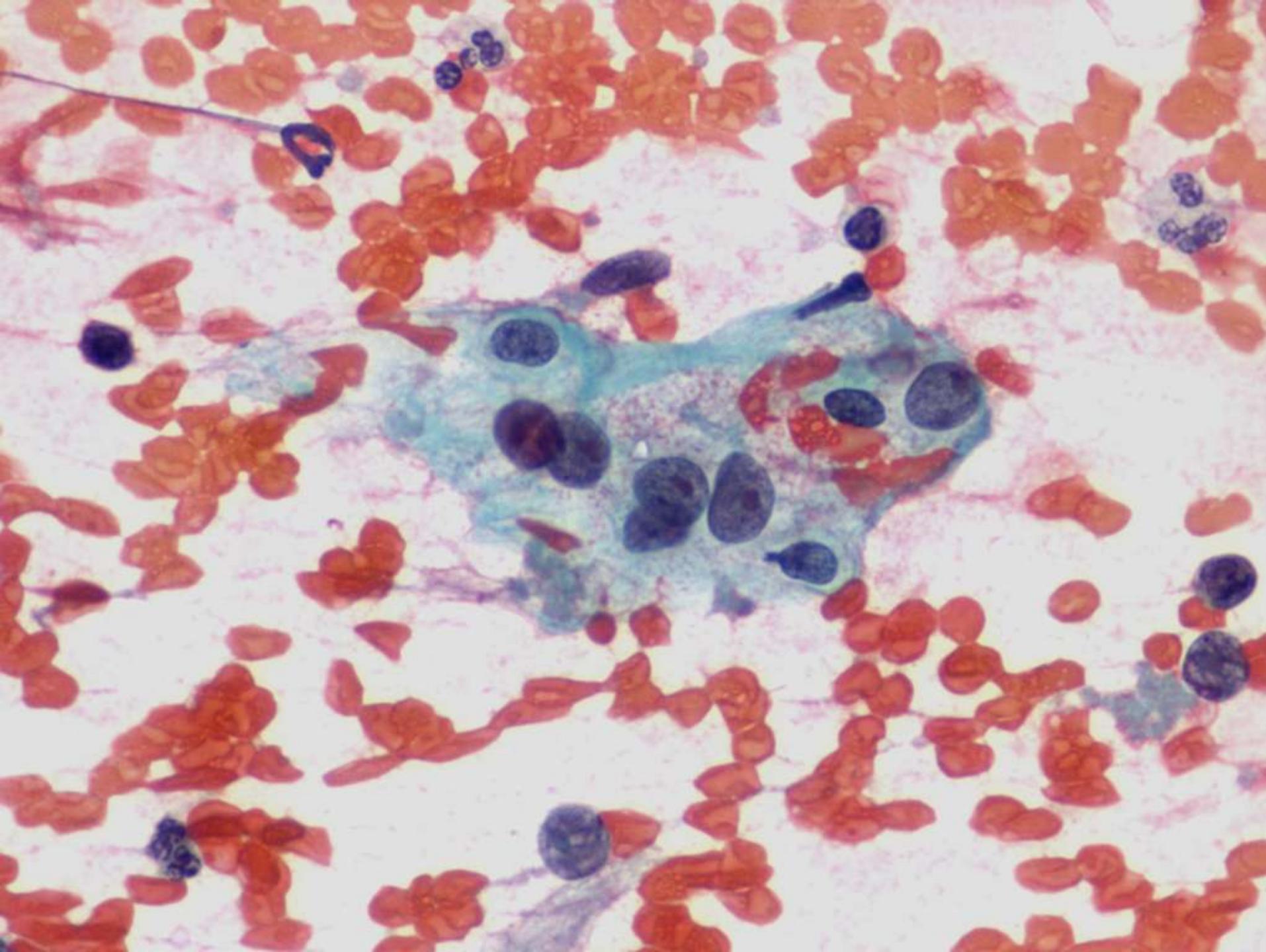


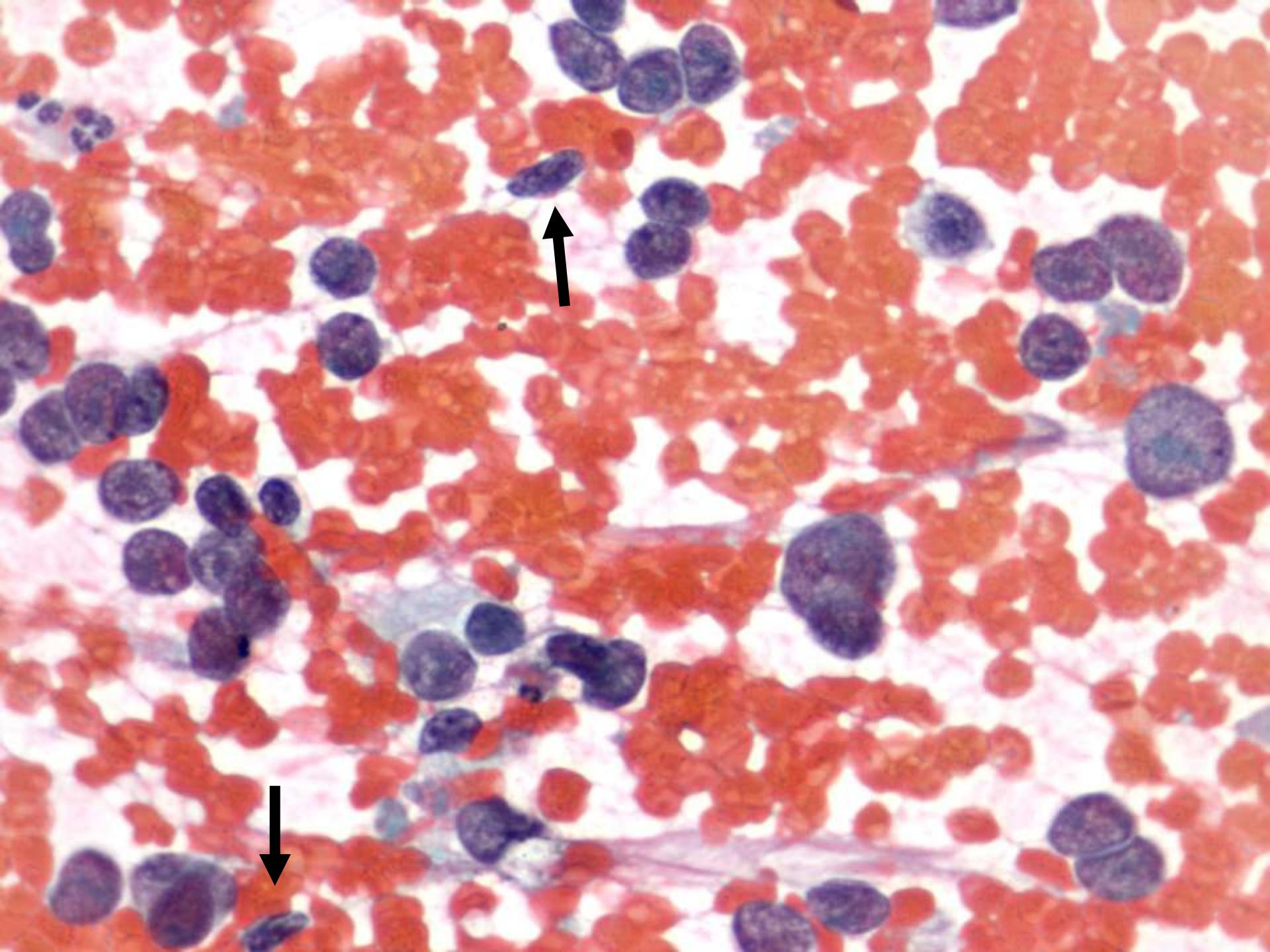


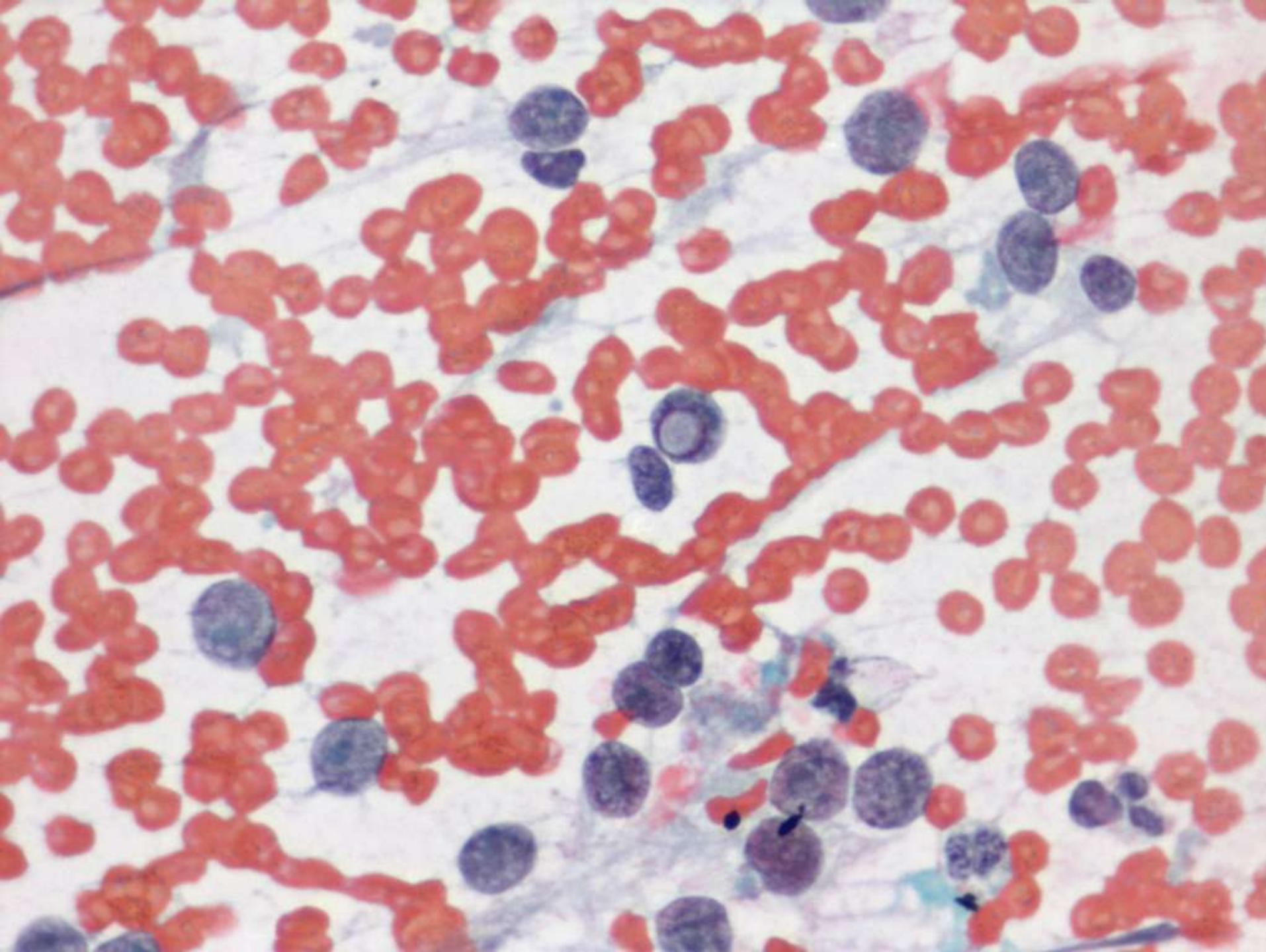


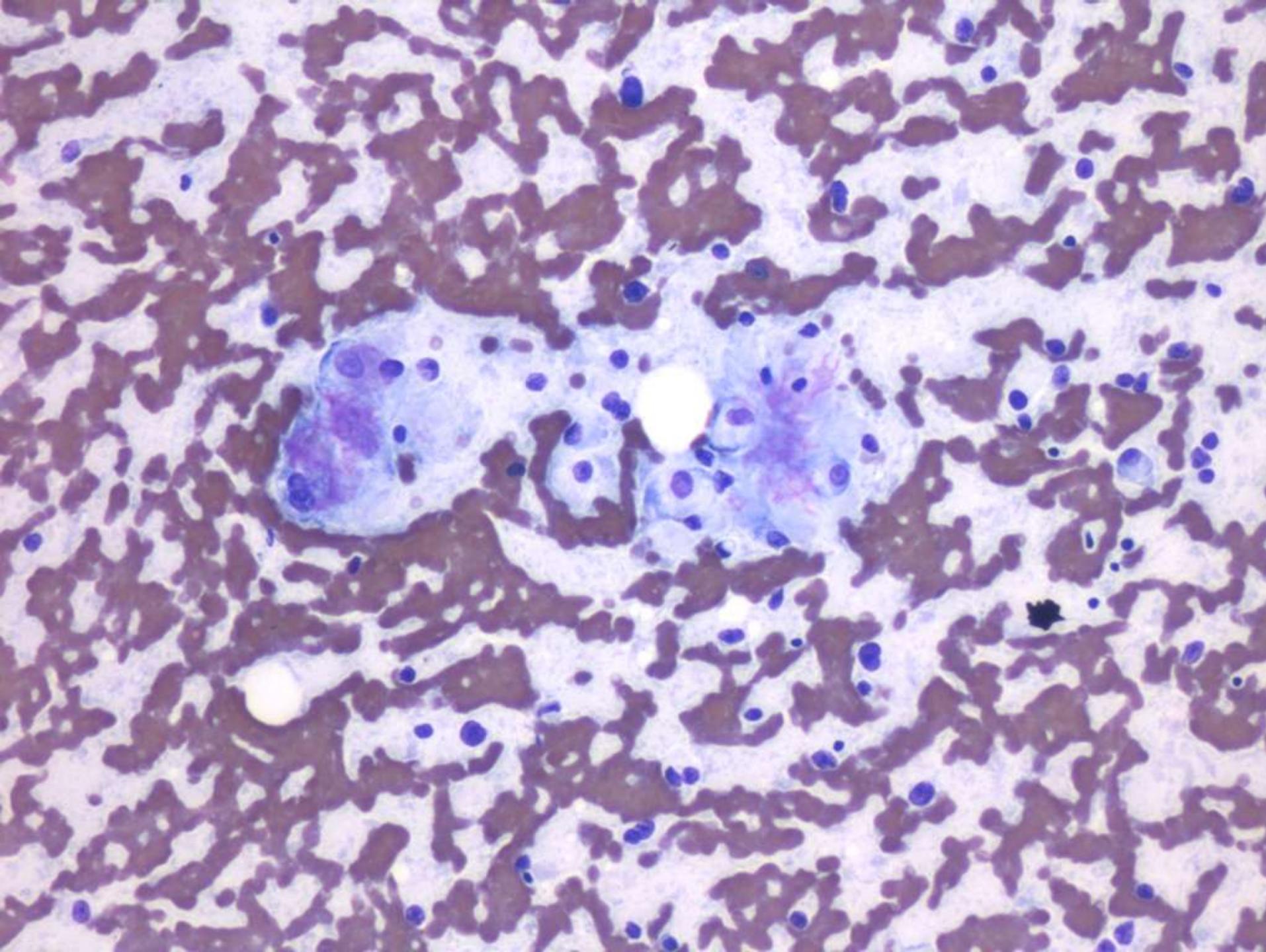


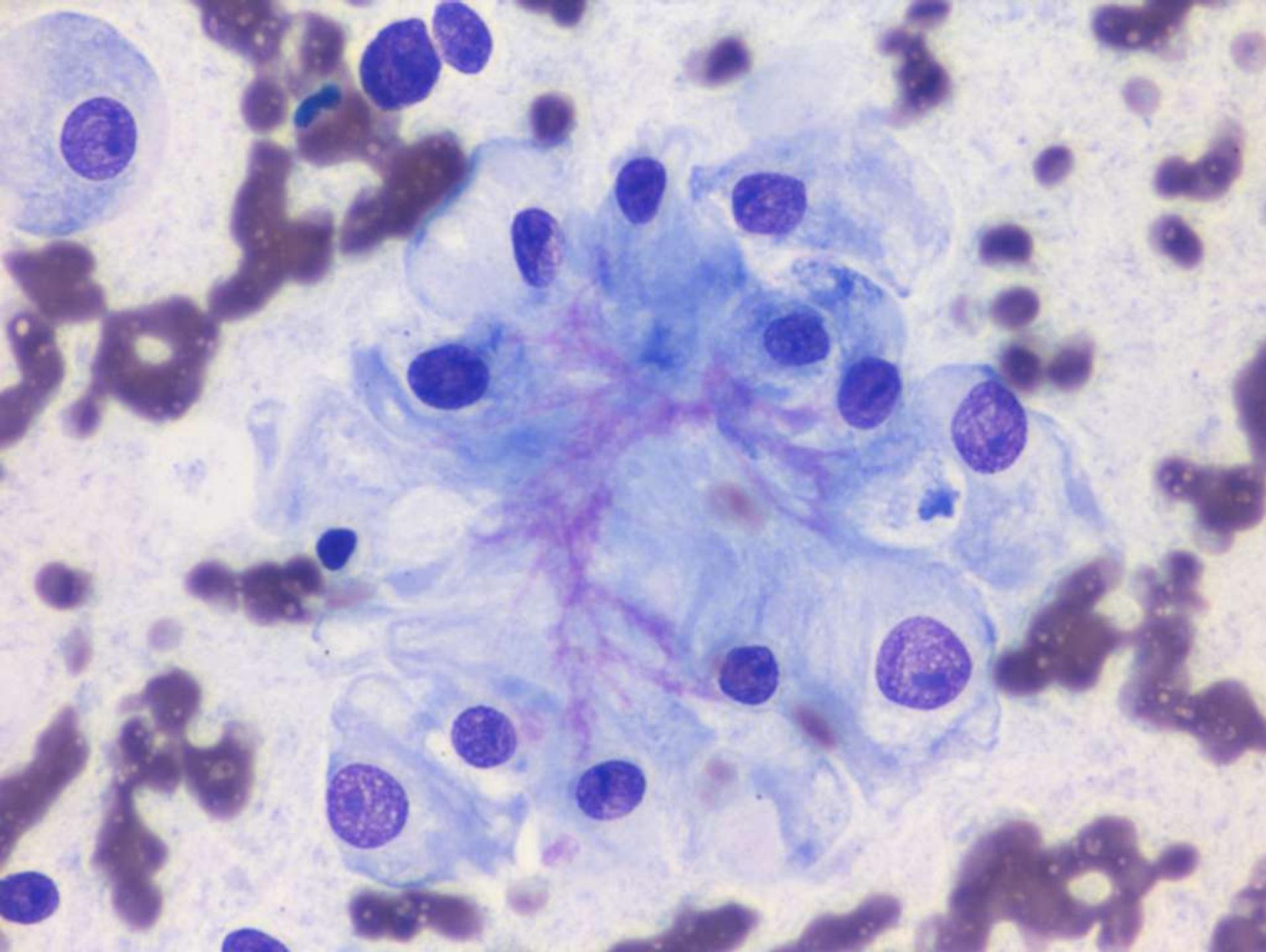


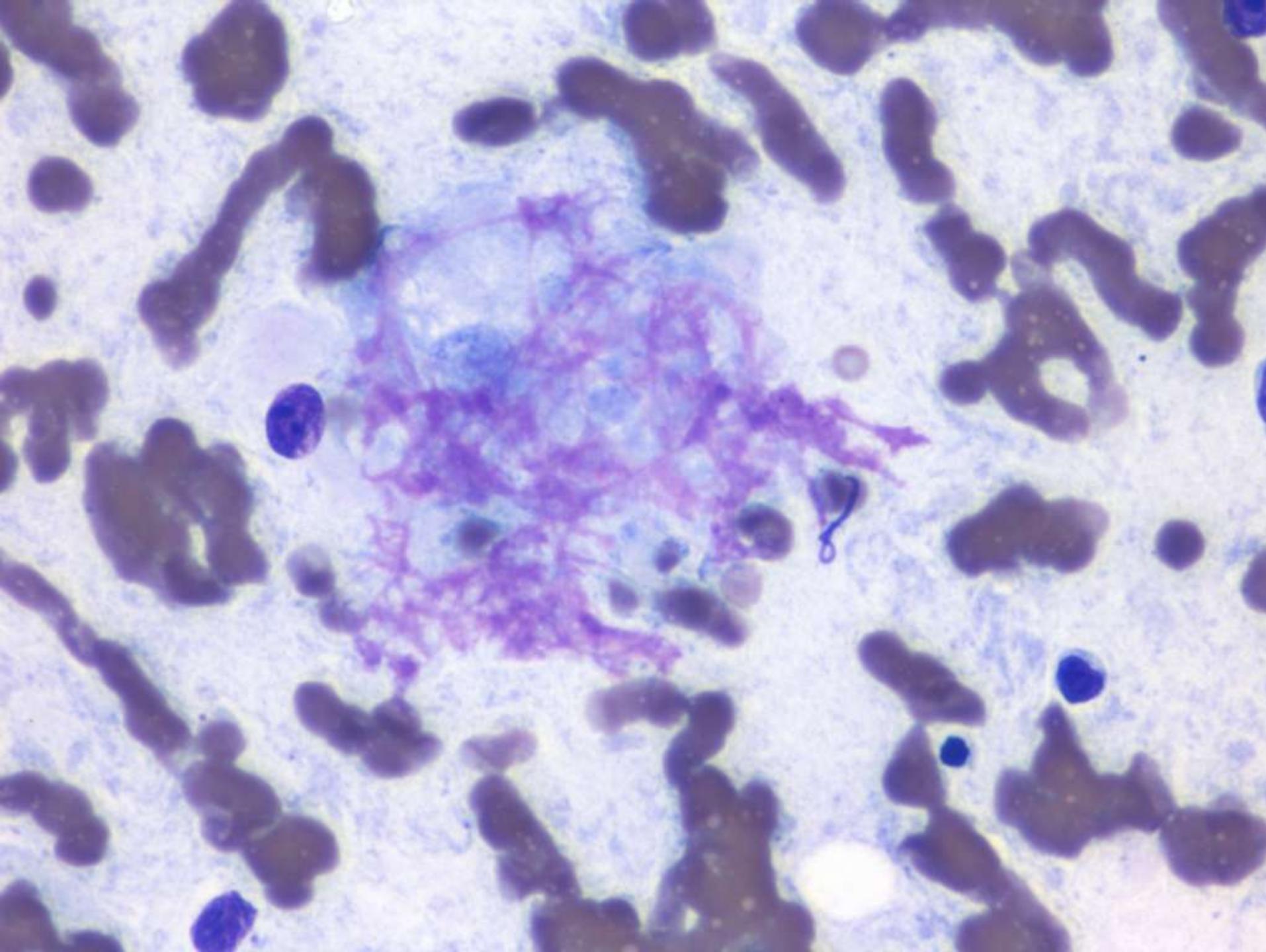


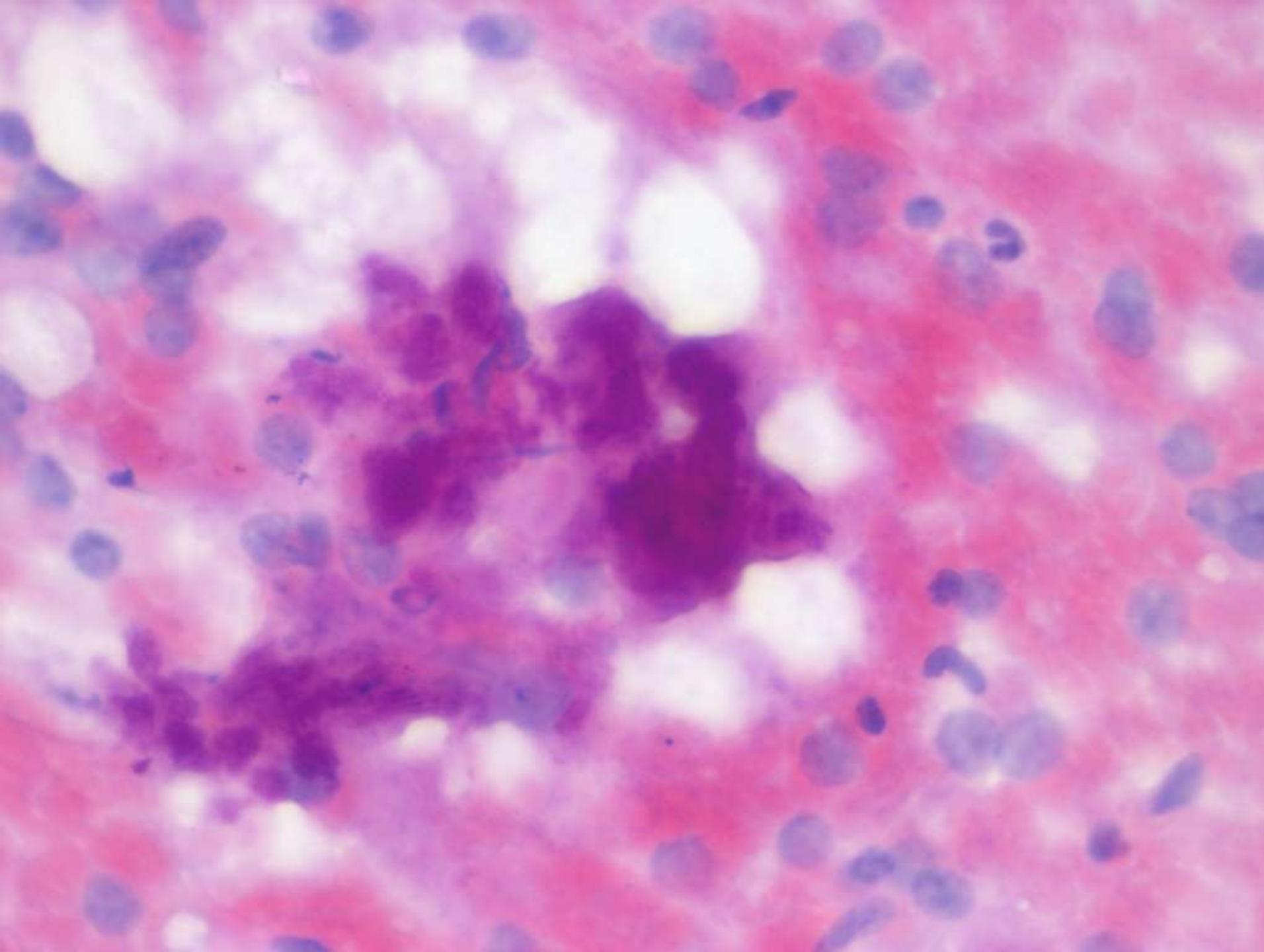




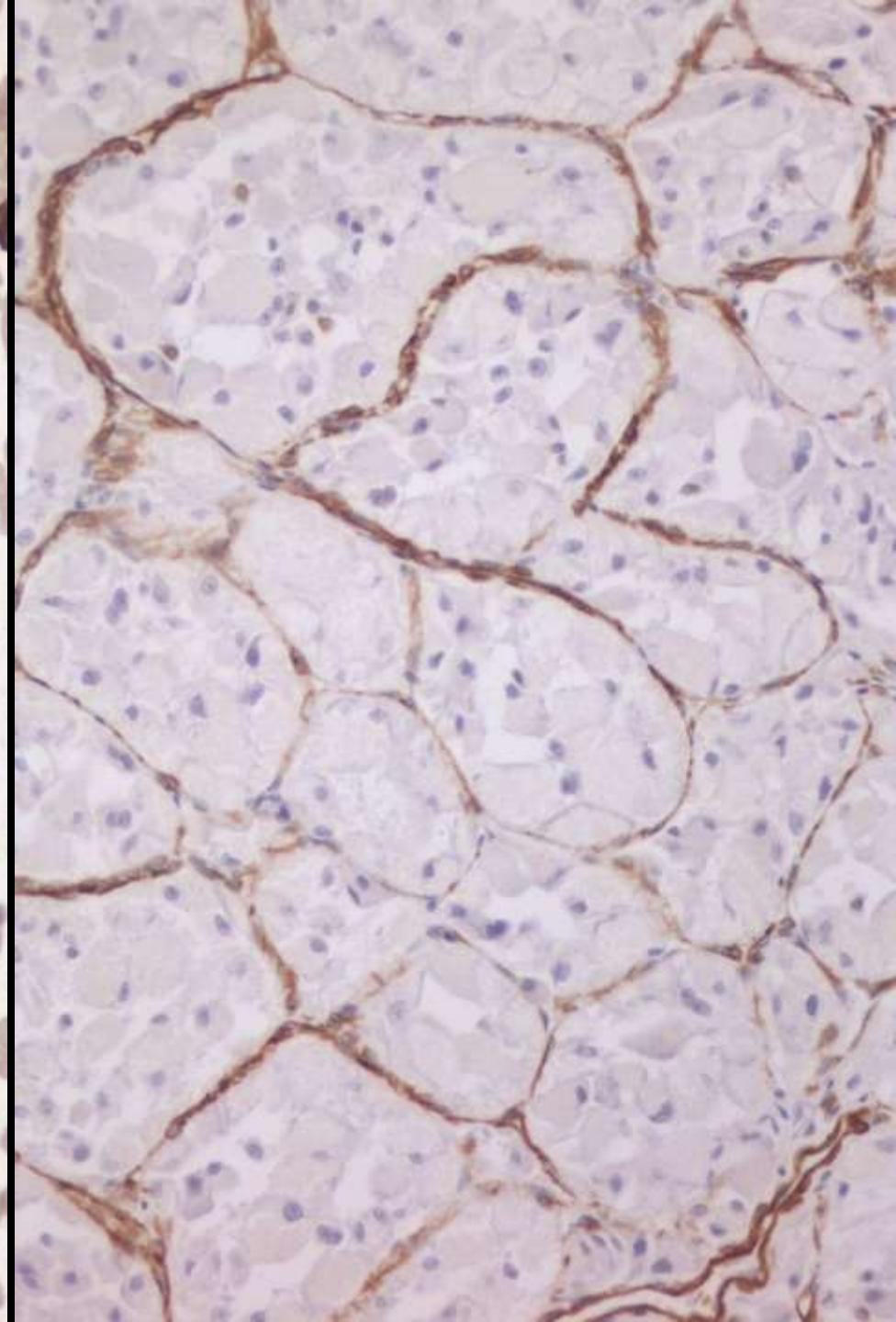
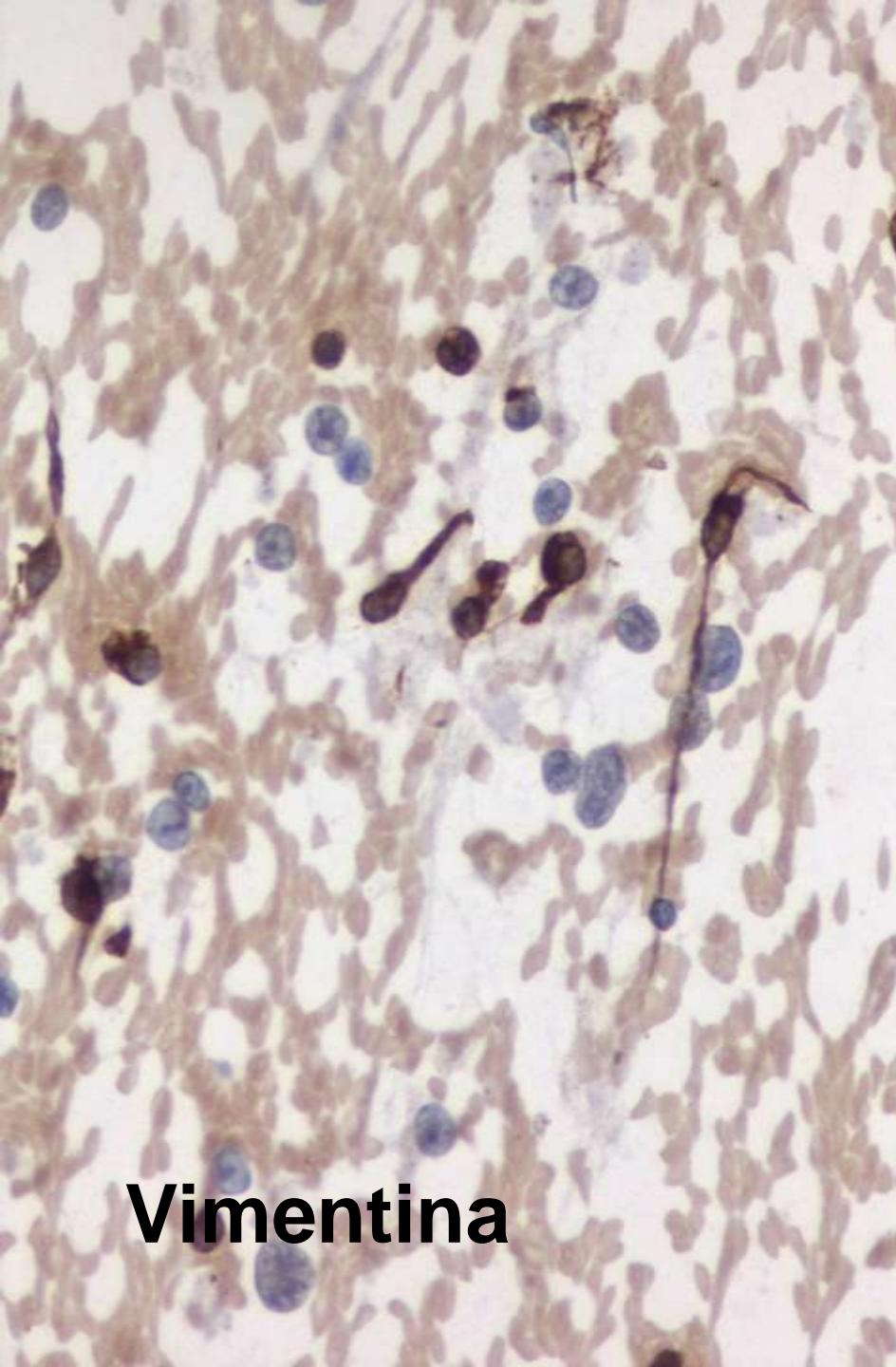




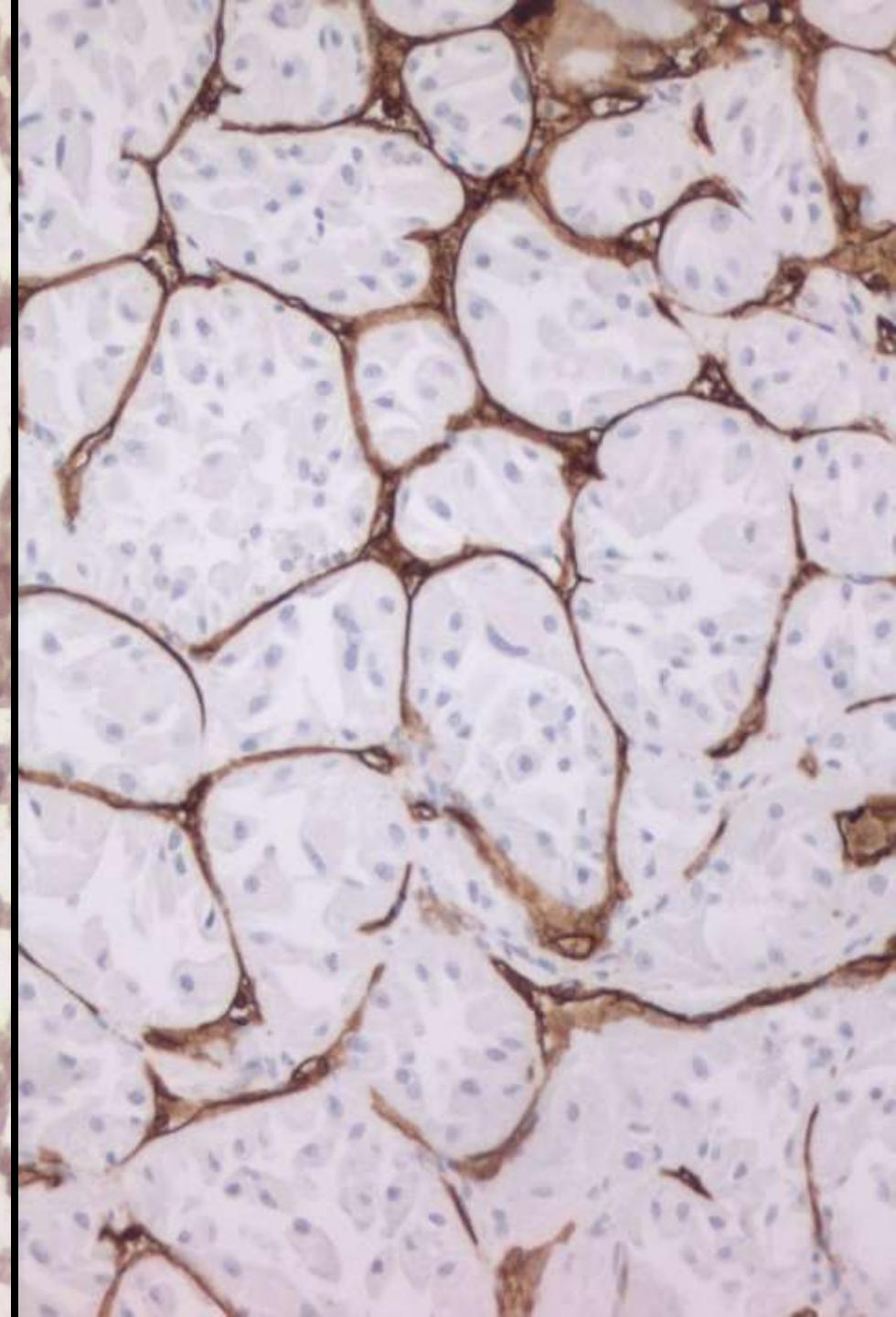
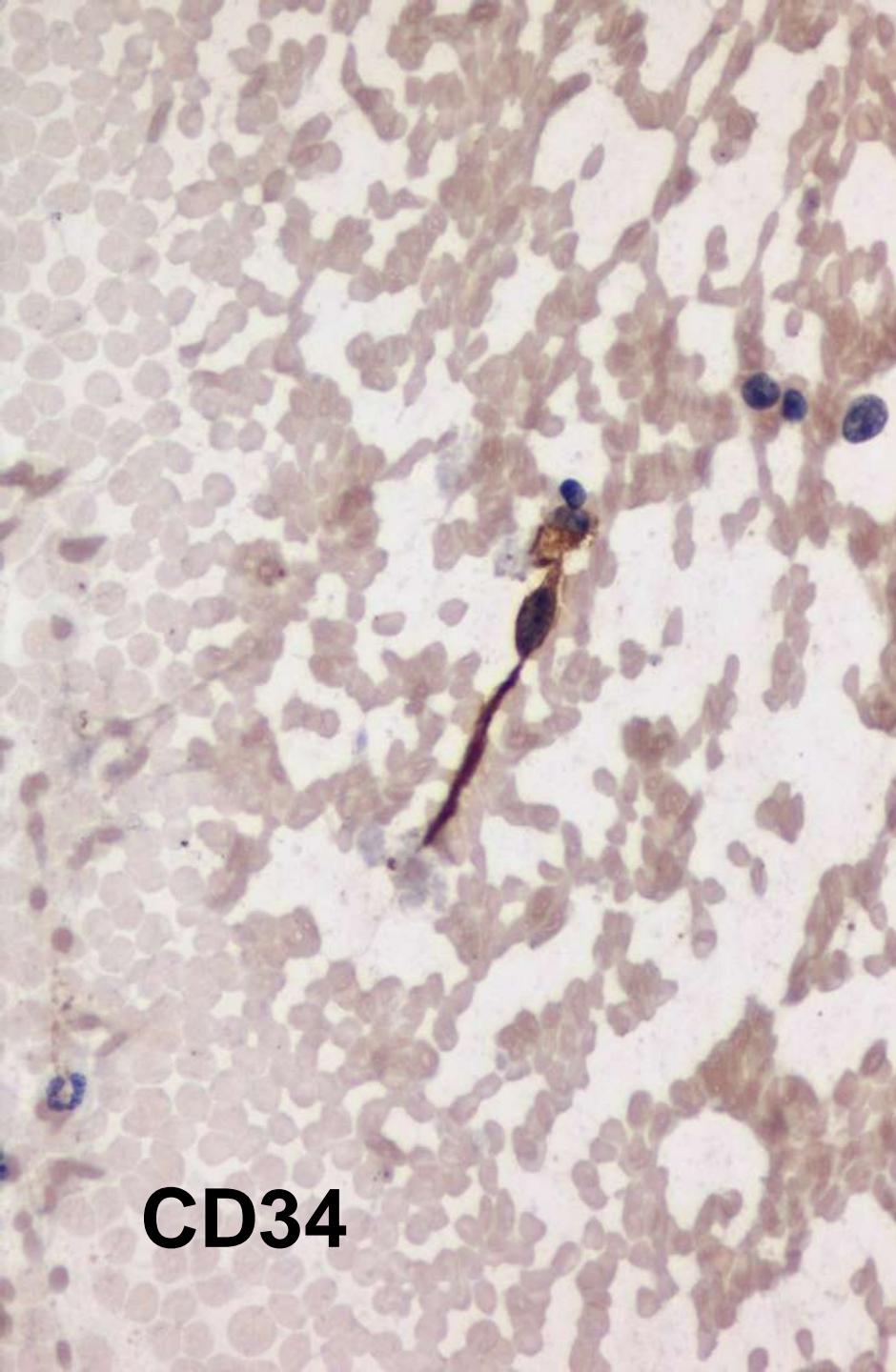


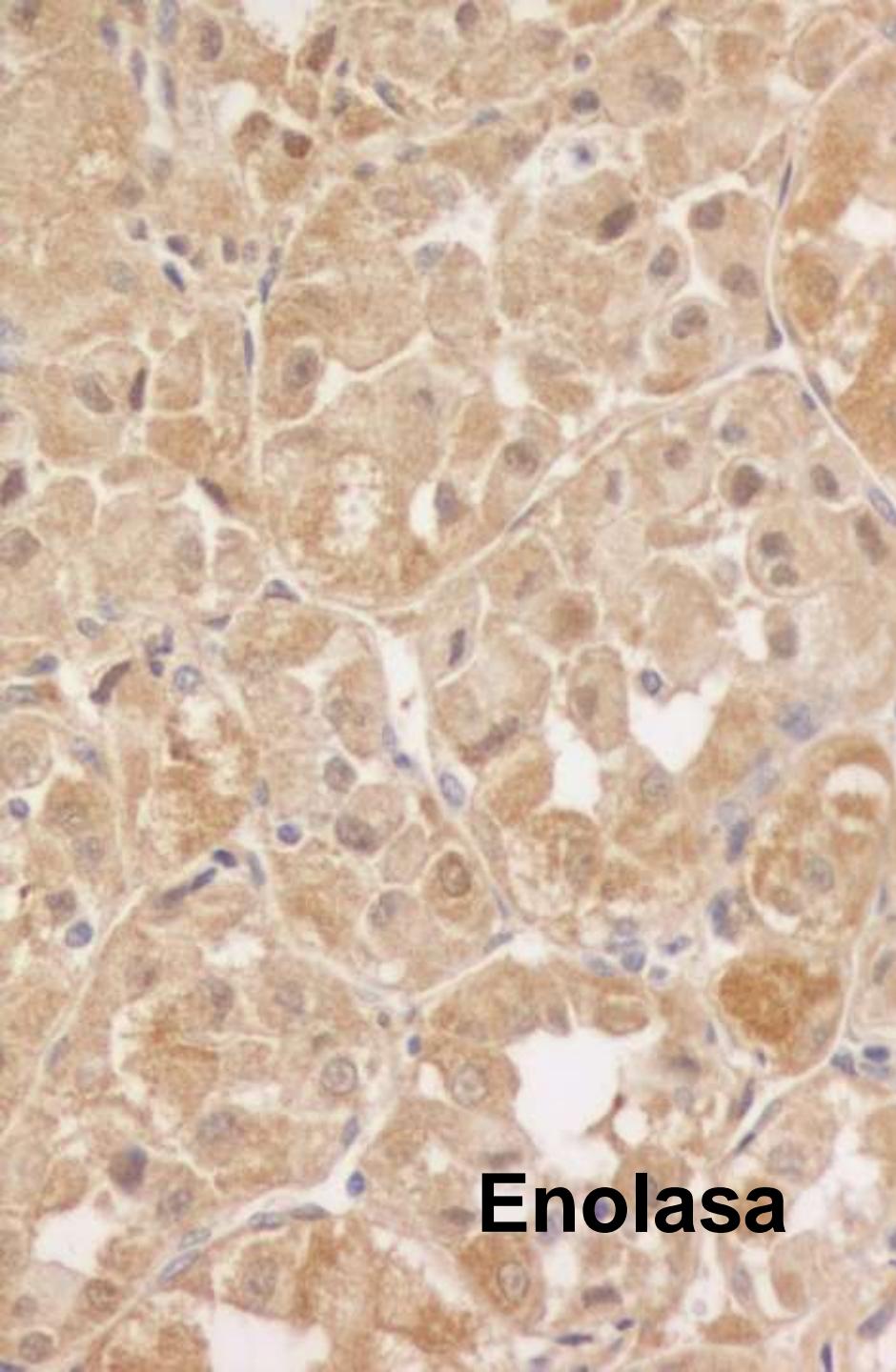


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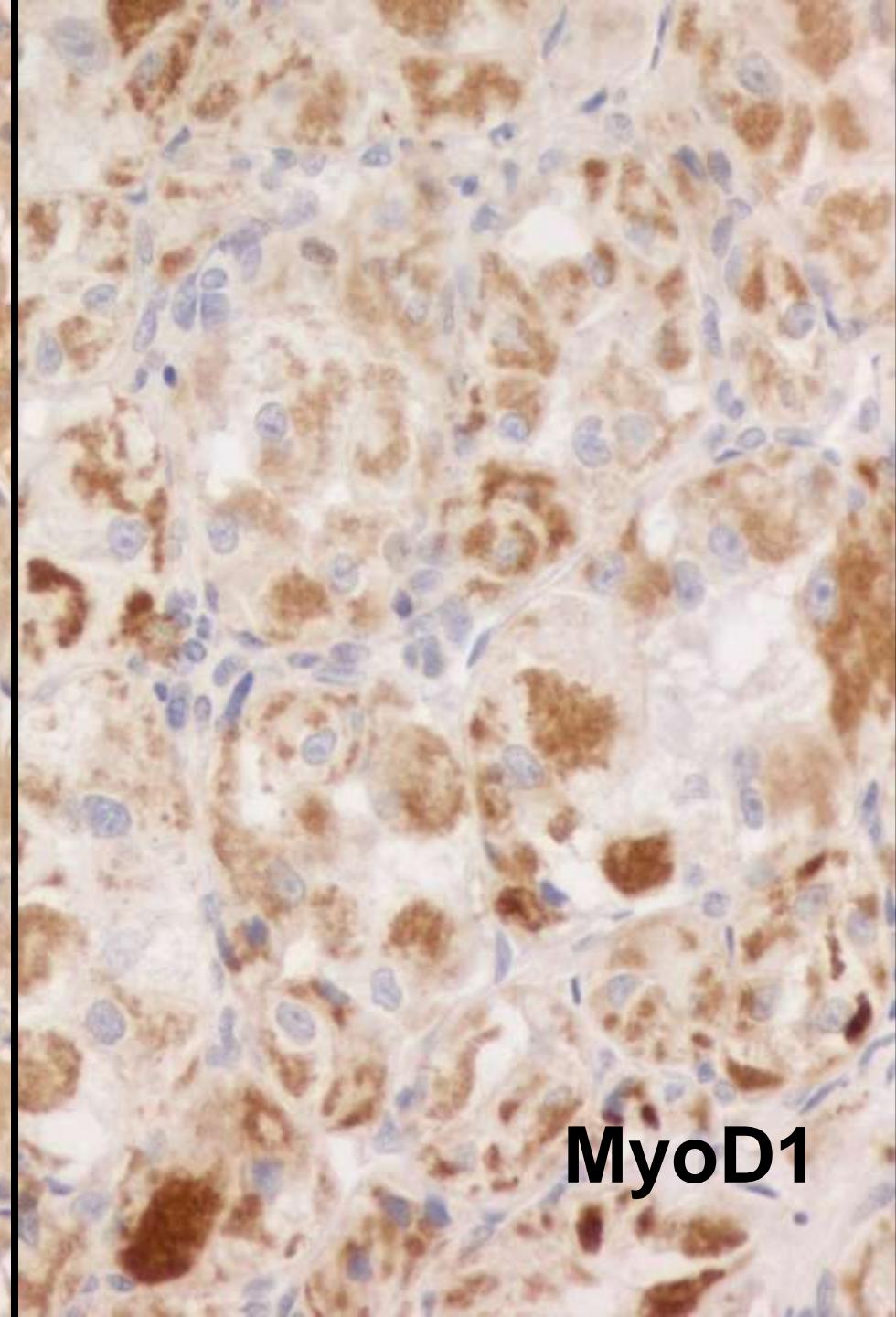


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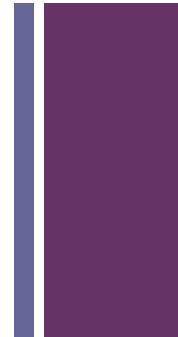
Enolasa



MyoD1

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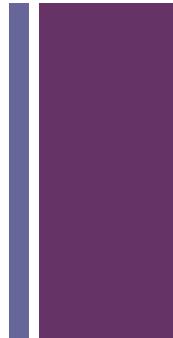
IHQ - negativos



- Citoqueratinas
- EMA
- S100
- Cromogranina A
- CD99
- Actina HHF35
- Desmina



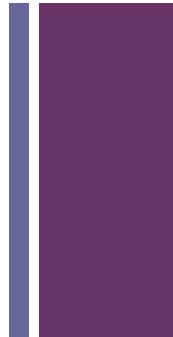
Diagnóstico Diferencial



- Sarcoma epiteliode
- Sarcoma de células claras
- Histiocitosis
- Tumor de células granulares
- Carcinoma de células renales
- Paraganglioma
- Melanoma



SAPB - clínica, epidemiología....

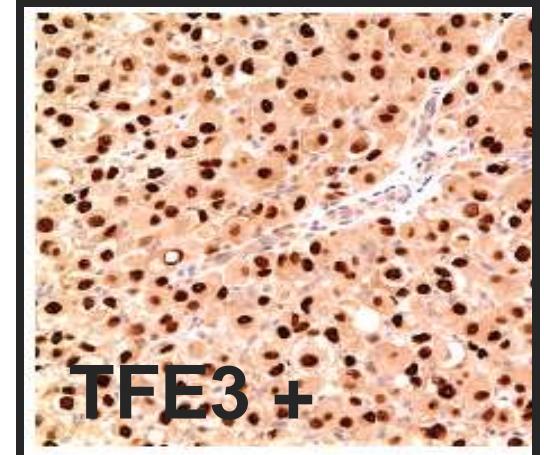
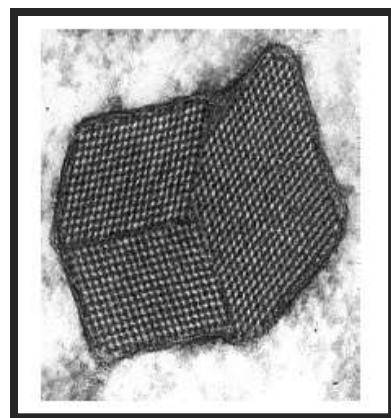
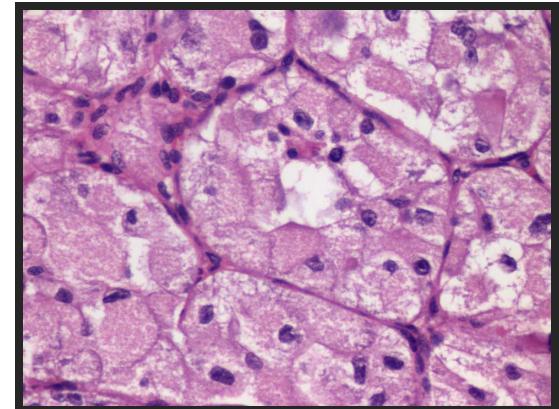


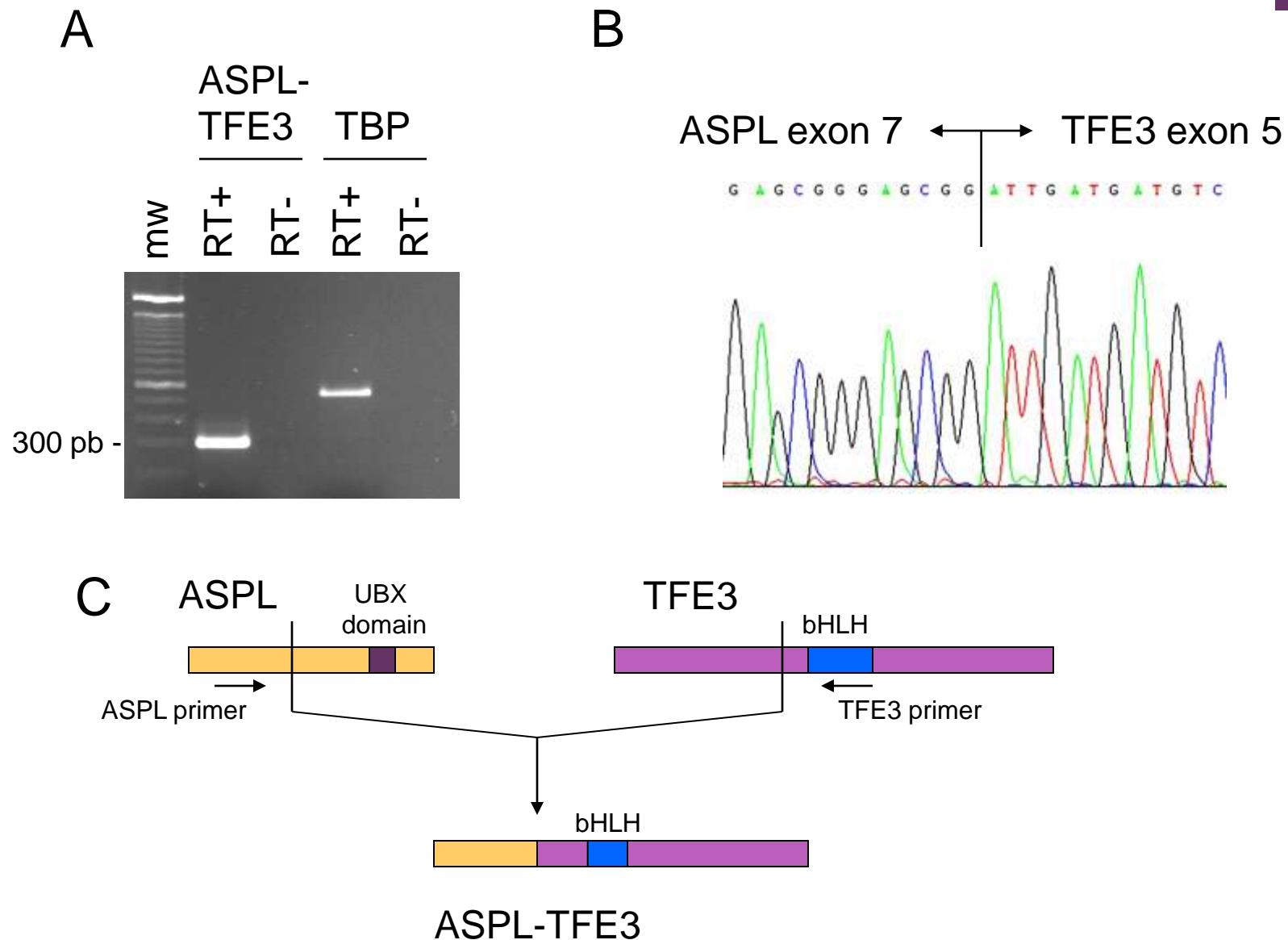
- Descrito en 1952 por *Christopherson et al*
- Menos del 1% de los sarcomas de partes blandas
- Tumor de histogénesis incierta
- Niños y adultos jóvenes, M > H, extremidades inferiores
- Curso clínico insidioso
- Metástasis tardías (pulmón, cerebro, hueso)



SAPB - diagnóstico

- Historia clínica
- Estudios radiológicos (RMN)
- PAAF o biopsia
 - IHQ
 - Ultraestructura
 - Genética







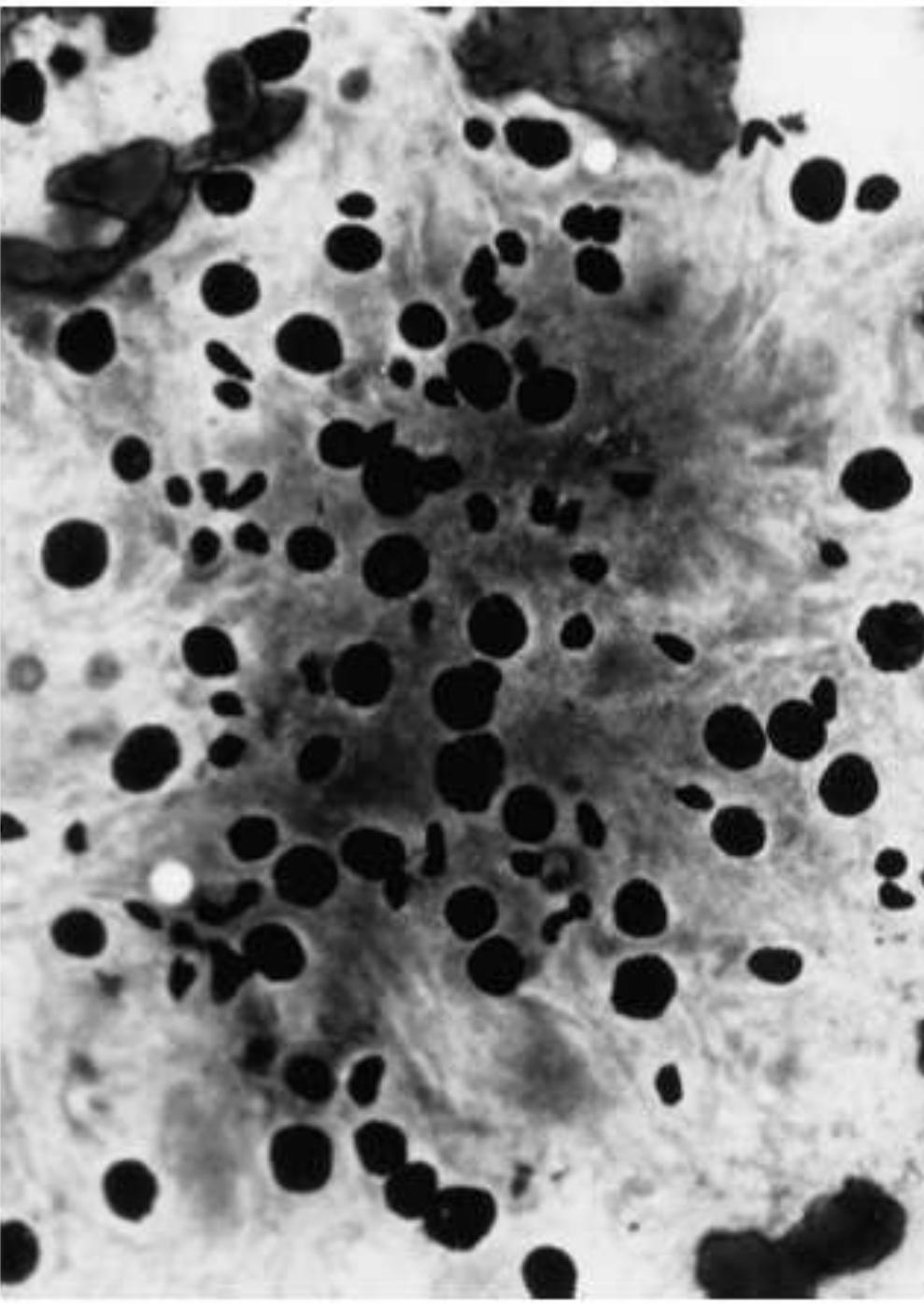
SAPB - tratamiento y pronóstico

- La extirpación completa con márgenes libres debe ser considerada la terapia de elección
- El principal factor pronóstico es la posibilidad de realizar una resección quirúrgica completa
- Ningún rasgo histopatológico permite predecir el pronóstico
- Factores favorables: menor edad, enfermedad localizada y pequeño tamaño del tumor



PAAF en SAPB

- Machhi J, Kouzova M, Komorowski DJ, Asma Z, Chivukala M, Basir Z, Shidham VB. Crystals of alveolar soft part sarcoma in a fine needle aspiration biopsy cytology smear. A case report. *Acta Cytol.* 2002 Sep-Oct;46(5):904-8. PMID: 12365228]
- Logrono R, Wojtowycz MM, Wunderlich DW, Warner TF, Kurtycz DF. Fine needle aspiration cytology and core biopsy in the diagnosis of alveolar soft part sarcoma presenting with lung metastases. A case report. *Acta Cytol.* 1999 May-Jun;43(3):464-70. PMID: 10349382
- Husain M, Nguyen GK. Alveolar soft part sarcoma. Report of a case diagnosed by needle aspiration cytology and electron microscopy. *Acta Cytol.* 1995 Sep-Oct;39(5):951-4. PMID: 7571976
- Drachenberg CB, Papadimitriou JC. Alveolar soft part sarcoma. A case report with correlation of fine needle aspiration and ultrastructural cytologic features. *Acta Cytol.* 1991 Nov-Dec;35(6):746-52. PMID: 1950328
- Shabb N, Sneige N, Fanning CV, Dekmezian R. Fine-needle aspiration cytology of alveolar soft-part sarcoma. *Diagn Cytopathol.* 1991;7(3):293-8. PMID: 1879268]
- Ordonez NG, Hickey RC, Brooks TE. Alveolar soft part sarcoma. A cytologic and immunohistochemical study. *Cancer.* 1988 Feb 1;61(3):525-31. PMID: 3276381]
- Persson S, Willems JS, Kindblom LG, Angervall L. Alveolar soft part sarcoma. An immunohistochemical, cytologic and electron-microscopic study and a quantitative DNA analysis. *Virchows Arch A Pathol Anat Histopathol.* 1988;412(6):499-513. PMID: 3129865]
- Kapila K, Chopra P, Verma K. Fine needle aspiration cytology of alveolar soft-part sarcoma. A case report. *Acta Cytol.* 1985 Jul-Aug;29(4):559-61. PMID: 3861045]
- Nieberg RK. Fine needle aspiration cytology of alveolar soft-part sarcoma. A case report. *Acta Cytol.* 1984 May-Jun;28(3):198-202. PMID: 6587695]



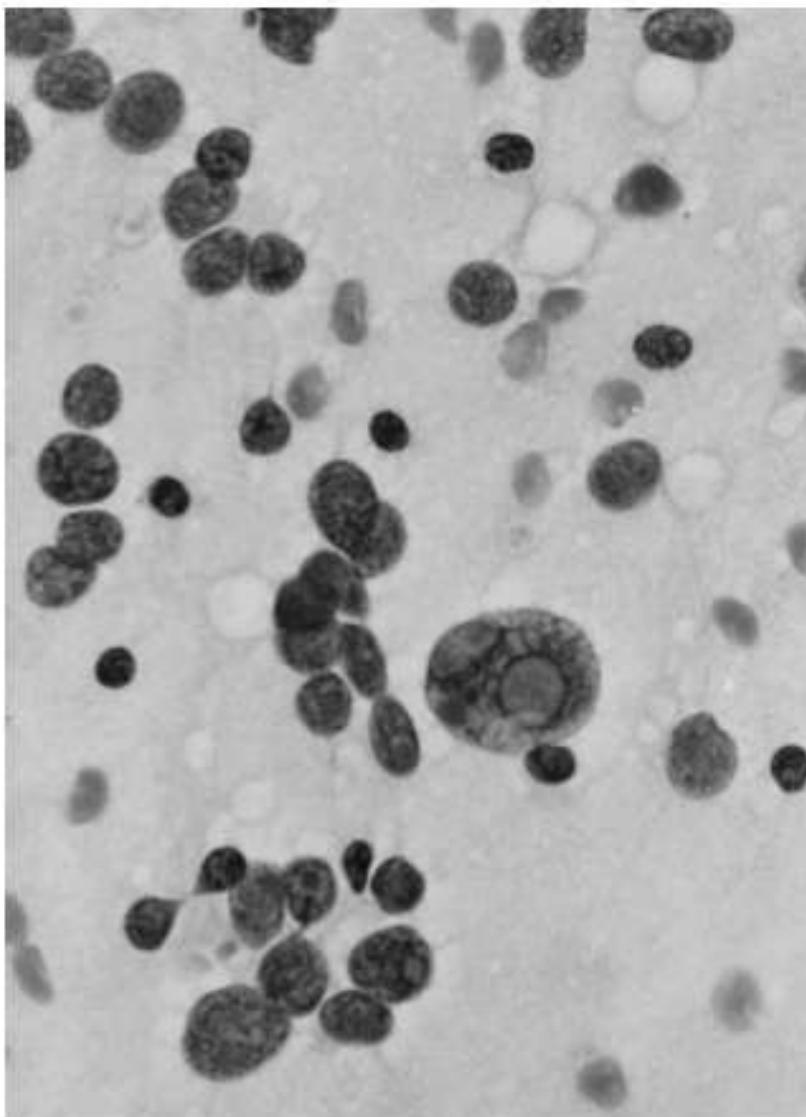
Cytologic Features of Alveolar Soft Part Sarcoma: Report of Three Cases

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Cytopathology of Alveolar Soft Part Sarcoma

A Report of 10 Cases

Paul E. Wakely, Jr, MD¹; James E. McDermott, MD²; and Syed Z. Ali, MD³

Cancer Cytopathology December 25, 2009

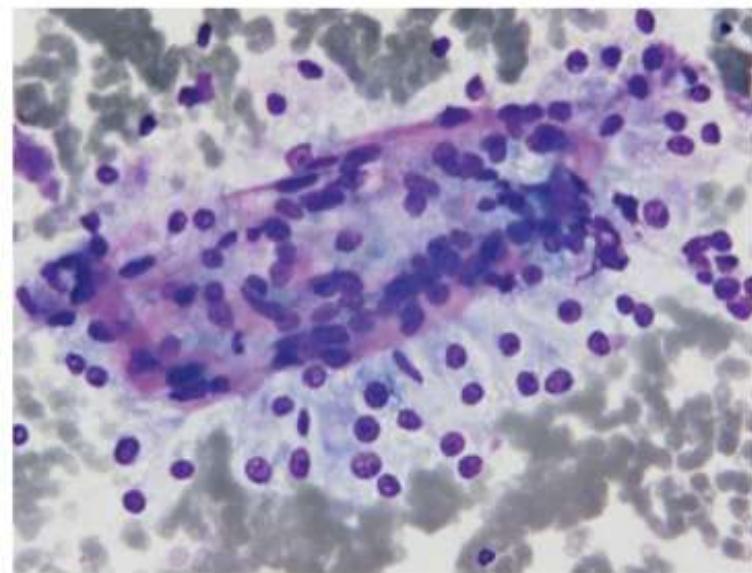


FIGURE 1. Cells are distributed in a vague acinar arrangement with a circular strand of metachromatically staining basement membrane-like stroma (Romanowsky stain, original magnification $\times 40$).

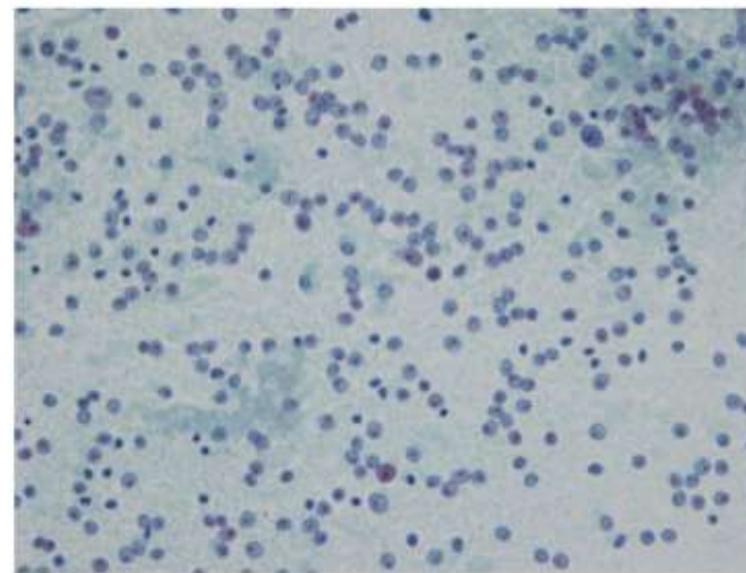


FIGURE 2. Cell fragility leads to a sheet of numerous, dissociated, large, bare nuclei with discrete acidophilic macronucleoli. Note the fragmented granular cytoplasm in the smear background (Papanicolaou stain, original magnification $\times 20$).



SAPB - conclusiones

- El reconocimiento de los rasgos citológicos típicos del SAPB en PAAF permite realizar un diagnóstico preciso de la lesión
- La IHQ permite excluir otras entidades consideradas en el diagnóstico diferencial
- La ultraestructura y la genética son un complemento valioso en el diagnóstico específico de este tumor